Cystic schwannoma of the trochlreal nerve mimicking a brain-stem tumor

Case report


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A case is reported of a rare cystic schwannoma of the fourth cranial nerve which was interpreted as a probable intrinsic brain-stem lesion. The clinical approach to brain-stem tumors in terms of empirical treatment or surgical biopsy is discussed.

KEY WORDS · brain neoplasm · cystic schwannoma · brain stem · trochlear nerve

Schwannomas of cranial nerves are common intracranial tumors, constituting up to 8% of intracranial neoplasms. The vast majority of schwannomas occur on the acoustic nerve, with less common involvement of the trigeminal nerve and other cranial nerves. They are rarely found on motor cranial nerves. Schwannomas of the fourth cranial nerve (trochlreal nerve) are extremely rare, only three previous cases having been reported. The present report documents the findings in a fourth case, which was also unusual in that the patient presented with a cystic lesion mimicking a brain-stem tumor.

Case Report

This 18-year-old female college student presented with a 6-month history of intermittent drooping of the left eyelid and occasional diplopia.

Examination. She had a mild left ptosis and a mild and variable pupillary inequality. There was no evidence of papilledema and extraocular movements were normal. There were no other neurological abnormalities and the general examination was unremarkable, with no stigmata of von Recklinghausen's disease. Investigations, including serum electrolyte and coagulation studies and visual and brain-stem evoked responses, were normal. Tests for myasthenia gravis were negative. A computerized tomography (CT) scan was reported as showing an intrinsic brain-stem glioma (Fig. 1 left).

A course of radiotherapy was proposed and was about to commence when the patient's parents sought a second opinion. In view of the atypical history and some uncertainty about the CT findings, further high-resolution CT scanning was carried out. This showed a prepontine tumor with both solid and cystic components (Fig. 1 right). Both portions enhanced strongly on contrast administration. The solid component closely approximated the posterior cavernous sinus. The attached 2.5-cm cystic portion lay in the prepontine region and indented the adjacent pons. There was some flattening of the fourth ventricle. It could not be definitely ascertained whether the tumor was intrinsic or extrinsic to the brain stem.

Based on the radiological appearance, the differential diagnosis included a large posterior communicating artery aneurysm, a meningioma of the cavernous sinus, and a brain-stem glioma with a necrotic center. Left vertebral and left internal and external carotid angiography was performed. This showed a mass lesion in the region of the pons and midbrain causing displacement and elevation of the left posterior cerebral artery, the superior cerebellar artery, and the posterior communicating artery. No neovascularity was associated with the mass. The neoplasm was thought to have both infratentorial and supratentorial components. The angiographic and CT findings combined to favor an extrinsic lesion; however, an intrinsic cystic lesion was still considered a possibility. A magnetic resonance image was obtained to resolve the problem. This showed a clearly extrinsic...
Cystic schwannoma of the trochlear nerve

FIG. 1. **Left:** This initial computerized tomography (CT) scan was reported as showing an intrinsic brain-stem neoplasm (short arrow). There was some distortion of the fourth ventricle (long arrow). **Right:** Repeat CT scan showing a partially cystic, partially solid mass in close association with the brain stem (arrowhead). This scan was obtained on a GE 9800 scanner after administration of a double dose of intravenous contrast material.

cystic mass which was impinging upon the adjacent brain-stem structures (Fig. 2).

Operation. A left subtemporal craniectomy was performed. On retraction of the temporal lobe and free edge of the tentorium, a well-demarcated preoptic tumor was exposed. The left third cranial nerve was elevated and compressed by a cystic mass. The tumor arose from the fourth cranial nerve and entered the tentorium along with the nerve. The neoplasm was dissected free of the brain stem and was removed along with a portion of the trochlear nerve. The tentorial portion was removed piecemeal. Postoperatively, the patient recovered well and was discharged with a partial left third-nerve palsy which had improved over the short postoperative course. She did not complain of double vision.

Pathological Examination. The fragments of tumor had a firm rubbery consistency. Some had a smooth, slightly curved appearance consistent with the wall of a cyst. Microscopically, the fragments consisted of predominantly Antoni type A tissue (Fig. 3) with spindle cells arranged in bundles and occasionally in structures resembling Verocay bodies. Immunostains for S-100 protein were positive. The cyst showed no evidence of epithelial lining, but was defined by condensed tumor stroma. This histology was diagnostic of a schwannoma.

Discussion

Brain-stem lesions, and those lesions thought to arise from the brain stem, often cause a clinical dilemma. The problem is a significant one, particularly in children. Tumors of the central nervous system still constitute the largest group of solid neoplasms seen in childhood. 10% to 20% of these are brain-stem tumors. In adults, tumors of the pons and midbrain account for 1% of all intracranial tumors, although the...
percentage of all brain-stem tumors occurring in adults in some series is as high as 36%.21

Historically, surgical biopsy of these lesions has resulted in high morbidity.20 This fact, combined with increasing confidence in radiographic techniques for diagnosis, has led many clinicians to suggest radiotherapy or chemotherapy as a primary method of treatment without the necessity of a preliminary histological diagnosis.3-7,10,14,16,18,22 Although this approach has many protagonists, some would follow a more aggressive approach.1,3,8 All would agree that this position is tenable only in institutions where the clinical and radiological expertise, coupled with appropriate equipment, can insure that the diagnosis of malignant brain-stem neoplasm is correct. If any doubt as to the nature of the tumor exists following clinical and radiographic investigation, a biopsy is warranted. In fact, biopsy of the tumor offers many advantages. A tissue specimen is obtained so that grading of the tumor may be performed, although this may be difficult due to sampling limitations.13,19 If a tumor is present, drainage of cystic components and removal of exophytic portions may improve both survival time and quality of survival.2 A more precise knowledge of tumor type and grade may also aid in judgment of the efficacy of current treatment and assessment of any future therapeutic modalities. Perhaps it is of equal importance to rule out any benign, potentially curable tumors that would not respond to irradiation. The present case is an illustration of that point. It is not unique; schwannomas of other cranial nerves have also been mistaken for intrinsic brain-stem tumors.11 Other benign lesions such as neuroepithelial cysts, epidermoid cysts, and dermoid cysts also arise in this region and should be excluded.

References

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