CYSTIC TERATOMAS AND TERATOID TUMORS OF
THE CENTRAL NERVOUS SYSTEM IN INFANCY
AND CHILDHOOD

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The teratomas and teratoid tumors of the central nervous system have
been the subjects of numerous publications. Most of these have
been single case reports or systematic studies of the literature. The
tumors have come, therefore, to be regarded as curiosities unlikely to be
encountered by the neurosurgeon or to be available for detailed histological
comparisons. Furthermore, the wide variations in structure and clinical
behavior have made it difficult to draw conclusions in regard to treatment
and prognosis from one or a few cases. At The Children’s Hospital, there
have been 15 neoplasms of this group in 20 years.* With a series as large as
the present one, it is possible to survey a considerable range of histological
structure and of clinical behavior as well as to evaluate different forms of
surgical management.

Since certain tumors of the central nervous system have much histo-
logical resemblance to the group under discussion but present entirely differ-
ent surgical problems, it has been necessary to set arbitrary limits to the
series. Craniopharyngiomas and sacrococcygeal teratomas have been ex-
cluded, as have chordomas. We have included the dermoid cysts, however,
because they present neurosurgical problems similar to those of the cystic
teratomas and because it is our belief that they belong histologically and de-
velopmentally to the same group as the tumors containing more varied
organoid elements.

The 15 teratomas and teratoid tumors were found among 231 neoplasms
of the central nervous system operated upon at The Children’s Hospital
(6.5 per cent). Eight of the tumors were intracranial (4.1 per cent of all
intracranial tumors at this hospital) and seven were intraspinal (18 per
cent of all intraspinal tumors). In interpreting these statistics, it should
be borne in mind that The Children’s Hospital has no patients over 15
years of age. The pathological features of the intracranial and intraspinal
teratomas and teratoid tumors were identical and will be discussed together.
From the standpoint of surgical management, the problems were somewhat
different; the clinical features of the two groups will accordingly be consid-
ered separately.

* Three more patients with intracranial cystic teratomas have been added to the series since the
preparation of this report, making a total of 18.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Onset</th>
<th>Sex</th>
<th>Clinical Data</th>
<th>Location</th>
<th>Associated Anomalies</th>
<th>Operation</th>
<th>Histology</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10-8/12 years</td>
<td>M</td>
<td>Diplopia on looking to left. Increased intracranial pressure.</td>
<td>Third ventricle</td>
<td>None</td>
<td>1. Removal of cystic tumor from third ventricle. 2. Exposure of inoperable residual.</td>
<td>Stratified squamous epithelium, pseudostratified columnar epithelium, gland-like structures with various epithelia, smooth muscle, peripheral nerve, collagenous tissue.</td>
<td>Good immediate result; recurrence after 5 months. Death 10 months after operation.</td>
</tr>
<tr>
<td>2</td>
<td>8 days</td>
<td>M</td>
<td>Gradual increase in size of head.</td>
<td>Frontoparietal</td>
<td>None</td>
<td>Removal of cysts.</td>
<td>Cuboidal epithelium, pseudostratified columnar epithelium, choroid plexus, atypical blood vessels, collagenous tissue.</td>
<td>Temporary improvement. Living 18 months after operation.</td>
</tr>
<tr>
<td>3</td>
<td>10-10/12 years</td>
<td>F</td>
<td>Not related to teratoma.</td>
<td>Choroid plexus</td>
<td>None</td>
<td>Removal intraspinal meningioma.</td>
<td>Whorls of arachnoid cells with psammoma, stratified squamous epithelium, bone with marrow, nerve trunks, smooth muscle, collagenous tissue.</td>
<td>Incidental finding at autopsy.</td>
</tr>
<tr>
<td>5</td>
<td>6 months</td>
<td>F</td>
<td>Hemiplegia, coma, block.</td>
<td>Cerebellar</td>
<td>None</td>
<td>Removal mid-line posterior fossa tumor.</td>
<td>Hair, hair follicles, collagenous tissue; inflammatory and foreign body reaction.</td>
<td>Living 9 years after operation. Slight mental retardation.</td>
</tr>
<tr>
<td>6</td>
<td>5-10/12 years</td>
<td>M</td>
<td>Drowsiness, headache, vomiting.</td>
<td>Cerebellar</td>
<td>None</td>
<td>Evacuation of cyst.</td>
<td>Stratified squamous epithelium, edematous collagenous tissue.</td>
<td>Death 11 days postoperative.</td>
</tr>
<tr>
<td>7</td>
<td>3 years</td>
<td>F</td>
<td>Vomiting, increased pressure</td>
<td>Fourth ventricle</td>
<td>None</td>
<td>Posterior fossa exploration. Removal of cyst.</td>
<td>Stratified squamous epithelium, collagenous tissue.</td>
<td>Symptom-free 4 years postoperative.</td>
</tr>
<tr>
<td>8</td>
<td>Birth</td>
<td>F</td>
<td>Hydrocephalus</td>
<td>Frontal bilateral</td>
<td>None</td>
<td>Right frontal bone flap. Partial removal large tumor.</td>
<td>Serous glands, mucous glands, voluntary muscle, fat, cartilage, collagenous tissue, pseudostratified columnar epithelium, lymphatic vessels, Pacinian corpuscles.</td>
<td>Died 3 days postoperative.</td>
</tr>
<tr>
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<tr>
<td>11 R.M.</td>
<td>5 years</td>
<td>M</td>
<td>Back pain, paralysis lower extremities, C.S.F. block.</td>
<td>Lumbar</td>
<td>None</td>
<td>Laminectomy. Incomplete removal cystic tumor.</td>
<td>Stratified squamous epithelium, sebaceous glands, mucous glands, pseudostratified columnar epithelium, fat, collagenous tissue.</td>
<td>Living with few residual changes 4 years after operation.</td>
</tr>
</tbody>
</table>

