Glomus jugulare tumor presenting with increased intracranial pressure

Case report

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The authors report a case of glomus jugulare tumor presenting with papilledema and visual loss. The tumor was extremely vascular with significant shunting of arterial blood into venous sinuses. There was no intracranial extension of tumor, and papilledema resolved after removal of the lesion.

KEY WORDS • glomus jugulare tumor • papilledema • pseudotumor cerebri • increased intracranial pressure

PAPILLEDEMA is an unusual finding in patients with glomus jugulare tumor, and has been reported only in association with posterior or middle cranial fossa invasion producing mass effect with or without obstructive hydrocephalus.\(^1,2,3\) The patient presented herein had a glomus jugulare tumor with no intracranial component, but developed papilledema which resolved after removal of the lesion.

Case Report

This 51-year-old obese woman was initially evaluated for right-sided deafness and tinnitus in August, 1976. For approximately 6 months before admission she had noted decreased hearing and a rushing sound synchronous with her pulse in the right ear. Past medical history was unremarkable with the exception of obesity and a diagnosis of hypothyroidism made in 1957, for which she was taking thyroid replacement.

First Admission. Physical examination revealed a loud bruit over the right temporal region, mastoid bone, and base of the skull, a mild right peripheral facial weakness, and sensori-neural deafness in the right ear. Fundoscopic examination was normal. Polytomography showed erosion of the right petrous bone with enlargement of the jugular foramen. Cerebral angiography demonstrated an enormously vascular lesion replacing the right petrous bone. The mass was fed by numerous branches of the external carotid artery including a huge occipital artery, muscular branches from the right vertebral artery, and basal branches of both internal carotid arteries (Fig. 1). Rapid shunting of arterial blood into the right transverse sinus and reflux of contrast material into the sagittal sinus were noted.

Based on the clinical and angiographic findings, a differential diagnosis of arteriovenous malformation (AVM) versus glomus jugulare tumor was made. There was no apparent intracranial involvement, and minimal neurological deficit. We were uncertain whether the lesion was neoplastic, and decided that direct attack would be a formidable procedure with significant risk. We therefore elected to treat the patient conservatively with embolization of the right external carotid artery with Gelfoam. The embolization was accomplished without difficulty and it resulted in a considerably decreased flow through the lesion, as seen angiographically, and marked diminution of the bruit. After embolization the patient developed considerable pain in the mastoid region, which resolved in approximately 7 days. The facial nerve palsy was transiently worse and the hearing loss was unchanged.

The patient's condition remained stable until December, 1976, when she developed episodes of "blurring" and "darkness" of vision and at times total blindness in both eyes. These episodes increased in
severity, frequency, and duration, and she was re-evaluated in March, 1977.

Second Admission. Her examination was unremarkable except for a continuous machinery-like bruit over the right mastoid region extending throughout her head and down the right side of her neck. It was not quite as prominent as when she was seen initially, but definitely worse than immediately after the embolization. Neurological examination showed bilateral papilledema with venous engorgement, hemorrhages, and exudates. She had severely diminished visual acuity, with light perception only in the left eye, and markedly constricted fields and an enlarged blind spot in the right eye. She could count fingers with the right eye. The mild peripheral nerve weakness on the right side of the face was unchanged, and there was no hearing in the right ear. No other neurological abnormalities were noted, with the exception of mild nystagmus in all directions of gaze. Endocrine evaluation was performed, and no abnormalities were noted.

A computerized tomography scan showed small ventricles and no intracranial extension of the mass. Angiography was repeated, and again showed the extremely vascular lesion filling from both internal carotid arteries and the right vertebral artery, with reflux of arterial blood into the cerebral venous system.

A tentative diagnosis of pseudotumor cerebri was made. However, the possibility was entertained that the increased venous pressure resulting from the arteriovenous shunting through the lesion could account for the intracranial hypertension and papilledema. Cerebrospinal fluid (CSF) was normal for cells and protein. Intracranial pressure (ICP) was monitored through a lumbar subarachnoid catheter. The mean ICP was slightly elevated (20 to 25 torr) but the amplitude of cardiac and respiratory pulsations was markedly exaggerated with peaks up to 35 torr. Drainage of CSF and megadose dexamethasone therapy were instituted, but there was no improvement in vision after 5 days.

Operation. In April, 1977, the patient underwent surgery. Initially the right internal jugular vein and right external carotid artery were ligated in the neck. Posterior temporal and suboccipital craniectomy revealed a firm, encapsulated tumor, which eroded the right petrous bone and involved the middle ear with no intracranial extension. The tumor was extremely vascular, and alarming arterial hemorrhage occurred, but the mass was ultimately totally removed. The seventh nerve and the petrous portion of the carotid...
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artery were not seen. Histological examination revealed a glomus jugulare tumor of an angiomatous type.

Postoperative Course. The ICP and amplitude of the pulsatile waves returned to normal. The papilledema resolved although, disappointingly, the patient's vision remained unchanged. After surgery, she had a total peripheral facial nerve palsy on the right. Follow-up examination in July, 1978, showed that the vision in her left eye was unchanged, but her right eye had improved to the point at which she could read fine newspaper print with magnifying glasses. In addition, the facial nerve palsy had recovered nearly completely.

Discussion

Papilledema has been observed as a rare accompaniment of glomus jugulare tumor, occurring in between 0% and 4% of cases in published series. In all reported instances, there was intracranial extension of the tumor large enough either to elevate the ICP by producing mass effect or to obstruct CSF pathways producing hydrocephalus. In our case, there was no intracranial mass effect. We propose that the direct shunting of high-pressure arterial blood, which refluxed into the major venous drainage system, impaired venous drainage, thereby increasing cerebral blood volume and ICP. Furthermore, the elevated venous pressure probably also impaired the absorption of CSF.

Occasionally papilledema is seen in patients with AVM's. Most of these patients have hydrocephalus resulting from subarachnoid hemorrhage. However, it has been observed in patients with communicating hydrocephalus and in those without hydrocephalus. The mechanism of development of papilledema in the latter circumstances is probably the same as in our case. This is presumably due to the elevated venous pressure in the sinuses altering the absorption of CSF.

We believe that this is the first reported case of glomus jugulare tumor presenting as the pseudotumor cerebri syndrome. The fact that the papilledema and high-amplitude intracranial pulsations resolved after removal of the tumor strongly suggests that the neoplasm was the causative mechanism. The presence of papilledema in association with such a tumor does not necessarily indicate intracranial extension.

References

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