Epidermoid (Mixed) Tumors of the Central Nervous System

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It is generally conceded that the rare, mixed tumors of the central nervous system arise from epithelial cell rests. 5,17, 21,23,30

According to their origin from one or more germ layers, these tumors are usually subdivided into epidermoids, 6 which contain only epidermal tissue and dermis and lack dermal structures; dermoids, which consist of epidermis, dermis, and dermal glands; teratoids, 28 which contain recognizable tissues of two germ layers; and teratomas, 29 whose characteristic cells are derived from all three germ layers. In many of these complex tumors 26 derivatives from one or two germ layers tend to overgrow the others, so that the final analysis of the total number of germ layers present may be difficult.

Review of Published Cases

Intraspinal Tumors. The intraspinal epidermoid and dermoid tumors occur less frequently than the cranial variety, and the dermoid less often than the epidermoid. In their collection of 40 tumors of this sort, Boldrey and Elvidge 6 mentioned five intraspinal cases. Ingraham and Bailey 1,13 reported 21 of these tumors treated at the Children's Hospital in Boston. Eight were intracranial and 13 intraspinal, the latter including 5 dermoids, 5 teratoids, and 3 teratomas.

Isolated cases 5,12,13,16,19,22,26 brought the total of intraspinal epidermoid tumors to 90 in 1962 when Manno, et al., 18 reviewed and reclassified the group. Features such as long duration of symptoms, a radiographic film of diffuse enlargement of the spinal canal, 7,11 or the presence of a spina bifida occulta or a pilonidal sinus are pathognomonic of the spinal epidermoid.

Rewcastle and Francoeur 24 reported sex chromatin studies, which suggested that these tumors were not caused by a simple misplacement of normally-developing somatic cells, but rather were the result of a teratomatous mal-development. However, Van Gilder and Schwartz 29 have produced dermoid and epidermoid tumors from direct skin implants along the neuraxis of albino rats, evidence which supports the epithelial cell rest theory.

Crani al and Intracranial Tumors. Rand and Reeves 21 reported 21 intracranial epidermoids and two dermoids. In 1942, Black 3 found 205 reported cases of intracranial epidermoids. Grant and Austin 8 published 22 cases of cranial epidermoids, and found the most common sites of the intradural group to be the cerebellar pontine angle and the parapituitary region. Baumann and Bucy 2 described three cases of paratrigeminal tumors in relatively young persons. Scott 27 published a case of an epidermoid of the lateral ventricle, and Keville and Wise 14 recorded 17 patients with intracranial epidermoids and two with dermoid tumors. Two additional cases of epidermoid tumors of the brain were reported by Rosenbluth and Lichtenstein 25 in 1960. Unless calcification is present, the intracranial mixed tumors are seldom suspected preoperatively.

Generally speaking, the embryonic rest tumors of late inclusion are more benign and offer a more favorable prognosis than those derived from more than one germ layer. Some may be easily removed, others cured after a series of operations; some remain quiescent even though incompletely removed, while others are so extensive or invasive that radical surgery is not possible. We are reporting three additional cases: one intracranial and two intraspinal.

Case Reports

Case 1. P.P.P., an Army Air Force sergeant, noted left-sided tinnitus in March, 1943. This was followed by progressive hearing loss, diplopia, left-sided ataxia, and weakness of the left side of the face. There was

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also an associated left fronto-occipital headache. The pertinent features of the examination included left-sided deafness, a peripheral left-facial weakness, hypesthesia of the left trigeminal nerve, papilledema, and both horizontal and vertical nystagmus. The vestibular examination disclosed an absence of response on the left with a diminished reaction of the vertical canals on the right. Audiometric tests confirmed the left-sided deafness. It was believed he had a left-sided acoustic neurinoma.

Operation. On August 3, 1945, at the Hammond General Hospital, Modesto, California, through a unilateral approach, a large bluish pontine cyst was uncovered in the cerebellar pontine angle. It contained thick, grayish-red mucoid material which coagulated rapidly; 40 cc were collected. The capsule extended anteriorly as far as the fifth cranial nerve, which was clearly seen. While the seventh nerve was intact, it appeared compressed. All of the cystic material was removed by suction, but the thin texture of the capsule which incorporated the seventh and eighth cranial nerves, prevented its complete excision. However, at the conclusion of the procedure it was believed that only a small portion of the lining membrane had been left behind.

Microscopic Findings. The microscopic diagnosis of the material removed, made by E. L. Benjamin, M.D., was that of an epidermoid cyst. Under low-power examination he observed the delicate linear structure of a thin-walled cyst. Higher magnification demonstrated an internal lining one to two cells thick; the cells were somewhat flattened. The basophilic nuclei were round to oval, and their long axes appeared to be parallel with the lining of the cyst. Externally, a rather delicate arrangement of collagen connective-tissue fibers lay parallel to the cyst membrane.

