CLINICAL OBSERVATIONS ON TUMORS OF THE PINEAL REGION

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SINCE Blaue's original report, tumors of the pineal region have presented a fascinating challenge in diagnosis, pathology, and therapy. In most areas, as Cushing stated, "... they are so few in number and so variable in their histological composition that their life history and the possible relation of any one type to the condition known as 'pubertas praecox' is imperfectly understood." However, as previously reported, these tumors occur relatively frequently in Japan. The present report concerns the clinical features, therapy and results in 24 such cases. The pathological classification was: pinealoma, 15; "double" tumor, 5; teratoma, 2; astrocytoma, 1; and meningioma, 1.

As often noted, the greatest incidence is in males under the age of 25 (Fig. 1). In this series, 21 of the patients were males and were under 25 years of age. The oldest patient, a woman with meningioma, was aged 31 years. The initial symptom was headache in 21 cases, usually associated with nausea and vomiting. Other complaints were: diplopia, 22; visual disturbances, 15; tinnitus, 9; dysacusis, 8; and vertigo, 8. The duration of symptoms was less than 6 months in 14 cases and was over a year in only 2 instances, the longest being 26 months.

Examination revealed papilledema in 21 cases, with secondary optic atrophy in an additional case. Limitation of upward gaze (Parinaud's syndrome) was noted in 16 patients, half of these having absent pupillary reactions. Some form of disturbance of the 3rd cranial nerve was found in 18 patients, in all but 2 being bilateral. By contrast, the trochlear nerve was involved in only 4 instances, and the abducens in 2. The 7th and 8th cranial nerves were affected in 4 and 7 cases respectively, and the 5th in 5. Nystagmus of various types was present in 9 instances. Ataxia was evident in 16 cases, of cerebellar type in 6. In 7 instances there were sensory disturbances, usually lateralized. Hemiplegia occurred only twice but abnormal Babinski's responses appeared in 5 cases and hyperreflexia with clonus in 8.

Disturbances in the psychic and mental spheres were not uncommon. Clouding of consciousness occurred in 7 cases, usually episodic in relation to severe headaches. Evidences of excitability and impatience were found in 10 patients, while 4 became dull and apathetic. Metabolic abnormalities were of various types, both obesity and emaciation being observed; the latter was especially striking in 8 instances. Polyuria and frequency were not unusual. Pubertas praecox was present in 2 patients while in a third similar changes developed following removal of an astrocytoma in the pineal region. Vascular hypotension below 100 mm. Hg systolic was noted in 3 instances.
TREATMENT

The first documented surgical approach to a pineal tumor was made in 1913 by Krause through the subtentorial space without success. During the past 40 years the usual procedure has been either that of Dandy or through the splenium of the corpus callosum, or the transventricular approach of Van Wagenen. Horrax, in 1937, reported a successful removal of a 70-gram pinealoma by a modification of the latter method. More recently the trend has been toward radiation therapy after some form of ventricular shunt, because of the high mortality associated with attempts at surgical excision. Assessing the operative hazards, it is apparent that they fall into three categories: preparation, the operation itself, and post-operative management. The problems, and our methods toward their solution, will be considered under the above headings.

Preparation is directed primarily toward early reduction of increased intracranial pressure, which was present in 22 of the 24 cases. This is accomplished by controlled ventricular drainage over a period of 4 or 5 days, with appropriate antibiotic protection. Since tissue swelling may be present in the tumor-bearing area, cortisone is given daily in amounts of 100–150 mg. Many of the

Electroencephalography was done in 15 instances with only 2 normal results. The others showed frequent slow waves, usually of high voltage and generalized, except in 4 patients who presented lateralizing neurological signs.

Roentgenography disclosed calcification in the pineal region in 15 instances, including a teratoma containing teeth and cartilage (Fig. 2). Spreading of sutures and increased convolutional markings were present in 4 patients under 10 years of age. Ventriculography revealed symmetrical hydrocephalus and a defect in the pineal region of the 3rd ventricle in all but 1 case (Fig. 3). Slight displacement or irregular enlargement of the adjacent lateral ventricles were noted in some instances of infiltrative tumors. Angiography supplied valuable information concerning collateral venous channels for the veins of Galen in some cases.
patients are severely dehydrated before admission; rehydration is begun as soon as ventricular drainage is established, but not before this time. An adequate caloric intake is also maintained during this period.

