CHOLESTEATOMA OF THE CENTRAL NERVOUS SYSTEM
FOUR CASES OF EPIDERMOID TUMORS

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Intraspinal epidermoid tumors comprise only about one quarter of the epidermoids of the central nervous system. [1,2] In 100 cases of central nervous system tumors operated on and verified by biopsy, we isolated only four cholesteatoma cases. Of these, three were intracranial and one intraspinal. We thought it worthwhile to publish an account of these seldom seen tumors with a brief description of their peculiar clinical properties.

Case Histories

Case 1: H. M., age 41, was admitted to the hospital with painful tics in the frontal region and clonic contractions of the left side of the face. Five years before, he had had a discharge from his ear, which was treated with antibiotics. Four years later clonic contractions began on the left cheek, with a vertigo which forced him to lie down.

Examination: He was fully conscious and cooperative. His pupils were equal and reacted normally to light and distance. Nystagmus appeared especially on the right horizontal gaze. Left corneal reflex was negative. Hypalgesia on the left maxillary and mandibular division, left central facial palsy, diminution of hearing of the left ear, lateralization of Weber sign toward the right, and diminution of air and bone conduction were noted. Rebound phenomen were observed on the left upper extremity, as well as hypotony, asymmetry, asynergy and quick fatigue of the left arm. Hypotony of the left leg and awkwardness on the same side in the wheel knee test were noted. Bilaterally, patella and Achilles reflexes were positive and there was a bilateral extension response of the plantars. The Rombergs sign was positive with a lateral pulsion towards the left, and typical cerebellar gait.

Instead of an uneventful postoperative course, the patient died of a purulent meningitis, perhaps due to a careless lumbar puncture.

Case 2: H. Y., a 35 year old man, was admitted with symptoms of continuous dizziness, staggering gait, and blackout spells. One day, three years before, his legs had felt numb and he had been unable to walk. Later he showed signs of giddiness, nausea, and on

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several occasions vomiting. Shortly after, a tinnitus of the right ear developed, and despite improvement in this symptom, the ear became gradually deaf.

Examination: The patient was lucid and cooperative. On percussion, the left parietotemporal region was sensitive, even painful. Visual acuity was bilaterally 6/10. The bilateral secondary optic showed atrophy in the initial stage. The bilateral course showed horizontal nystagmus. Corneal reflexes were present. Right facial hypalgesia was present with continuous clonic contractions at the left commissure. There was hypotony, asymmetry, and asynergy of the right arm. Diminished tonus of the right lower extremity was noted. The wheel knee test showed awkwardness on the right. Tendon reflexes were normal bilaterally.

The case was diagnosed as a tumor of the right cerebellar lobe, eventually a right cerebellopontile tumor, and an exploration of the posterior fossa was made. When the dura mater was reached by a suboccipital craniotomy, its strong pulsation surprised us. After opening the membrane, the folia cerebelli did not look enlarged but had a hard consistency. While evacuating the cisterna magna, deflection of the tonsillea exposed a mass with a waxy consistency, and a pearly appearance, which proved to be a cholesteatoma. It was noted that this tumor had filled the fourth ventricle completely, enormously extending its cavity. The mass weighed 165 grams. After an uneventful postoperative course, the patient was discharged lacking right cerebellar coordination. This later improved greatly (see Figure 1).

Figure 1. Cholesteatoma extirpated from the fourth ventricle, weighing 165 grams.

Case 3: A. E., a 35 year old man, entered the hospital complaining of painful tics on the left side of his face at half hour intervals. Five months before he had had a series of very painful tics in the maxillary region which were considered due to decayed teeth. However treatment did not result in alleviation of the pain. A neurologist, thinking that his case was trigeminal neuralgia, referred him to us.

Examination: The patient was lucid and cooperative, sight 8/10 bilaterally, proptosis was present, pupils were equal, and reaction to light normal. The left half of the face was edematous and asymmetrical, no other abnormalities of the other cranial nerves were found. Extremities were normal.

In this case we decided to perform a Dandy operation as the origin of the typical neuralgia attacks was thought to be due to a left cerebellopontile angle lesion. After the usual left suboccipital craniotomy, left ventricular puncture and evacuation of the cisterna magna, the left cerebellum was easily retracted and a mass with pearl-like bodies on it, growing under the cerebellar surface, was seen and identified as a cholesteatoma. After opening its capsula the contents were evacuated easily with a spoon. Because of profuse bleeding from the capsula, no attempt was made to extirpate it. After an uneventful postoperative course, the patient was discharged without any complaints. Histological diagnosis was epidermoid tumor.