* Previously reported by Ingraham.4  
† Previously reported by Ingraham4 and Kubie and Fulton.5
PATHOLOGICAL CONSIDERATIONS

Tables 1 and 2 show the incidence in which mesodermal, entodermal and ectodermal derivatives were represented among the 15 tumors. The simplest contained only stratified squamous epithelium undergoing keratinization and its accompanying stroma of collagenous tissue. In the series, there were tumors varying from these dermoids to neoplasms composed of a great variety of tissues developing from all three germ layers.

As seen grossly, whether at operation or at autopsy, the lesions had much more in common than in histology. A striking characteristic of the solid portions of the lesions in the gross was the texture of the tissue. It was firm and grayish-white, at times with areas discolored yellow by previous hemorrhage. This was an indication that the tumors contained sufficient collagenous connective tissue to determine the texture of their solid portions and was a great assistance to the surgeon in differentiating them from tumors of the glioma group. In the case of neoplasms made up only of keratinizing epithelium and collagenous tissue, the latter seemed best interpreted as a stromal response. When the tumor contained bone, cartilage or other mesodermal derivatives as well as epithelium, it was impossible to tell how much of the collagenous tissue was an element of the tumor itself and how much was stroma, incited to proliferation by other elements.

Some of the teratomas were sharply demarcated from the adjacent nervous structures and could be readily dissected away from them. Others, and especially in the intraspinal group, blended so closely with the brain or spinal cord that no plane of cleavage could be identified. Histologically, the blending was the result of a mixture of reactive gliosis with the connective tissue of the tumor. In one instance (Case 4), the tissue elements were themselves invasive, the neoplasm being composed of a mixture of neuroblastoma and rhabdomyosarcoma. Special features of this and certain other tumors are discussed in the individual case reports.

In all instances, some part of the tumor was cystic. The cysts were usually multiple (Figs. 2 and 12) and varied in size from microscopic structures (Case 3) to huge, tense masses of fluid comprising a large part of the cranial contents (Case 2). The character of the fluid varied greatly, not only from tumor to tumor but also occasionally from cyst to cyst within a single tumor. In some instances (e.g., Case 2), the fluid was pale yellow and clotted. If a small quantity of such fluid was obtained from a cyst or by aspiration through a needle inserted through a suture or burr hole, it would be indistinguishable from that of a gliomatous cyst or late subdural hematoma. When a cyst of a teratoma contained this type of fluid, it was usually present in much larger amounts than would be expected in either of the two other conditions. At times, the fluid in a cystic teratoma was deep yellow and clotted, like the first type. Here again, the differentiation from the fluid of certain gliomatous cysts or of one stage of subdural hematoma could not be made on the character of the fluid alone. More suggestive of the diagnosis of cystic teratoma or teratoid tumor was a cloudy, whitish fluid with a
yellow tinge. This was not cloudy because of blood but because it contained cells. The fluid was not purulent, and few leukocytes could be demonstrated. It was cloudy in most instances because of keratinized cells cast off by stratified squamous epithelium lining the cyst. This is the type expected in dermoid cysts but may also occur in any teratoma in which there is a cyst lined by keratinizing epithelium. In Case 15, the cells in the fluid were columnar and had cilia, as described by Kubie and Fulton.\textsuperscript{5} In still other teratomas, a chocolate-colored, opaque, stringy, gelatinous fluid was obtained; at times, this had a reddish tinge. While it is possible that fluid of such character might be found in a necrotic glioblastoma multiforme, we have seen it only in instances of cystic teratoma or teratoid tumor.

At least two factors help to determine the character of the fluid in these cysts. One is the nature of the cells which form the lining—whether or not they produce keratinized epithelium or other desquamated cells, whether or not they secrete mucin, whether or not they are necrotic. The other factor is the presence or absence of hemorrhage. If there has been recent hemorrhage, a reddish tinge may be expected; if old, the fluid will be yellow or brown. Because of the variability in the character of the fluid in cystic teratomas and teratoid tumors, it is usually unwise to depend wholly on the gross appearance of the fluid when considering the diagnosis in the operating room. A stained smear of the fluid will often show keratinized cells, ciliated epithelium or other elements which make the diagnosis certain at an early stage in the operative procedure. The presence of hair within the cyst when opened at operation, of course, establishes the nature of the lesion without further procedure, but is uncommon (Case 5).