Operation is planned to minimize hemorrhage and disturbances of the autonomic nervous system. Both objectives are promoted by the use of hypothermia to about 27°C, induced under ether anesthesia. Autonomic blocking agents such as chlorpromazine and promesadine are given at this time. The considerable depth of the surgical target and its adjacent vascular structures is a clear indication for an ample craniotomy exposure, usually in the occipitoparietal region, according to the method of Dandy. Cortical veins entering the sagittal sinus posterior to the rolandic vein are occluded and divided and the cerebral hemisphere is retracted gently from the falx. The ventricular drainage tube permits displacement of ventricular fluid during this maneuver, providing ample exposure of the corpus callosum. This structure then is divided, giving access to the portion of tumor within the posterior part of the 3rd ventricle. Absolute hemostasis should be obtained before removal of the tumor is begun. For large tumors occupying the upper quadrigeminal region, the falx and tentorium are divided to provide further exposure. The deep venous drainage system is now assessed, with special reference to the great and lesser veins of Galen and their relations to the tumor. Proper handling of these structures is the technical "heart" of the operation and the unsatisfactory results in this series were related chiefly to venous injury and hemorrhage in this region. Unilateral occlusion and division of the lesser veins of Galen are indicated for safe access to some tumors and occasionally it may be permissible to ligate the great vein if collateral anastomoses have already been established. Important information on this point may be obtained from preliminary angiography. The consistency of the tumor is often the crucial factor in determining the technical indications at this stage. Soft tumors can be removed by suction with minimal injury to the veins, which may traverse the tumor. The firm tumors tend to form deep furrows, in which the veins are lodged, requiring ligation before the tumor can be removed. The 3rd ventricular portion of the tumor is then lifted carefully with a small spatula, identifying and securing any deep vascular connections. If the tumor is firm and very large, it may be necessary to use digital palpation to identify the cord-like vessels along its hidden surface. All save 2 of the tumors were encapsulated.

The operation was done by the method of Dandy in 12 cases, the transventricular approach of Van Wagenen in 6, and a combination of the two procedures was used in 1 instance. The Torkildsen shunt operation was done in 2 cases. The remaining patients died before a surgical attack upon the tumor was accomplished. The tumor was thought to have been removed completely in 16 patients while only partial excision was possible in 3. Injury to the veins of Galen occurred in 11 cases, with 9 deaths, compared to 3 deaths in the 8 cases in which such injury was avoided.

Postoperative management plays a major role in the success of all intracranial operations, but especially of pineal surgery. Controlled ventricular drainage is continued until the pressure returns to a satisfactory level. The ventricular tube is less likely to become clogged if it is placed in the frontal horn; it should be anchored firmly. Excessive loss of ventricular fluid may lead to unconsciousness and seizures, so that abnormally low pressure should be avoided. If ventricular drainage must be continued more than 20 days, another site is selected for the tube, to avoid infection. Persistent high ventricular pressure is an indication for some type of fluid-shunt procedure. Hyperthermia of 39°C or above occurred in all but 1 patient. Among the 11 patients whose temperature did not exceed 39°C, there were 6 survivors. Of the 8 patients with more severe hyperthermia, there was only 1 who survived. The combination of high fever and prolonged unconsciousness usually was an indication of blood in the ventricular system, with a poor prognosis. Measures used to combat hyperthermia included antibiotics, autonomic
blocking agents, and external cooling with multiple ice bags. Oral feedings are instituted as early as possible.\textsuperscript{12,13}

RESULTS

Prior to 1955, when ventricular drainage, hypothermia, and the use of cortisone and autonomic blocking agents were introduced, there were 8 patients. Of these, 6 had surgical attack upon the tumor, 1 ventriculography only, and 1 had a ventriculocisternal shunt. All of these patients died.

In the more recent series there were 16 patients, 13 of whom had surgical excision of the tumor; 2 died following ventriculography. In 1 of these an inoperable situation\textsuperscript{20,21} was found at autopsy, with neoplastic invasion of the lateral and 3rd ventricles (Fig. 4). The other patient, with a 32-gram tumor, became unconscious after excessive reduction of ventricular pressure and expired. The remaining patient was thought to have a cerebellar tumor, the small pineal neoplasm being undetected by ventriculography. Extirpation of the tumor was successful in 7 instances. Five of these patients are living, 2 to 18 months after operation. One died after reoperation for a recurrent glioblastoma at 7 months; the original diagnosis was astrocytoma. The other patient\textsuperscript{19} died 20 months after operation, with recurrent symptoms. In the 6 instances of operative fatality the critical factor appeared to be injury and consequent ligation of the veins of Galen in 5. One of these patients had a 102-gram teratoma (Fig. 5), and another a 35-gram heavily calcified meningioma (Fig. 6). The remaining patient died from infection, associated with a cerebrospinal-fluid fistula.

SUMMARY

The clinical features, therapy and its results, in 24 patients with tumors in the
pineal region are described. The tumor types were: pinealoma, 15; "double" tumor 5; teratoma, 2; astrocytoma, 1; and meningioma, 1. Surgical extirpation was done in 19 cases with satisfactory results in 7. Special emphasis is placed upon pre- and postoperative management.

REFERENCES


Fig. 6. Roentgenogram of skull showing densely calcified 35-gm. meningioma.