Osteochondroma of the Base of the Skull Causing an Isolated Oculomotor Nerve Paralysis

Case Report Emphasizing Microsurgical Techniques

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Intracranial chondroma and osteochondroma are rare tumors. Chorobski, et al., in 1939 collected 25 intracranial chondromas from the literature and his personal series. Aronson and Otis in 1962 found 25 additional cases and reported one case of their own. Gabrielsen and Kingman in 1964 reported one case. Falconer, et al., in 1968 added two cases. Our case is unique because a review of the literature has disclosed no previous report of a similar lesion causing an isolated oculomotor nerve paralysis.1,5,6,10,14,16,17,20,21

Case Report

A 25-year-old woman was admitted to Harbor General Hospital on June 25, 1967, with the chief complaint of a droop of her right eyelid and double vision. In January, 1966, she began to have diplopia on occasions when she suddenly looked to the left. This cleared spontaneously and had not been a problem until December, 1966, when she noticed drooping of her right eyelid; within a few months this had progressed to virtual inability to open the right eyelid and persistent double vision.

The patient was known to have Ollier's disease (enchondromatosis) which in her childhood had required multiple surgical procedures, on both knees and the left ankle to correct the deformities.

Examination. The abnormalities in the general physical examination were limited to the slightly deformed knees. Neurological examination revealed an isolated paralysis of the right oculomotor nerve with a complete ptosis and a dilated, nonreactive pupil (Fig. 1, left). Visual acuity and visual fields were normal. The fundi were normal. There were no other significant neurological findings.

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Blood sugar was 95 mg%; tests for venereal disease were negative. The cerebrospinal fluid was normal. Tensilon test was negative, and the electroencephalogram was normal.

Radiological Studies. Skull films showed an irregular calcification in the area adjacent to and slightly posterior to the right posterior clinoid process (Fig. 2). No bony changes in the skull were identified. Chest x-ray showed calcified and cystic changes of the right fourth rib and the left seventh and eighth ribs and the right scapula. X-ray of the pelvis and lower extremities showed multiple changes of the bones, demonstrating expansion of the cortex and some loss of trabecular pattern. Brain scan with Technetium99m was normal. Bilateral carotid and vertebral angiography and pneumoencephalography did not show significant abnormalities.

Operation. On July 5, 1967, a right temporal craniotomy was performed under general anesthesia. The right temporal lobe was elevated slowly from the floor of the temporal fossa until the free margin of the tentorium was identified. The tumor was apparent in the region of the point where the free margin of the tentorium attached to the posterior aspect of the petrous bone. The binocular operating microscope was introduced using 10 power magnification. The arachnoid was freed from the dome of the tumor, which presented as a cystic-appearing mass. The right third cranial nerve could not be seen at this time. Dissection about the tumor was continued and a window was then cut in the wall of the tumor for internal decompression. The contents of the tumor consisted of a gelatinous material, which was easily removed with suction, and stony fragments of calcific material ranging from 2 to 5 mm in diameter. The tumor was moderately vascular and was evacuated with a
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Fig. 1. Left: Preoperative photograph of the patient showing a complete right third cranial nerve paralysis. Right: Postoperative photograph of the patient 6 months after removal of the osteochondroma.

A combination of curettage and suction, revealing a mass of approximately $2 \times 3 \times 2$ cm. The right third cranial nerve was identified under the surgical microscope on the medial side of the tumor, evidently having been displaced medially and thinned like a ribbon. It was possible to separate the tumor capsule from the third cranial nerve and to follow the nerve in its entirety from beneath the posterior clinoid process to the brain stem. The tumor actually arose from the tip of the right petrous bone and was completely removed with its capsule. The wound was closed in a routine fashion. The pathological diagnosis on microscopic examination was osteochondroma.