We do not feel that our study of the 15 tumors in the present series contributes anything toward an understanding of the histogenesis of the condition. Neoplasms of this group, in the central nervous system and elsewhere, have long been regarded as the result of belated and abnormal proliferation of pluri- or totipotential cells misplaced in early embryonic life. It is as difficult to find an alternative for this view as it is to obtain direct evidence bearing on its validity. Perhaps one reason why the dermoids of the central nervous system have been set apart in most publications from the other tumors that have here been grouped together is because they contain tissues derived only from the ectoderm along with a connective-tissue stroma, developed in response to the presence of the ectodermal structures. Since the nervous system adjacent to the dermoid is also ectodermal, there has been the suggestion that the factors inducing the development of the tumor occur much later in embryonic, or even in postnatal, life. So far as can be determined from the present series, the dermoid forms one end of a scale of increasing complexity, with the highly varied tridermal tumors at the other.

Vonderahe and Niemer\textsuperscript{6} have published an interesting study of the relationship of intracranial lipomas to teratomas and propose a correlation between the histology of the tumors and the stage of embryonal development at which the tumor originated. They say, "Lipomas developing some time
after gastrulation contain only adipose tissue; others, developing shortly after gastrulation, possess more highly differentiated mesodermal derivatives such as bone; still others, developing before or during gastrulation, possess neuroectodermal as well as mesodermal elements. There is thus a transition from simple lipoma to the more complex teratoid tumors and teratomas.” In the same way, the tumors in this series seem to vary from one another only quantitatively, and in the embryonic potentialities of their cells of origin. Russell7 has shown that certain pinealomas are actually atypical teratomas but can be proved so only on detailed histological study. The histological possibilities of the teratoma group appear to be more varied, the more they become understood.

CLINICAL ASPECTS—INTRACRANIAL GROUP

From the clinical point of view, the character of the symptoms and the problems of differential diagnosis of intracranial teratomas and teratoid tumors were those of any space-occupying lesion in that location. There were local effects, depending on the site of the tumor, and pressure effects, resulting from the increase in volume of the cranial contents. Nothing was encountered that would suggest the pathological diagnosis preoperatively, unless an associated congenital anomaly was present. This occurred occasionally in the intracranial group (Case 5) but more often in the intraspinal group. The presence of a congenital anomaly in a patient with signs and symptoms of a space-occupying cerebral lesion does not establish the diagnosis of teratoma or teratoid tumor, but makes the chances of finding this lesion somewhat greater than in its absence. It would be unusual to find a glioma in a patient with a skull defect or superficial malformation of the head, but hemangiomas or similar tumors might well be encountered.

On the whole, the symptoms of the tumors in the series developed slowly (e.g., Case 5) not only because the neoplasms were slow in growth but also because children tolerate increased intracranial pressure better than adults. There was no evidence that the cysts filled suddenly with fluid. While there was the possibility of a large hemorrhage into a pre-existing cyst with consequent enlargement of the volume of the tumor, this was uncommon. Case 4 was an exception, in that symptoms developed rapidly. This was the result of the histological character of the tumor, which was invasive and contained numerous mitotic figures.

Roentgenograms of the skull might possibly show calcified areas suggesting the presence of bone or other organoid structures in tumors of this group. In none of the patients in this series was it possible to make a definite diagnosis of the type of lesion from the roentgenograms. There was evidence of increased intracranial pressure and, in some, internal hydrocephalus was present. Ventriculograms or encephalograms demonstrated filling defects indicating the size and location of the tumors.

The intracranial cystic teratomas and teratoid tumors varied a great deal from the operative point of view. Some were clearly demarcated by a con-

FIG. 2. Case 1. A cystic teratoma after removal from the third ventricle. There are several cysts of different sizes separated by dense tissue.

FIG. 3. Case 1. The histological appearance of an area from the tumor in Fig. 2. A cyst lined by keratinizing epithelium and another lined partly by columnar and partly by stratified squamous epithelium, and a group of gland-like structures are separated by collagenous tissue. Phloxine-methylene blue stain, camera lucida drawing, $\times 125$. 
nective-tissue capsule and could be readily dissected away from the adjacent nervous structures (e.g., Cases 1 and 5). On the other hand, the tumor in Case 4 was so intensely adherent to the brain substance that no plane of cleavage could be found. Most instances fell between these extremes. However, in dealing with such tumors as those in Cases 2 and 8, it was impossible to define the relation to the nervous tissue because the tumors filled a large portion of the cranial cavity. The easily separable tumors offer a good prog-

Fig. 4. Case 1. A complicated duct-like structure with many branches. Some of the cysts appeared to have originated from distension of similar cell-groups. Phloxine-methylene blue stain, X112.

nosis. Much less can be done for the neoplasms intimately adherent to the nervous tissue, though even here much can be accomplished unless the tumor is histologically malignant or unless it is of such large size that any surgical approach is hopeless.

Of the 8 patients with intracranial teratoma or teratoid tumor, 1 is living more than five years after operation; 2 are living for periods less than five years; and 5 are dead.

