Cranioopharyngioma of the cerebellopontine angle

Case report

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The authors report a case of craniopharyngioma presenting in the right cerebellopontine angle. The tumor was removed totally. The purpose of this report is to record this very unusual localization of a craniopharyngioma.

KEY WORDS • craniopharyngioma • cerebellopontine angle tumor

CRANIOPHARYNGIOMAS account for approximately 2% to 3% of all intracranial tumors. They are mainly considered to be suprasellar tumors. We are reporting a case of craniopharyngioma in a very unusual anatomic location, and consequently associated with very different clinical manifestations.

Case Report

This 14-year-old boy was hospitalized for evaluation of progressive loss of hearing in the right ear for 2 years. In addition, he had complaints of diplopia and pain in the neck radiating over the shoulders.

Examination. The physical examination was normal. Neurological examination confirmed bilateral marked papilledema and decreased pain perception over the distribution of all branches of the right trigeminal nerve. The right corneal reflex was nearly absent. A slight peripheral paresis of the right facial nerve was noted. Hearing was diminished on the right side. There was mild sway with the Romberg test. In light of these findings, a right cerebellopontine mass was suspected, and investigations were carried out accordingly.

Skull films and brain scanning were normal. Audiometric examination revealed normal findings in the left ear. Pure-tone audiometry of the right ear disclosed a sensorineural hearing loss, especially prominent in the higher frequencies (2000, 4000, and 6000 cycles/sec). Tone decay was markedly positive. Speech discrimination test was performed at 75 dB and was found to be 60%. Recruitment testing was negative. Bekésy audiometry disclosed a Type I pattern. Computerized tomography showed a right cerebellopontine mass (Fig. 1 left).

Operation. Craniectomy through a right parame
dian incision was undertaken with the patient in the sitting position. Extramedullary exploration revealed a cystic component of the tumor, and 10 to 15 cc of yellowish-brown oily fluid was aspirated. The tumor itself was round and encapsulated. It contained macrocalcifications. The fourth, fifth, seventh, eighth, and lower cranial nerves were identified and preserved. The tumor, approximately 5 × 3 × 3 cm in size, was totally removed by microsurgical technique (Fig. 1 right).

Postoperative Course. The patient had a third nerve palsy in the early postoperative period which did not improve, and he developed diabetes insipidus in the 2nd postoperative week. Aside from these complications, the patient was in a good condition and self-sufficient.

The microscopic examination (Fig. 2) yielded the surprising diagnosis of a craniopharyngioma. The specimen was reevaluated and the diagnosis was confirmed by four pathologists from different hospitals. Radiotherapy was decided upon after the histopathological diagnosis.

Discussion

The great majority of craniopharyngiomas (94%) are located in the suprasellar area. Like other tumors of this region, craniopharyngioma is usually manifest by increased intracranial pressure, visual disorders, endocrine disturbances, and mental impairment.
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FIG. 1. Left: Preoperative computerized tomography scan showing a solitary mass at the right cerebellopontine angle. The cystic component is seen lateral to the tumor. Right: Postoperative scan made 20 weeks after surgery.

Craniopharyngioma is considered to be a nonfunctional, noninvasive, and nonmetastasizing tumor; however, it may increase in size, thus producing the above-mentioned symptoms by compression and distortion of the adjacent structures. The first invasive craniopharyngioma reported in the literature was described in 1952 by Iyer. Postmortem studies revealed direct invasion of the anterior border of the pons. The tumor had replaced the structures in the hypothalamic region and had grown into the third ventricle and upper midbrain. A second case of craniopharyngioma invading the intra-axial brain tissues was that of a boy who had been treated both by surgery and irradiation. The patient died 12 years after the initial diagnosis, and necropsy showed massive invasion into the deep gray areas and almost total replacement of the midbrain and pontine structures. Only rarely has extension of the tumor been reported to be toward the temporal fossa, the posterior fossa, or the subfrontal and intracerebral tissue.

In the series of Petito, et al., tumor extension was into the anterior fossa in 5% of cases, into the middle fossa in 2%, and into the posterior fossa in 4%. Tumors were rarely situated entirely within the third ventricle, and originated in the sphenoid bone, in the nasopharynx and neighboring areas, or involved the midportion of the midbrain, thus causing Weber's syndrome. In addition, a few odontogenic craniopharyngiomas have been reported in the literature.

In Olivecrona's series of 108 craniopharyngiomas, lack of balance and Romberg's sign were observed in five patients, ataxia in two, nystagmus in seven, and nerve palsy in one. Love and Marshall have observed cerebellar ataxia in two patients and seventh nerve palsy in three in their series of 100 cases of craniopharyngioma. In spite of these posterior fossa symptoms, our review of the literature has failed to reveal a craniopharyngioma presenting as a primary cerebellopontine angle tumor. Our review of the literature dealing with large series of cerebellopontine angle pathology has shown that, aside from the common tumors of this localization (such as acoustic neurinomas, meningiomas, fifth nerve neurinomas, epidermoid tumors, metastases, chordomas, and brain-stem gliomas), rare tumors like lipoma, hamartoma, ceruminoma, and syphilitic gumma have been observed in the cerebellopontine angle. However, a craniopharyngioma has not been reported in this location.

The nature of the complications we faced suggests the extension of the tumor from the suprasellar region. We think that the tumor migrated from its original suprasellar location to this very unusual position through the tentorial incisura, and that a small portion of it remained in the suprasellar region at the time of diagnosis. During the manipulation and retraction of the tumor, this part of the tumor was disturbed, which resulted in the postoperative complications.

References


Manuscript received June 10, 1983. Accepted in final form October 10, 1983. Address reprint requests to: Nur Altinörs, M.D., Kızıllırmak Sokak 17/9, Bahaniklar, Ankara, Turkey.