Follow-Up. When examined again in 1948, the patient continued to have occasional left-sided headaches but there was no papilledema and no bulging of the suboccipital craniotomy. He believed his hearing was improving. In 1952, 7 years postoperatively, the patient reported his hearing had improved and that he was essentially unchanged. In 1966, Edward S. Connolly, M.D., of San Francisco, re-explored and evacuated the cyst. The patient tolerated the procedure well and experienced a benign postoperative course.

Case 2. R.L.B., a 41-year-old telephone engineer, had been afflicted with urinary difficulties including cystitis since childhood. He first recalled intermittent low-back discomfort in 1933. As the result of a transurethral resection in March, 1947, his urinary symptoms were slightly improved. During this operation four attempts at spinal anesthesia were unsuccessful. Numbness of the buttocks and rectum, noted about 4 days postoperatively, subsided spontaneously. For 2 months before our examination, he had continuous low-back pain and bilateral sciatica that incapacitated him for any steady work.

Examination. The salient features in the examination at the Santa Barbara Cottage Hospital included a sacral birthmark, a lower sacral pilonidal cyst, and atrophy of both calves. Both ankle jerks were absent; there was weakness of extension of the feet as well as impaired straight-leg raising. Except for a faint trace of protein in the urine, the laboratory findings were normal. Roentgenograms of the lumbosacral spine disclosed spina bifida in the first sacral segment and diffuse enlargement of the lumbar canal.

A myelogram showed a block at L-2. No spinal fluid was obtained at low lumbar levels. Because of the long history and the presence of both spina bifida occulta and a pilonidal sinus, it was believed the large obstructive mass in the region of the cauda equina was an epidermoid or dermoid tumor.

Operation. A large sausage-shaped, pearly tumor incorporating the nerve roots of the cauda equina was removed at lumbar laminectomy October 5, 1948. It had enlarged the lumbar canal and had the gross appearance of an epidermoid tumor. After careful dissection, very little abnormal tissue remained.

Microscopic Findings. The pathological findings were described by William O. Russell, M.D. Microscopically, there was a broad zone of parakeratosis and hyperkeratosis in the stratified squamous epithelium. In one part a defect was seen in the stratified squamous epithelium. Here there were many multi-nucleated giant cells and large masses of macrophages. The diagnosis was epidermoid tumor.

Postoperative Course. The postoperative course was uneventful, and in view of the
involvement of the cauda equina, the patient revealed little weakness. His progress has been satisfactory. He has experienced no significant back or leg pain, and showed no objective sensory loss. The calf muscles remained atrophic and the Achilles reflexes absent. In March, 1952, with the exception of continued inability to raise himself on his toes, he demonstrated good power in the legs. In March, 1966, 18 years postoperatively, he was working regularly and had had no return or progression of symptoms.

Case 3. B.L.H., an 8-year-old girl, was admitted to the St. Francis Hospital, Santa Barbara, on August 11, 1952, with a complaint of increasing pain and weakness of the legs of 7 months' duration. Examination revealed a birthmark in the sacral area. Both legs were weak and the patellar and Achilles reflexes could not be obtained. X-rays demonstrated minor congenital anomalies of the first and second sacral neural arches. A myelogram showed complete obstruction at the second lumbar level.

Operation. A lumbar laminectomy August 19, 1952, led to the removal of an intraspinal epidermoid of the cauda equina, extending from the first to the fourth lumbar region. The tumor was attached to one of the nerve roots, which was sacrificed. William J. Sheehan, M.D., made the pathological diagnosis of an epidermoid of the cauda equina. The sections showed stratified squamous epithelium arranged in a corrugated pattern beneath which there was dense, acellular, connective tissue containing evidence of acute and chronic inflammation. An amorphous, laminated, and faintly basic-staining substance had sloughed off from the superficial layers of the squamous cells.

Postoperative Course. The postoperative course was uneventful. The patient was gratifyingly relieved of her pain. When re-examined September 20, she had been going to school and was comfortable. The patellar and Achilles reflexes were still absent, but no motor or sensory weakness was demonstrable. She has remained relatively symptom-free during the succeeding 12 postoperative years.

Clinical Features

In 20 cases of intraspinal dermoid and epidermoid tumors collected from the literature, Gross\textsuperscript{10} found the average duration of symptoms to be 10 years, and the average age at the time of operation to be between 30 and 40 years. Such a delayed appearance of neurological signs and symptoms is best explained by the slow growth and soft content of these tumors. Often the x-rays show a diffuse fusiform enlargement of the canal extending over several vertebral levels.\textsuperscript{7-11}

The combination of a long duration of symptoms, radiographic evidence of pressure erosion, and the presence of a spine bifida occulta or a pilonidal sinus are characteristic of the intraspinal tumors. These findings led to the correct preoperative diagnosis in our Cases 2 and 3.

Summary

We have reported three cases of mixed (epidermoid) tumors of the central nervous system, all treated successfully. We have reviewed other significant reports and theories on this subject.

The combination of a long duration of symptoms (pain and weakness) with spine bifida occulta and x-ray evidence of diffuse enlargement of the spinal canal can often lead to a preoperative diagnosis.

References