The following case reports indicate the range of variation in clinical behavior and pathological findings in this group of tumors.
Abstract Case 1, C.R.W. (296,488). This patient was referred to the hospital at the age of 10 11/12 years with a history of frontal headaches associated with sinusitis and ptosis of the left eyelid. For 3 months there had been double vision on looking to the left. The positive findings on physical examination were ptosis of left eyelid, papilledema with retinal hemorrhages, and marked tachycardia with a loud systolic murmur. There was moderate leukocytosis and elevation of the sedimentation rate. Roentgenograms showed a small area of calcification in the pineal region slightly to the right of the midline. Under avertin-ether anesthesia exploration was attempted but there was a marked fall in blood pressure before the scalp incision had been completed. He was very soon in a state of severe shock, operation was interrupted and the blood pressure was restored to normal levels only after the most heroic measures including the intravenous administration of large amounts of eschatin. At a later date he was given eschatin preoperatively as well as throughout the procedure and it was possible to turn down the bone flap and make a subtemporal decompression. Operation was again interrupted as signs of shock appeared. At a third session it was possible to amputate the right occipital pole, traverse the corpus callosum and remove an intact partially cystic tumor from the 3rd ventricle (Fig. 2). It is interesting to note that the pulse rate immediately became slower, blood pressure stabilized, and there was no suggestion of shock after the removal of the tumor. It was assumed at this time that the tumor had been entirely removed and the patient was sent home in good condition 3 weeks after operation. He returned to school and was symptom-
free for 5 months, at the end of which time he complained of headache and he was readmitted to the hospital with high intracranial pressure. Ventriculography revealed the presence of a large mass in the 3rd ventricle which was found to be continuous with the adjacent brain tissue and quite inoperable. It seemed evident at this time that the large mass previously removed was a relatively free and benign portion of a larger invasive tumor. This impression was later confirmed by autopsy. The temporary relief of block in circulation of cerebrospinal fluid and relief of hypothalamic pressure had been sufficient to restore the patient to a normal physiological level.

Pathological Study. The soft, grayish-pink tumor measured 3.8×3.2×2.3 cm. (Fig. 2). The cysts ranged from 0.2 to 1.5 cm. in diameter. Their walls were thin but varied in translucence. A few hard, white nodules, 0.8×0.8×0.4 cm. in size, were found on one surface of the mass. On section, the tissue had a lobulated appearance. The cysts contained clear fluid and were separated by fibrous septa, which were almost gristly in consistency.

The microscopic sections showed a variety of structures. The larger cysts were lined by stratified squamous epithelium with well marked keratinization, despite the clear appearance of the fluid. Other smaller cystic spaces consisted of duct-like structures lined by pseudo-stratified columnar epithelium resembling that of the respiratory tract. The epithelium was much infolded and at times showed an abrupt transition to the stratified squamous type (Fig.
3. Small glands of irregular outline were the dominant tissue element in certain areas. They were lined by low cuboidal epithelium. A few other spaces were lined by various types of epithelium, at different places resembling somewhat colonic, transitional and ependymal epithelia. The collagenous stroma contained bundles of smooth muscle and peripheral nerve. At times, the connective tissue was arranged like that in neurofibromas. Lymphocytes were present in fairly large numbers in the stroma and there were a few cells with large pale nuclei suggesting reticulum cells. No mitoses or areas of invasion were found (Figs. 3, 4, 5 and 6).

Comment. Serious physiological instability was caused by the presence of the tumor in the neighborhood of the third ventricle and pressing upon the hypothalamus. The surgery necessary for the removal of the neoplasm would probably have been impossible without the use of adrenal cortical extract. When this was given in amounts sufficiently large to restore the patient's physiology to somewhere near normal, the operative procedure could be carried out safely. The remarkably small number of successful operations on tumors of the pineal region serves to emphasize the gravity of surgery in this location when the physiological disturbances have not been corrected.

Despite the encapsulation of the tumor and its benign appearance histologically, there was evidence of recurrence five months after operation and death one month later. In the interval, the patient was in good health. From the standpoint of useful function, therefore, the quadratic defect caused by the removal of the occipital lobe was less serious than the choking of the optic discs, which was relieved by the operative procedure.

Abstract Case 2, R.T. (290430). This infant was admitted to the hospital at the age of 8 days because of a gradual increase in the size of the head. Delivery was normal and at full term. Nothing unusual was noted immediately after birth but within a few days it was apparent that the head was growing rapidly, particularly the "back" of the head. The patient lay in a position of opisthotonos, the right side of the head down, moving the extremities vigorously. The fontanelles were large and the sutures widely separated, the left coronal more than the right. The right ventricle was tapped and pressure found to be elevated. Ventriculography showed dilatation of the ventricular system with a shift to the right. A large bone flap was turned down and a huge cystic mass was exposed (Fig. 7). The multilocular cyst appeared to fill the major portion of the left half of the cranial cavity. The cysts were evacuated and part of the wall removed but in view of the extensive damage to the brain already evident no attempt was made at complete removal.

Pathological Study. The material removed for biopsy consisted of a deep red, opaque piece of cyst wall, measuring 0.9×0.8 cm. The outer surface was smooth, while the grayer inner surface had a small brownish, nodular excrescence upon it and was traversed by several large, congested blood vessels.