Case 4: A. B., a 9 year old child, was admitted to the hospital with complaints of difficulty in walking and low backache. The backache started eight months before admission and a few days later radiated toward her right leg. Increasing pain obliged her parents to consult a doctor who prescribed drugs with no apparent benefit. Later treatment in an orthopedic clinic relieved her pain to a degree but was abandoned upon return of the pain. Upon consulting a neurologist, she was advised to see a neurosurgeon and was admitted to our clinic. This patient was seen by the eminent English neurologist, Dr. Hugh Garland.

Examination: On the lumbar region there were annulary angiomatous spots with mild hypertrichosis. The region was sensitive to percussion. As an antalgic, the patient's position tended toward the right with the lumbar muscles strongly contracted. Abdominals were positive. Active and passive movements were normal with a good leg stretching test, knee jerks were positive bilaterally. Ankle jerks were diminished on both sides and the plantars were extended. There was bilateral impairment of both superficial and deep sensations. The child's gait was painful and staggering. Routine examinations were normal.

X – ray examination showed a spina bifida between the eleventh dorsal and fourth lumbar vertebrae and enlargement of the vertebral canal with extreme thinning of the pedicles of the first lumbar to third lumbar vertebrae (Figure 2). A myelography (using 4 cc of "myodil" injected sub-occipitally) showed that the opaque material, producing a cupula image and lateral infiltrations at the first lumbar vertebra, totally filled the remaining vertebral canal down to the "cul-de-sac" (Figure 3).

All of these observations were supposed due to a congenital medullary tumor and we decided to operate on the child.

A laminectomy between the second lumbar and the fifth lumbar vertebrae was performed and after opening the dura, an intramedullary mass of pearl-like particles, measuring 5x2x1 cm was seen (Figure 4). The capsula was totally removed. After incision and evacuation, the histological diagnosis was epidermoid tumor (Figure 5). The patient was discharged after an uneventful postoperative period. Control examinations after eight months and again after two years showed no weakness of her legs, no difficulty walking and the sphincters were normal.

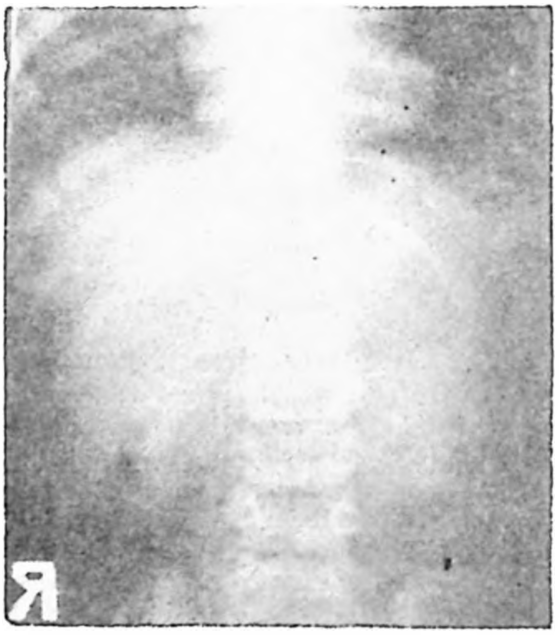

Figure 2. Anteroposterior direct x-ray examination of the vertebral column in case 4.

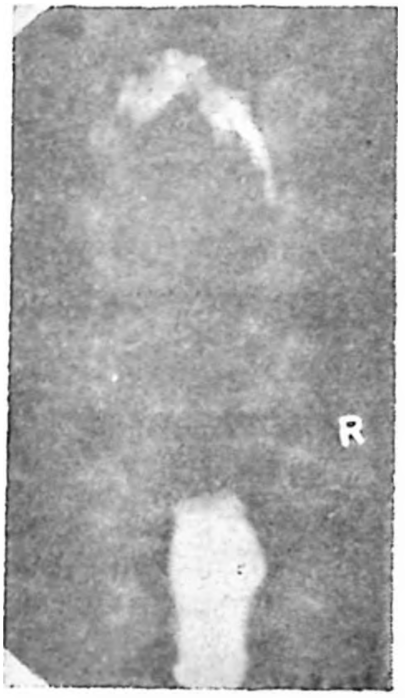

Figure 3. Typical cupola image and filling defect in a case of intramedullary cholesteatoma.

