CASE REPORT

Solitary Fibrous Tumor of the Cerebello-Pontine Angle

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ABSTRACT—A case is presented of solitary fibrous tumor occurring in the cerebello-pontine angle. There have been only two other reported cases of a solitary fibrous tumors in this region. Imaging studies showed the tumor to be characteristic in shape and position of an acoustic tumor. However, at surgery the tumor was found to have a “rock hard” consistency. Solitary fibrous tumor differs from acoustic schwannoma and meningioma in its histopathological features and in this case, regrowth, after incomplete excision, was extremely rapid.

Solitary fibrous tumor is a rare neoplasm that has been reported in numerous anatomical areas. It was first described in the pleura, where it was thought to arise from submesothelial fibroblasts. Intracranial solitary fibrous tumor is very rare. A case is presented occurring in the cerebello-pontine angle (CPA). To date there has been only one other report, without detailed clinical features,¹ of a solitary fibrous tumor in this region.

CASE HISTORY

A 50-year-old female presented with a 2-year history of progressive left-sided hearing loss. There was no vertigo, dysequilibrium, or tinnitus. Clinical examination was normal except for a sensorineural hearing loss (Fig. 1). Speech discrimination was 60% with maximum amplification.

Computed tomography (CT) (Fig. 2) revealed a 2-cm lesion in the CPA, consistent with the clinical diagnosis of acoustic tumor. At translabyrinthine surgery, 1 month later, the tumor was found to be extremely firm, indeed almost “rock hard” in consistency. It proved im-

possible to debulk the tumor by known standard methods, however, reduction in bulk was attained by steadying the tumor with a pair of forceps with one hand and using a number 11 scalpel with the other. Frozen sections gave a diagnosis of schwannoma. At the end of the procedure, a small residuum of extremely firm tumor was left attached to the facial nerve, as further dissection would have resulted in its loss. Facial function recovered to House–Brackmann grade 2.

At 1-year review the tumor was visible on CT. The patient was reviewed on a regular basis and by 5 years a significant tumor growth had taken place (Fig. 3) and minor cerebellar signs had developed.

At further resection via a translabyrinthine approach, the greater part of the tumor was found to be of a much softer consistency than previously encountered. However, there remained a residuum of extremely tough nature, which could not be removed without certain loss of the facial nerve. Frozen section, at the time, was inconclusive. Postoperative recovery was prolonged with an increase in cerebellar signs. At 10 weeks the patient had resumed an independent existence with a House–Brackmann grade 2 facial nerve function.

Figure 1. A pure tone audiogram demonstrating a severe sensorineural hearing loss across all frequencies.

Figure 2. This axial CT slice taken through the posterior fossa shows a strongly enhancing rounded mass in the left cerebello-pontine angle. The angle between the mass and the posterior face of the petrous temporal bone is acute and appearances are strongly suggestive of an acoustic neuroma.

Figure 3. This axial CT slice taken through the posterior fossa after intravenous contrast shows a substantial recurrence of the tumour in the left cerebello-pontine angle. A left-sided mastoidectomy defect is also shown.

HISTOPATHOLOGY

The specimens obtained in both 1993 and 1998 showed a spindle cell tumor without atypia or mitoses. Macroscopically, the tissue was white, and so hard that intraoperative smears could not be made. Microscopically, there were alternating sclerotic and cellular areas, the former being more prominent in the first biopsy. The cells in the sclerotic areas had elongated, often wavy nuclei reminiscent of schwannoma. A weakly positive stain for S-100 protein and the fact that a small nerve entered and merged with the tumor at one edge added to the resemblance to schwannoma (Fig. 4). However, the cells had inapparent cytoplasm and were separated from one another by dense intercellular collagen (Fig. 5), unlike the abundant fibrillary cytoplasm seen in typical schwannoma. In the cellular areas, the cells had more cigar-shaped nuclei and some wispy cytoplasm, though there was still abundant intercellular collagen (Fig. 6). The possibility of fibrous meningioma was considered, but there were no whorls or psammoma bodies and epithelial membrane antigen stain was negative. Im-