Microscopically, the wall of the cyst was found to consist of coarse collagenous tissue and large endothelial-lined spaces. At a few points, the surface of the cyst was lined by a single layer of low cuboidal epithelium which immediately overlay groups of large blood vessels. In most portions of the cyst wall, however, the connective-tissue fibers formed the lining. Projecting into the cavity of the cyst were many long, fine, filamentous processes covered with epithelium similar to that of the cyst wall. Within most of the processes were distended blood vessels, being separated from the epithelium only by their endothelial layers. The picture, therefore, simulated in many respects a congested choroid plexus. A probable diagnosis of teratoid tumor was made on the basis of a cystic neoplasm with epithelial elements and an abnormal vascular tissue.
Comment. Case 2 illustrates the type of teratoid tumor for which surgery offers least. Occupying a large portion of the cranial cavity, the tumor could not be attacked radically. The most that could be done was to evacuate all accessible cysts with the hope of palliation.

Abstract Case 3, R.B. (154906). A 10 10/12 year old girl was admitted to the hospital because of weakness of the right arm of 2 weeks' duration. Physical examination showed weakness of the right leg, diminished sensation over the right forearm, right exophthalmos and choroiditis. There was a café au lait spot on the trunk and a small cutaneous nodule in the left axillary region. This was removed and identified as a neurofibroma. Roentgenograms showed bilateral calcification of the choroid plexus. Laminectomy was done and a meningioma removed from the cervical canal. Subsequently a cerebellar exploration was done to relieve a block caused by what appeared to be a tumor of the right cerebellar hemisphere, sections of which showed gliosis but included a number of psammoma bodies. The patient died 2 years after the operation.

Pathological Study. The final autopsy diagnoses were: brain tumors, multiple; meningioma of cervical spinal cord; teratoid tumor of choroid plexus; congenital defects in septum pel- lucidum; sclerotic atrophy of cerebellum; syringomyelia; cutaneous neurofibroma.

The present study was concerned only with the teratoma of the choroid plexus, an incidental finding in a patient with several clinically more important lesions of the nervous system. This description is confined to the region of the lateral ventricles and choroid plexus. The lateral ventricles were slightly distorted, giving the appearance that the right basal
nuclei were pushed upward and medially, decreasing the size of the right lateral ventricle. They were not dilated. The ependymal surfaces were smooth. The choroid plexus in both lateral ventricles had a rounded, nodular appearance instead of the usual frond-like structure (Fig. 8). Between the lateral ventricles, there were multiple small defects in the septum pellucidum.

Some few portions of the choroid plexus had a normal histological appearance. The knob-like projections seen in the gross were composed of large nests of arachnoid cells arranged in whorls. There were numerous calcified psammomas within the arachnoid cell clusters. The appearance was that of a series of minute meningiomas, adjacent but usually discrete. However, there were other tissue elements near the arachnoid cell nests. A small cyst was lined by stratified squamous epithelium; its lumen contained desquamated cells which were partially keratinized (Fig. 9). At another point, a mass of bone was found. This was well calcified and had clearly defined Haversian systems. Fatty marrow was seen within the bone and small clusters of hematopoietic cells were present (Fig. 10). Several groups of myelinated nerve trunks lay within the tumor and were regarded as a part of it, since there was no evidence of invasion and since nerve trunks of this type would not be expected in the region where they were found. There was a stroma of collagenous tissue throughout the neoplasm. In spite of the resemblance of large parts of the tumor to meningioma, it was classified as a teratoid tumor because a cyst lined by stratified squamous epithelium, nerve trunks and bone with marrow were also present.

Comment. The tumor in Case 3 was an incidental finding in the midst of bewildering neuropathological abnormalities. It accounted for the calci-
Cystic Teratomas and Teratoid Tumors

of two encountered among 231 tumors of the central nervous system at The Children's Hospital.

From the pathological point of view, the tumor in Case 3 was of considerable interest. Arachnoid villi are present normally along the septum pellucidum and hyperplasia of arachnoid villi is a fairly common finding, but the masses in this tumor achieved a degree of hyperplasia not seen under ordinary conditions. The presence of other organoid elements in association with the meningioma-like cell nests made the diagnosis of teratoid tumor necessary. No other tumor in this series had clusters of arachnoid cells among its tissue elements.
Abstract Case 4, A.M. (260086). A 10 7/12 year old boy was referred into the hospital because of vomiting spells of 3 months' duration. He vomited once or twice a day throughout this time and had lost 10 pounds. Vomiting was not associated with nausea and it was not projectile. Eyegrounds were normal. Roentgenological examination of the gastrointestinal tract showed marked prominence of the rugae interpreted as hypertrophic gastritis (Fig. 11). On the 18th hospital day he complained of double vision and shortly thereafter minimal choking of the discs could be seen. A diagnosis of midline cerebellar tumor was made and exploration carried out. A soft, reddish-gray tumor mass was exposed by incising the vermis and the major portion of the tumor removed but the remainder was clearly continuous with the wall of the 4th ventricle. The patient made a satisfactory recovery, was given extensive radiation therapy and was free from symptoms for nearly a year. He then went downhill very rapidly and died outside the hospital.