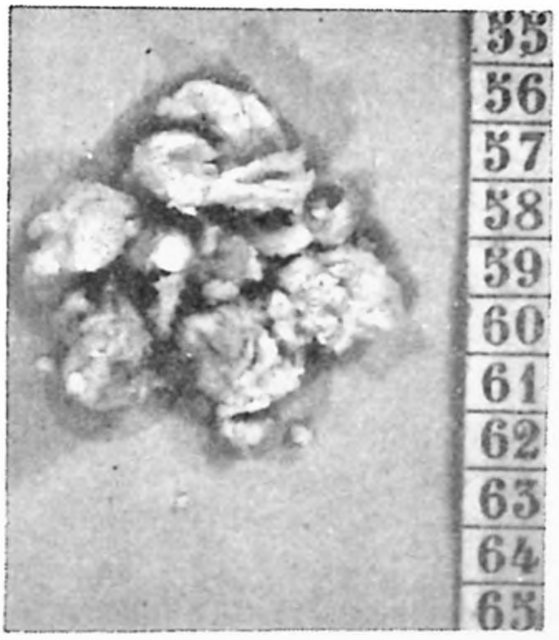

Figure 4. Macroscopic appearance of the intramedullary cholesteatoma.

Figure 5. Microscopic appearance of the same tumor.

Discussion

Epidermoid and dermoid tumors are formed from skin elements. These tumors should not be confused with the cholesteatomas of the middle ear which take their origin from inflammatory desquamation, or with intraspinal epidermoids which arise after repeated lumbar puncture for the treatment of tuberculous meningitis.

In some cases of congenital epidermoid tumors other developmental anomalies may also be found on the midline. In the four patients whose cases were published by French [3] one had hare-lip; one facial asymmetry; and the other two, dermoid cysts of the nasal tip. Defects most frequently observed are dermal sinus, hypertrichosis at the site of the tumor, skin defects, angiomatous spots, and spina bifida. In the case which we presented as intramedullary epidermoid tumor, all of these congenital defects except dermal sinus were present.

Intraspinal epidermoid tumors are seen usually earlier in life than intracranial ones, the most frequent age span being between 15 and 25. According to Jefferson and Smalley [4] 17 per cent of all cranial epidermoid tumors are located in the diploe. Intradiploic tumors can be easily diagnosed in direct radiological examination by their typical image. They cause very sharply delineated bone defects.

Intracranial epidermoid tumors are most frequently located at the pontile angle, the base of the brain, the suprasellar region, and interventricularly.

Preoperative diagnosis of intradural epidermoid tumor is not possible in a majority of cases. Direct x-ray examination usually reveals only that there may be thinning of the anterior clinoids in suprasellar epidermoid tumors. Even the absorption of the chiasmatic sulcus and widening of the optic foramina may be diagnosed epidermoid. When tumors are located at the angle between the pons and the cerebellum they may cause an erosion of the petrous pyramid. On encephalography, the spongy appearance of these lesions is characteristic. [5-7] Weinberger and Dyke [5] consider this caused by air invasion of the spaces in the wall of the tumor. Lindgren [6] and Hauser [8] reported that the only space occupying lesion which gives a characteristic image is a cholesteatoma.

Intraspinal epidermoid tumors are usually a single lesion, but multiple cases have also been reported. [9,1] These multiple cases are either primary or the result of implantation in the subarachnoidal space due to rupture of the primary lesion.

Surgery is the treatment for these tumors. The tumor with its capsula must be totally removed to prevent recurrence. In many cases, evacuation of the contents of the tumor and partial excision of the capsula is satisfactory because these tumors grow very slowly and the necessity for reexploration is very rare. In two of our cases our method of treatment was partial excision and up to this time there have been no signs of recurrence. In the case of the intramedullary epidermoid tumor, the tumor was extirpated totally with its capsula.

Total extirpation is usually feasible in cases of intradiploic tumors, though in these, the affected bone must be totally resected. [10]

Meticulous care must be taken to avoid any implantation of small tumoral pieces into the ventricles or the subarachnoidal space because these pieces can cause an aseptic meningitis. [11, 5] This lesion is the result of chemical effects of cholesterin. Sometimes a small tumoral fragment can enter the ventricular cavity and obstruct the "aqua ductus Sylvi".

In the first case we presented a previously existing aseptic meningitis, which was probably converted into a septic lesion as a result of the lumbar puncture.

Summary

In this paper we presented four cases of cholesteatoma of the central nervous system, one of which was intramedullary and three of which were were intracranial. We discussed the clinical picture and the results of surgical treatment in our cases.

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