munoperoxidase stains for vimentin, and CD34 (clone QBEnd/10) were positive, and Dako cytokeratins 19 and MNF 116, S-100 protein, desmin, smooth-muscle actin, and glial fibrillary acidic protein were negative. Progesterone receptors were positive in the majority of tumor cell nuclei. Oestrogen receptors were negative and Ki-67 proliferation antigen was positive in less than 10% of tumor cell nuclei. Electron microscopy showed mesenchymal-type spindle cells in a collagenous stroma, with occasional primitive intercellular junctions, but no specific feature of meningotheial, Schwann cell, or muscle differentiation. The histology was typical of solitary fibrous tumor of the meninges.

DISCUSSION

Acoustic tumor (vestibular schwannoma) is the most common tumour in the CPA. Meningioma accounts for about 5% of tumours² and various other tumors in this area have been reported. These include arachnoid cyst, lipoma, facial nerve schwannoma,

Figure 4. Section of the resected tumour in 1993 showing bland, "wavy" spindle cells resembling schwannoma, with a small myelinated nerve entering the tumor. Unlike typical schwannoma, there is abundant intercellular collagen. Dilated, thin-walled vessels are seen. (Hematoxylin and eosin, original magnification $\times 20$.)

Figure 5. Recurrent tumour biopsied in 1998. Hypocellular sclerotic area of tumor. (Hematoxylin and eosin, original magnification $\times 20$.)

metastases, teratoma, epithelial cyst, cavernous haemangiomas, and choroid plexus papilloma.²⁻⁴

CT and magnetic resonance imaging (MRI) are generally, but not always, able to differentiate these lesions.

CT and MRI (Figs. 2 and 3) in this case was suggestive of an acoustic tumor, but there was some minor stippling on CT (not demonstrated on photography) due to calcification, which is more common in, but not exclusive to, meningioma.⁵

The term "solitary fibrous tumor" was first used to describe a group of benign pleural tumours.⁶ Subsequently, such tumors have been described in the abdominal cavity, orbit, upper respiratory tract, soft tissue, salivary gland, infratemporal fossa, and spinal cord. It has been noted that extrapleural solitary fibrous tumors exhibit benign behavior and only rarely recur or manifest malignant behavior.⁷

The essential features⁷ are those of a circumscribed tumor of bland spindle cells with little cytoplasm, closely admixed with collagen fibers. The cell arrangement is not distinctive and mitotic rate is low. Alternat-

ing hypocellular and hypercellular areas are characteristic. Positive immunoperoxidase staining for CD34 is found in 80–100%, depending on the antibody used, MY 10 being more sensitive than QBEnd/10. The result of positive progesterone and negative oestrogen receptors and low proliferation marker (Ki-67 antigen) in this case was consistent with other reports of intracranial solitary fibrous tumors.^{1,7}

Intracranial solitary fibrous tumors have rarely been reported^{1,7-9} and there have been only two cases of a solitary fibrous tumor reported in the CPA; that histologic report described seven cases of solitary fibrous tumor of the meninges, five of which were intracranial.

Acoustic tumor recurs rarely,¹⁰ even when a small tumor segment remains, an experience mirrored in the St. Vincent's Hospital department. Meningioma similarly can be resected completely from the CPA, whereas meningioma in the skull base can be difficult to excise completely.¹¹ In this case of solitary fibrous tumor, a small residuum was left *in situ* to preserve the facial nerve, but the tumor otherwise was resected fully and subsequent regrowth was extremely rapid.

Figure 6. Hypercellular area of recurrent tumor with elongated, blunt-ended nuclei, but inapparent cytoplasm. The intercellular material is all collagen. The tumor resembles fibrous meningioma, but there are no whorls or psammoma bodies. (Hematoxylin and eosin, original magnification $\times 20$.)

CONCLUSION

Solitary fibrous tumor of the meninges is very rare. It differs from acoustic tumor and meningioma in its histopathological features and in this case, regrowth, after incomplete excision, was extremely rapid. At surgery the tumor was found to have a "rock hard" consistency and imaging studies (CT) show the tumor, which was characteristic in shape and position of an acoustic tumor.

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