Pathological Study. The portion of tumor removed at the first operation was composed of soft, gray fragments of tissue measuring in aggregate $2 \times 2 \times 0.5$ cm. and a thin piece of membrane, $1.2 \times 0.5$ cm.

Histologically, the tumor was made up of two types of cells. The first type had oval, somewhat vesicular nuclei closely packed with chromatin and a very small amount of cytoplasm; mitotic figures were occasionally present. Occasionally neurofibrils could be demon-

![Fig. 10. Case 3. Bone with fatty marrow containing small groups of hematopoietic cells in a teratoid tumor of the choroid plexus.](image-url)
strated in these cells. The cells grew in sheets and were accompanied by a definite but scanty connective-tissue stroma. The second type of cell was found where the tumor had invaded the cerebellum and was mixed with otherwise relatively normal cerebellar tissue. Some of these cells were of medium size with darkly stained nuclei. Numerous ones were much larger—two or three times as large as Purkinje cells. The giant cells had a single nucleus surrounded by a large amount of relatively clear cytoplasm in which cross striations could occasionally be seen. In some places, there were cells with short, stubby extensions of cytoplasm containing very clearly defined cross striations. Mitoses were frequent in cells of this sort. This second type of cell was that of a malignant tumor of striated muscle. It appeared that the first type

![Fig. 11. Case 4. Roentgenogram showing hypertrophic gastritis in a patient with a malignant intracranial teratoid tumor. Note the greatly enlarged rugae. The "gastritis" did not change significantly after successful surgical attack on the tumor.](image-url)

of cell was of neurogenic origin and completely different from the second type. The presence of two distinct types of cells in a neoplasm with a cyst led to the diagnosis of malignant teratoid tumor.

*Comment.* This tumor differed greatly from the other members of the series in being composed of cellular elements that were highly undifferentiated and invasive. The frozen section made during operation showed only poorly differentiated cells with many mitotic figures. Both the operative
appearance of the tumor seeding the meninges and the frozen section were suggestive of medulloblastoma. The permanent sections left no doubt as to the presence of further malignant elements and placed the lesion in the category of invasive teratoid tumors. One type of cells was the neuroblast in various stages of differentiation. The other was a striated muscle cell as seen in rhabdomyosarcoma. Both mesodermal and ectodermal elements participated in malignant growth, and no adult tissue structures were found.

The tumor in Case 4 was hopeless from the surgical standpoint, not because of large size or of inaccessible location, but because of its pattern of cellular growth. This is the second example of a malignant teratoid tumor in the series. The rarity of such lesions precludes any evaluation of the efficacy of treatment.

It is interesting to consider in this instance the relationship of this infiltrating fourth ventricle tumor to the hypertrophic gastritis. There was no vomiting during the postoperative survival period until the terminal phase. It must be assumed therefore that the prominence of the gastric rugae was the result of the long-continued vomiting before operation or was merely an incidental finding. Other children who have been vomiting for a long period of time have not ordinarily shown these changes. In an attempt to clarify the relationship, a second G.I. series was done six months after operation when the boy was perfectly well. At that time the findings were exactly the same as they were preoperatively.

Abstract Case 5, C.A.G. (208099). This patient was admitted in coma following the sudden development of paralysis of the left arm and leg. Examination showed the head to be enlarged (19 1/4 in. circumference), fontanelle tense, and just below the external occipital protuberance there was a small red area which became dimpled when the skin was moved. Fundi showed blurring of the disc margins. Roentgenograms showed separation of the sutures. Ventricular taps revealed enlargement of both lateral ventricles and a block between the ventricles and the spinal subarachnoid space. Following these diagnostic taps the block seemed to be temporarily relieved and the patient became alert and responsive. At this point a positive Babinski response could be demonstrated on the right. A cerebellar exploration was carried out and a firm, pale tumor of variable consistency exposed. It lay chiefly in the midline but extended somewhat into the left cerebellar hemisphere. In the depth of the tumor there were several long blonde hairs. The mass was completely removed.

Pathological Study. There were eleven fragments of tumor tissue available for examination, varying from 1.2 × 1.0 × 0.5 cm. to 0.4 × 0.3 × 0.2 cm. in overall dimensions. The portions representing cyst lining were smooth and grayish-pink in some parts and in others grayish-brown with strands of hair attached to the surface. The solid parts were gray or grayish-red with considerable variation in texture and appearance.

Histologically, the cyst was lined by rather coarse connective tissue with well formed hair follicles in the layer immediately subjacent. There was a rich infiltration with inflammatory cells and some multinucleated giant cells of foreign body type were in this region. These features were interpreted as reaction to keratinized cells, even though stratified squamous epithelium could not be demonstrated. Definite proof of the original character of the cyst lining was not obtained, and the conclusion as to its nature was based on the presence of hair, hair follicles and an inflammatory reaction similar to that seen in degenerated sebaceous cysts of the skin. The other parts of the tumor were composed of collagenous tissue with some infiltration by lymphocytes and mononuclear cells as well as polymorphonuclear leukocytes in a few
foci. There was considerable variation in degree of vascularity, but all blood vessels were essentially normal in structure. No other types of tissue were encountered. There was no evidence of invasion, dedifferentiation of cells or areas of rapid growth. Portions of adjacent brain tissue removed with the tumor showed extensive reactive gliosis.