Postoperative Course. The patient had an uneventful postoperative course and was discharged from the hospital on July 13, 1967, the eighth postoperative day. Follow-up visits showed rapid recovery of the right oculomotor nerve function. Three weeks postoperatively, she was able to open her right upper eyelid slightly, and by 3 months there was no ptosis. Movements of the right eye returned slowly to normal, and the right pupil became smaller and reactive to light within 6 months after the operation (Fig. 1, right).

Discussion

The rarity of intracranial cartilaginous neoplasms is well known. Cushing found only three osteochondromas in his series of
2033 cases. He mentioned that these were at the base of the skull projecting into the cranial chamber, producing symptoms by compression, invasion, or other complicating factors. Leitholf\textsuperscript{14} found four chondromas among 4135 brain tumors seen at the Serafiner-Lasarettet in Stockholm. At the Tonnis Clinic in Cologne, there were nine cases considered as chondromas of the basilar synchondroses among more than 6000 brain tumors.\textsuperscript{2,12,13,18} Occasionally, intracranial chondromas, as in our case, may be associated with similar lesions in the skeleton, especially long bones, and paranasal sinuses (List\textsuperscript{17} Cases 5, 6, and 7). These tumors appear to have four sources of origin within the cranial cavity,\textsuperscript{7} namely, from 1) the sphenoid, ethmoid, and occipital bones at the base of the skull; 2) the choroid plexus; 3) the dura mater, or more specifically from one of its three septum-like curtains, the falx cerebri, the falx cerebelli, and the tentorium; and 4) the arachnoid, especially in connection with meningiomas. List\textsuperscript{17} and others\textsuperscript{8,13} stated that the tumors arise from an embryonic rest retained along the basilar synchondroses.

The location of preference for these tumors appears to be either the parietal or frontoparietal area and the base or parasellar region.\textsuperscript{1,6} They may also involve the roof of the third ventricle, cerebellopontine angle, paranasal sinuses with intracranial extension and to the orbit.\textsuperscript{3,19,22} According to Kleinsasser,\textsuperscript{13} osseouschondromas and chondromas of the base of the skull occur in females more than in males and mostly from the second through the fifth decade of life. Clinically, the patient may have scanty symptoms, even when the lesion is large,\textsuperscript{4} possibly due to slow growth of the tumor. They may produce visual disturbance,\textsuperscript{20} ophthalmoplegia,\textsuperscript{16} or trigeminal pain.\textsuperscript{5}

Our patient had had symptoms referable to the tumor for about 16 months and developed a gradual isolated right third nerve paralysis without any other significant neurological findings. Green, \textit{et al.},\textsuperscript{10} reported that in only five (3.8\%) of 130 patients with isolated oculomotor nerve paralysis was the paralysis due to intracranial neoplasm (one hemangioblastic meningioma and four metastatic tumors). Skull x-ray is of little value in a non-calcified chondroma without bony changes. Klinger\textsuperscript{11} believes that chondromas produce a typical maculated shadow which derives from the calcifications and osteogenesis in tumors and often local erosion of the bone. In our case, the disease was suspected because of calcification of the region of the right posterior clinoid process, and the previously known enchondromatosis. Diagnostic studies were not helpful.

Pathologically, these tumors are benign. Malignant degeneration of the chondroma and osteochondroma, at the base of the skull, is rare.\textsuperscript{17}

Surgical removal of the tumor is the treatment of choice. Total excision of the tumor is not the rule, but even subtotal removal will give the patient many additional years of a useful life.\textsuperscript{6,17} In our case, the removal of the tumor was made possible by utilizing the binocular surgical microscope which was extremely helpful in minimizing the surgical trauma to the surrounding structures, particularly the right third cranial nerve.

**Summary**

We have reported a successfully treated case of osteochondroma of the base of the skull, originating in the right petrous bone and extending to the region of the right posterior clinoid process, causing an isolated right oculomotor nerve paralysis. We have emphasized the value of microsurgical techniques for a safe removal of the tumor.
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