Parasellar epidermoid tumor presenting as painful ophthalmoplegia

Case report

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In 1964, a 51-year-old man experienced a right abducens nerve palsy, which resolved spontaneously. In 1978, he developed painful ophthalmoplegia on the right with paresis of the right oculomotor and trigeminal nerves. Neuroradiographic evaluation was reported unremarkable, and the painful ophthalmoplegia was attributed to the Tolosa-Hunt syndrome. High-dose corticosteroids were administered with transient improvement of signs and symptoms. In 1979, cranial computerized tomography demonstrated a low-density lesion adjacent to the right cavernous sinus, which at operation proved to be an epidermoid tumor.

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PARAINFLUENT ophthalmoplegia with paresis of multiple oculomotor nerves, with or without involvement of the ipsilateral trigeminal nerve or sympathetic fibers, localizes the pathological process to the region of the cavernous sinus and/or superior orbital fissure. A wide variety of lesions may cause this clinical syndrome, including aneurysms,\textsuperscript{13,17,26} neoplasms,\textsuperscript{6,13,14,26,31} and inflammation.\textsuperscript{12,20,26,28} Idiopathic granulomatous inflammation in the cavernous sinus has been given the eponym Tolosa-Hunt syndrome.\textsuperscript{24} This syndrome is characterized by acute attacks of painful ophthalmoplegia, which may remit spontaneously and are extremely sensitive to corticosteroid therapy. The Tolosa-Hunt syndrome is a diagnosis by exclusion, and should be made only after an exhaustive search for other causes. The following report describes a rare intracranial tumor simulating the Tolosa-Hunt syndrome.

Case Report

This 51-year-old man experienced the abrupt onset of right-sided headache and horizontal diplopia in June, 1964. The headache resolved over 1 week, but the diplopia persisted. Five weeks later he was found to have a right abducens nerve palsy, but was otherwise neurologically intact. Plain skull films were within normal limits, and over a period of 6 months the diplopia resolved.

In November, 1978, the patient reported the onset of a “dull ache” in the right periorbital region. Three weeks later he noted horizontal diplopia and droopiness of the right upper eyelid. He was diagnosed as having sinusitis, and was begun on a tapering course of corticosteroid therapy. He reported marked improvement of the periorbital pain and diplopia; however, when the corticosteroids were discontinued, his symptoms recurred.

In December, 1978, the patient was hospitalized. He had partial right oculomotor nerve palsy with ptosis, ophthalmoparesis, and mydriasis. There was decreased sensation over the first two divisions of the right trigeminal nerve. The right trochlear and abducens nerves were intact, and the remainder of the neurological examination was unremarkable. Skull radiographs, cranial computerized tomography (CT), and four-vessel cerebral angiography were reported within normal limits. Cerebrospinal fluid examina-
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Fig. 1. Left: Axial computerized tomography demonstrating a right parasellar low-density lesion with remodeling of the sphenoid bone (arrow). Center: Magnified coronal view delineates a lobulated tumor with compression of the right cavernous sinus. Right: Scan obtained 1 year earlier reveals a low-density lesion (arrows) in the right parasellar region.

The patient showed limitation of elevation and adduction on the right side. Ptosis was 2 mm on the right side. Decreased sensation over the first division of the right trigeminal nerve was noted. The remainder of the general physical and neurological examinations were unremarkable.

Cranial CT revealed a low-density lesion in the right parasellar region, compressing the cavernous sinus (Fig. 1). On the basis of this finding, the neuroradiographic studies performed 1 year earlier were reviewed. The plain skull films were normal, but the CT scan disclosed a right parasellar low-density lesion (Fig. 1 right). Carotid angiography demonstrated medial displacement and opening of the right carotid siphon (Fig. 2).

Operation. A right frontotemporal craniotomy was carried out. The tip of the temporal lobe was resected.
Epidermoid tumor causing ophthalmoplegia

ed, and elevation of the frontal lobe allowed exposure lateral to the optic nerve and internal carotid artery. A large extradural mass protruded laterally from the cavernous sinus, with an intradural extension upward, that lay lateral and posterior to the carotid artery. The mass was incised and its “cheesy” contents evacuated. The dura was opened widely over the extradural mass, and a large amount of the same material was removed. The capsule of the intradural and extradural components was then stripped out, although some fragments remained adherent to the wall of the cavernous sinus. A segment of this adherent capsule was sent for histopathological study (Fig. 3).