Comment. This tumor was the only one in the series in which hair was demonstrated within the cyst at operation and the diagnosis made thereby without further notice. From the surgical point of view, the lesion was a simple, benign, cerebellar tumor. The patient remained well for ten years. She now has no objective residual changes but appears somewhat retarded.

CLINICAL ASPECTS—INTRASPINAL GROUP

As with the intracranial teratomas and teratoid tumors, the symptoms produced by the intraspinal members of the series were those of a space-occupying lesion at the level involved. With the intraspinal tumors, however, the association of a congenital abnormality was the rule rather than the exception (Cases 10, 12, 13, 14, and 15). Since these anomalies affected the vertebrae, they could be easily demonstrated roentgenologically and often by physical examination as well. The bony defects might or might not occur at the same level as the tumor. They were of considerable importance in suggesting the diagnosis of teratoma or teratoid tumor preoperatively. Usually the anomalies were compatible with normal function of the spine once the tumor had been removed and seldom required operative treatment. Occasionally, also, naevi or pigmented spots in the skin at the level of the lesion were regarded as somewhat suggestive of teratoma or teratoid tumor. None of these congenital abnormalities, when associated with signs of a space-occupying lesion, was conclusive proof of the nature of the lesion, but was merely suggestive. We have not regarded congenital anomalies of other organ systems, such as the cardiac or genito-urinary systems, as being in any way related to the incidence of the tumors under consideration.

Roentgenograms were chiefly useful in this group of patients for the demonstration of the presence and extent of an anomaly of the vertebral column. It is conceivable that a definite diagnosis might be made roentgenologically by the demonstration of calcified organ-like portions of the tumor. As in the intracranial group, no instance of this has occurred in the present series. Other roentgenographic studies for further localization and definition of the extent of the lesion were made as indicated as with any intraspinal tumor.

When the intraspinal teratomas and teratoid tumors were exposed at operation, they were found to vary greatly from one another. They might be extradural or intradural. The neoplasms were sometimes encapsulated and could be removed without injury to the spinal cord. The effects of these tumors on the spinal cord were rather like those of neurofibromas in similar locations. They were those of pressure and progressive interference with the vascular supply of the spinal cord. In other instances (e.g., Cases 10 and 11), a large cystic mass was found, the wall of which was not separable from the
spinal cord over a considerable area. These were treated by the evacuation of the cyst and removal of as much of the wall as could be carried out without damaging the nerve tracts further than had already been done by the tumor. In Cases 10 and 11, only a flat ribbon of spinal cord was left, which looked so seriously damaged that any return of function could hardly be antici-

![Image of spinal cord tissue](image_url)

Fig. 12. Case 9. A, exterior, and B, cut surface of a cystic intraspinal teratoma.

pated. In spite of the discouraging appearance of the spinal cord at the end of the operation in each patient, there was complete recovery of function in Case 10 and almost complete recovery in Case 11. Furthermore, the recovery has been maintained for ten years in Case 10 and four years in Case 11. Even though some of the cyst was certainly left behind in each patient, there has been no clinical evidence of recurrence at any time. This appears to be the most encouraging experience to emerge from the present study. No matter how discouraging a cystic intraspinal teratoma or teratoid tumor may appear when exposed at operation, it is in the best interests of the patient to evacuate all accessible cysts and to remove as much of the solid portion as can be accomplished without traumatizing the spinal cord. Even
though the final appearance of the operative field is such as would lead the surgeon to give a very grave prognosis if the lesion had been any other form of neoplasm, it is often possible for patients with partially removed teratomas and teratoid tumors to recover spinal cord function and live for many years—perhaps indefinitely—without clinical evidence of recurrence.

Of the 7 patients with intraspinal teratomas and teratoid tumors, 2 are living more than five years; 4 are living for periods less than five years; and 1 is dead.

The following case reports are representative of the series of intraspinal cystic teratomas and teratoid tumors.

Abstract Case 10, C.S. (199355). This case has been reported in another publication.4

The patient was admitted to the hospital at the age of 2 5/12 years because of weakness of the arms and legs of 72 hours' duration. Seventeen days before admission she had been seen and treated for pain in the neck which appeared after a fall from the kitchen table. Roentgenograms made at that time showed absorption of the lamina of the 5th cervical vertebra interpreted as osteomyelitis. A Thomas collar had been applied for immobilization. At the time of entry she was acutely ill with flaccid paralysis of arms and legs with anesthesia except

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Fig. 18. Case 18. Camera lucida drawing of a hair follicle from a cystic intraspinal teratoma.
over the shoulders and neck, where there was hyperesthesia. A diagnosis of compression of the cervical cord was made and laminectomy was done (Dr. R. U. Light). Lumbar puncture done at the time of operation revealed a complete block. A cystic mass was exposed, extending from the 2nd cervical to the 1st thoracic vertebrae. Ten to 12 cc. of yellow, gelatinous fluid were removed from the cyst and a nodule of firm tumor about 1½ cm. in diameter excised. This left the cervical portion of the cord merely a flat ribbon lying on the anterior wall of the spinal canal. Postoperatively there was rapid and complete return of function.

