Facial Nerve Schwannoma of the Cerebellopontine Angle: A Diagnostic Challenge

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ABSTRACT

Facial nerve schwannomas are rare lesions that may involve any segment of the facial nerve. Because of their rarity and the lack of a consistent clinical and radiological pattern, facial nerve schwannomas located at the cerebellopontine angle (CPA) and internal auditory canal (IAC) represent a diagnostic and therapeutic challenge for clinicians. In this report, a case of a CPA/IAC facial nerve schwannoma is presented. Contemporary diagnosis and management of this rare lesion are analyzed.

KEYWORDS: Facial nerve schwannoma tumors, cerebellopontine angle (CPA), facial paralysis, magnetic resonance imaging (MRI)

Facial nerve schwannomas are uncommon benign tumors. About 300 cases have been reported to date. Although these tumors may involve any segment of the facial nerve, a predilection for the geniculate area has been reported.¹⁻³ Cerebellopontine angle (CPA) facial schwannomas are infrequent, accounting for less than 20% of facial schwannomas⁴ and about 3% of CPA tumors.⁵

In addition to their rarity, several factors make CPA facial schwannomas a diagnostic and

therapeutic challenge for clinicians. First, their clinical presentation is variable and facial function can be normal. Second, multiple radiological patterns have been reported and none of them are pathognomonic. The timing and best approach for surgical treatment are also controversial. We present a patient with a CPA facial nerve schwannoma that extended into the internal auditory canal (IAC) and analyze the contemporary diagnosis and management of this uncommon disease.

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CASE REPORT

A 56-year-old woman presented with a 1-year history of unsteadiness, accompanied by hearing loss and tinnitus in the right ear. She had no history of neurofibromatosis, previous facial nerve surgery, or head trauma. Examination showed very mild weakness on the lower face. Audiometric studies revealed a moderate high-frequency sensorineural hearing loss with a 70% speech discrimination score at 80 dB. Preoperative magnetic resonance imaging (MRI) showed an enhancing cerebellopontine angle (CPA) tumor with considerable extension to the IAC (Figs. 1, 2). Volitional electromyography was unremarkable. Evoked electromyography revealed a decreased peak-to-peak amplitude of the compound muscle action potential on the right side (1.5 mV) compared to the left side (2.5 mV).

The patient underwent a retrosigmoid exploration. The vestibular and cochlear nerves were

Figure 1 Axial T1-weighted, gadolinium-enhanced MRI at the level of the right IAC, showing a 20 x 10 mm tumor with necrotic areas. The tumor is concentric to the IAC.

Figure 2 Coronal T1-weighted, gadolinium-enhanced MRI at the level of the right IAC. The lesion involves both the CPA and the IAC. No enhancement of the geniculate fossa is present.

pushed posterosuperiorly by the tumor, which originated from the facial nerve (Fig. 3). The tumor, including the involved facial nerve segment, was resected completely. A sural nerve graft was interposed between the proximal and distal ends of the facial nerve. The latter could be seen at the fundus of the IAC. Fibrin glue was used to stabilize the anastomosis (Fig. 4).

Histologically, the lesion was characterized by interlacing bundles of spindle-shaped cells arranged in palisades (Fig. 5). Microcystic and hemorrhagic areas were present. The tumor showed no necrotic areas or mitosis. A schwannoma was diagnosed based on the pathologic analysis.

Postoperative audiometry showed no residual hearing, and the patient sustained complete facial paralysis. Fifteen months after surgery the facial nerve was House-Brackmann grade IV.

Figure 3 Operative photograph (right ear) of a retrosigmoid approach showing the cochlear and vestibular nerves (arrow) displaced posterosuperiorly by a facial nerve schwannoma. The posterior surface of the tumor has been partially coagulated.

DISCUSSION

In recent literature, CPA and IAC tumors represented a minor percentage of facial nerve schwannomas.^{4,6} Facial nerve schwannomas usually manifest with facial palsy, often with an insidious onset. Recurrent facial paralysis, hemifacial spasm, facial twitching, and facial pain are other presenting symptoms.⁷ Normal facial function has been reported in 27 to 54% of cases.^{6,8,9} A small but significant percentage of patients demonstrates some degree of facial dysfunction when evaluated by careful explo-

Figure 4 Operative photograph showing the nerve anastomosis in the IAC after complete tumor resection.