Postoperative Course. After craniotomy, the patient had a complete right oculomotor nerve palsy. Six months later, there was no longer any right ptosis, and diplopia was noted only in the extremes of vertical gaze. Although reduced sensation over the first division of the right trigeminal nerve persists, there has been complete resolution of the right periorbital pain.

Discussion

A variety of neoplasms in the cavernous sinus region may cause painful ophthalmoplegia (Table 1). To our knowledge, this is the first report of an epidermoid tumor causing an isolated parasellar syndrome. Epidermoid tumors comprise less than 2% of all intracranial tumors. In reviewing 100 cases, Lepoir and Pertuiset classified epidermoid tumors into three main groups according to their site of origin, and correlated them with the main blood vessels at the base of the skull and choroid arteries, as follows: retrosellar or vertebrobasilar; suprasellar, parasellar, or carotid; and intraventricular or choroid. Epidermoid tumors in the suprasellar region involve the visual pathways (optic nerves, chiasm, and tracts). In reporting 41 intracranial epidermoid tumors, MacCarty, et al., found seven suprasellar cases. All patients had abnormal visual fields, but the authors specifically stated that “none of the patients had involvement of the extraocular nerves.” Parasellar epidermoid tumors arise in the middle fossa and usually extend laterally, producing a clinical syndrome of the temporal or Sylvian region. Such cases typically present with temporal lobe seizures, accompanied by hemiparesis and, more rarely, by speech disturbances. Occasionally, epidermoid tumors may originate in the paratrigeminal region. These tumors arise extradurally along the petrous ridge and grow into the middle fossa, under the Gasserian ganglion, initially affecting the tri-

![Fig. 3. Photomicrographs of the biopsy specimen. Left: An epidermoid cyst (arrows) is attached to the dural wall (D) of the lateral cavernous sinus. The large myelinated nerve (N) is probably the ophthalmic division of the trigeminal nerve. H & E, × 63. Right: The epidermoid cyst wall is composed of stratified squamous epithelium without appendages. H & E, × 125.]

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<th>Type</th>
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<td>primary intracranial</td>
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<td>chordoma</td>
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*Cases summarized from Jefferson, Rucker, Thomas and Yoss, and Weinberger, et al.*
geminal nerve, but ultimately the optic, oculomotor, trochlear, abducent, facial, and acoustic nerves may become involved. A review of published reports of paratrigeminal epidermoid cysts showed that cranial nerve palsies were progressive, and did not remit spontaneously. Radiographic examination in all cases revealed erosion and decalcification of the petrous bone. In contrast, the skull films of our patient were normal.

Our case is instructive for many reasons. The patient initially developed a right abducent nerve palsy, in all likelihood due to tumorous compression; yet this deficit cleared spontaneously. Fifteen years later, he presented with a right cavernous sinus syndrome. Appropriate neuroradiographic studies were obtained, but they were incorrectly interpreted. The patient was believed to have the Tolosa-Hunt syndrome, and was begun on corticosteroid therapy. The salutary response to treatment appeared to support this diagnosis. Indeed, remission of signs and symptoms after 48 hours of corticosteroids has been suggested as a therapeutic trial in confirming the Tolosa-Hunt syndrome.44 However, improvement on corticosteroid therapy has been reported in cases of paratrigeminal neoplasms, including chordoma, giant-cell tumor, and lymphoma.76 In addition, the fact that signs and symptoms continued to recur in our patient as the corticosteroids were tapered made the diagnosis of Tolosa-Hunt syndrome suspect. A second CT scan clearly delineated a low-density lesion compressing the right cavernous sinus. The usefulness of CT in evaluating both the parasellar region and intratumor, and lymphoma.

It is now generally agreed that epidermoid tumors arise from ectodermal rests.57 These result from incomplete cleavage of neural and cutaneous ectoderm, occurring between the 3rd and 5th weeks of embryogenesis. Recent biomathematical analysis suggests that epidermoid tumors grow linearly, at rates approximating those reported for normal human skin, rather than exponentially, as most tumors do.9 Such a growth rate would be expected from tumors derived from a single layer of basal germinal cells. This slow growth has led to the observation that intracranial epidermoid tumors may cause an unexplained isolated cranial nerve palsy for years before more dramatic symptoms appear.80 The pragmatic importance of epidermoid tumors is emphasized because of improved detection with CT and the possibility of effecting a clinical cure by their removal.8,9

References

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