Pathological Study. Two portions of solid tumor were available for study. One measured 1.2×1 cm.; it was smooth, moist, and dull pinkish-gray with streaks of red. There was a small cystic area in the center. The other measured 1×0.5 cm.; it appeared granular and was irregularly mottled pink and pinkish-yellow.

Histologically, the tumor was composed of several varieties of tissue. The large cyst was lined by pseudostratified ciliated columnar epithelium, which was thrown into folds in places and which formed crypts in others. Beneath the epithelium, there was a loose network of collagenous connective tissue interspersed with numerous mucous glands. The region, therefore, resembled somewhat portions of the respiratory tract. Other areas were composed of hyaline cartilage, bundles of nerve fibers, fat, and glands with large lumina lined by cuboidal epithelium. There were several regions filled with astrocytes and microglia. These were located at or near the periphery of the tumor and were regarded as reactive gliosis, but the possibility that some of them were an integral part of the tumor could not be entirely excluded. In the region of gliosis, the blood vessels had swollen endothelia and very thick adventitia. There were no areas of rapid growth or invasion in any part of the material studied.

Section of a portion of dura removed from a region near the tumor showed congestion
Comment. The history of this patient illustrates the indolent growth and satisfactory clinical course of certain incompletely removed intraspinal teratomas. In spite of the fact that tumor was left behind at operation, there is still no evidence of recurrence after ten years. Histologically, the lesion was a teratoma without any areas of rapidly growing cells. The presence of ciliated epithelium relates it to the cases reported by Kubie and Fulton.

Abstract Case 11, R.M. (262598). This patient was admitted with complaint of low back pain and difficulty in voiding. Examination showed a thin, rather undernourished boy in no distress. His spine was held rigidly fixed and both lower extremities were flaccid with marked atrophy and little muscle tone. There was hyperesthesia corresponding to the distribution of the 5th lumbar segment. Lumbar puncture done at the beginning of laminectomy showed a complete block with a very high protein. A cystic mass was exposed extending from the 1st lumbar segment to the lower end of the canal. The cyst was evacuated of thick brown, gelatinous material, and the wall and some solid tumor removed. It was impossible to do a complete extirpation without causing further damage to the lumbar cord. He was discharged home very much improved 21 days after operation.

Pathological Study. Studies were carried out on a portion of soft, friable, pinkish-red tissue 0.5 × 0.3 × 0.2 cm. and a smaller fragment 0.1 in diameter.

Histologically, the tissue was composed of collagenous connective tissue of variable density in which were embedded discrete islets of keratinizing stratified squamous epithelium. In these regions, there was a large number of mucous glands and a moderate number of sebaceous glands but no hair follicles were present. There were a few areas of pseudostratified columnar epithelium. Fat cells were found in groups at numerous points. A smear made from the yellowish, mucilaginous cyst fluid showed erythrocytes and cellular debris. No keratinized cells were demonstrated.

Comment. Despite the extent of the lesion, the fact that tumor tissue was left at operation, and the severe compression of the spinal cord, this child is free from recurrence and has only very slight residual neurological abnormalities four years after operation. Histologically, the tumor was composed predominantly of keratinizing stratified squamous epithelium and skin appendages. It would, therefore, be regarded as a dermoid cyst had it not been for a few small groups of pseudostratified columnar epithelium. This is further evidence that dermoid cysts of the central nervous system should be considered with the teratomas and teratoid tumors. The presence of fat also links this tumor with the series described by Vonderahe and Niemer.

SUMMARY

Fifteen instances of cystic teratoma and teratoid tumor of the central nervous system in children have been described. Eight of these were intracranial and 7 intraspinal.

All contained one or more cysts. The tumors contained tissue elements in their solid portions varying from only keratinizing epithelium to mixtures derived from all germ layers. One tumor, made up of undifferentiated nerve and striated muscle cells, was highly malignant, freely invading the cerebellum and seeding the meninges.
There was nothing to suggest the diagnosis preoperatively unless an associated congenital anomaly was present. The diagnosis could be established at operation by demonstration of hair, by finding keratinized or ciliated cells in the cyst fluids or by frozen section.

The use of adrenal cortical extract proved to be a valuable adjunct in an operation at which a teratoma of the third ventricle was removed.

Two of the patients are living five years or more after operation without evidence of recurrence; 5 for less than five years without evidence of recurrence; 2 have evidence of recurrence and 6 are dead.

In two patients, partial removal of intraspinal teratomas was followed by excellent return of function without recurrence of symptoms over a long period of years.

To say a patient has a teratoma or teratoid tumor of the central nervous system is no more indicative of the clinical result to be expected than to say the patient has a tumor. Some are simply removed, some are cured after a series of operations, some remain quiescent even though incompletely removed, some are so extensive that radical surgery is impossible, and some are invasive and malignant.

REFERENCES