Figure 5 Photomicrograph of facial nerve schwannoma showing Schwann cell proliferation with the characteristic palisade organization.

ration, electrical studies, or both.¹⁰ Of 65 acoustic neuromas treated surgically at our institution, 5% were associated with abnormal clinical preoperative facial function.¹¹ Hearing loss and tinnitus are common among patients with facial nerve schwannomas. CPA and IAC facial schwannomas often show retrocochlear findings, increasing the probability of misdiagnosing these lesions as acoustic neuromas.¹²

CT imaging provides useful information for preoperative planning, but MRI is the method of choice for evaluating CPA lesions, including facial nerve schwannomas. These lesions appear hypointense relative to the brain on T1-weighted images, hyperintense on T2-weighted images, and enhance briskly with gadolinium. An isointense appearance on both T1- and T2-weighted images has also been reported.¹³ Enhancement of the geniculate ganglion and the distal facial nerve; the existence of a double component, one in the CPA/IAC and one in the middle cranial fossa; and an enhancing enlargement of varying thickness along a significant length of the nerve^{14,15} are very suggestive of a facial nerve schwannoma. Lesions confined to the CPA/IAC mimic acoustic neuromas.

Fagan et al¹² suggested some MRI signs that may help to distinguish CPA acoustic neuromas

from facial schwannomas. These authors stated that a posterior eccentricity to the axis of the IAC in CPA tumors smaller than 2 cm might help diagnose a facial nerve schwannoma preoperatively. No posterior eccentricity to the IAC was present in our patient (Fig. 1). In fact, a slight anterior placement could be seen. Moreover, in a review of our series of 65 acoustic neuromas, five cases had a posterior eccentricity to the IAC, but none of them corresponded to a facial nerve schwannoma.

The treatment of facial nerve schwannomas consists of removing the tumor with the involved facial nerve segment and restoring facial function with a cable graft or a nerve crossover graft. The surgical approaches are the same as those used to treat acoustic neuromas, that is, translabyrinthine, middle fossa, and retrosigmoid approaches, alone or in combination, depending on the size and location of the tumor. In their interesting study, Fagan et al¹² stated, "it is not possible to resect and graft a CPA facial nerve and preserve hearing" because CPA facial nerve schwannomas should be treated through a translabyrinthine approach. Curiously, their four cases were treated through a retrosigmoid approach, and no postoperative hearing results were reported. Although our patient's tumor extended to the lateral end of the IAC and her hearing could not be preserved, we believe that the retrosigmoid is a reliable approach to the CPA for resecting both acoustic and facial schwannomas, and hearing preservation should be attempted when possible.

The optimal timing of treatment for facial nerve schwannomas is also controversial. Because postoperative facial function is usually poor in these patients, some authors advocate a policy of close follow-up with serial MRI.¹⁶ Surgery would be indicated when facial paralysis progresses or when significant compression of vital structures is noted. Liu and Fagan¹⁷ have recently published an interesting report about conservative management of facial nerve schwannomas. In their study "conservative management" consisted of surgical exploration with or without partial tumor resection in 9 of 10

patients. These 9 patients had tumors confined to the CPA/IAC. In the remaining patient, the only one who did not require surgery, the MRI showed involvement of the geniculate ganglion. This study confirms the difficulty of diagnosing a CPA/IAC facial schwannoma before surgical exploration as no specific clinical or radiological tools are available for this entity. Because the outcome of a biopsy or partial resection on postoperative facial function may be unpredictable, intraoperative decisions should be individualized.

In summary, facial nerve schwannomas are rare CPA lesions. Facial dysfunction may alert clinicians to the possibility of a facial nerve tumor. Initially, however, the lack of a consistent clinical and radiological pattern usually leads to a misdiagnosis of acoustic neuroma, delaying the definitive diagnosis until surgery.

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Commentary

The authors present an interesting case of a 56-year-old woman with a right-sided cerebellopontine facial nerve neuroma that extended into

the internal auditory canal. The authors chose the retrosigmoid approach to preserve preoperative hearing. Intraoperatively, they found that the tumor involved the facial nerve as evidenced by the presence of the vestibular and cochlear nerves as shown on intact intraoperative photograph. These nerves were displaced superoposteriorly. The authors indicated that complete resection was achieved. That they could achieve a sural nerve graft is truly surprising, especially given that the tumor extended almost a centimeter into the internal auditory canal. Whether the anastomosis will provide some recovery of facial function remains to be seen. Not surprisingly, the patient had complete facial paralysis.

It is easy to play armchair quarterback; however, if we find what appears to be a facial neuroma during an approach to a cerebellopontine angle tumor, we seldom recommend obtaining a biopsy of these tumors, let alone resecting them. In our experience and that of others, complete facial paralysis would be guaranteed. If these small, benign tumors were to enlarge, radiosurgery or possibly open surgery could be considered.

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