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Case Report

A rare case of greater petrosal nerve schwannoma

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Abstract

Background: Facial nerve schwannomas include only 0.8% of all intrapetrous mass lesions, and schwannomas originating exclusively from the greater petrosal nerve (GPN) are extremely rare. To date, only 13 reports have been described. In this case, the tumor was thought to originate from the GPN on the basis of clinical, radiological, and operative findings.

Case Description: A 23-year-old girl presented an acute left facial palsy, a disturbance in tear secretion of the ipsilateral eye, and a left-sided conductive hypoacusia. Computed tomography (CT) scan and magnetic resonance imaging (MRI) showed an extradural mass in the left middle fossa. A subtemporal approach was performed and the lesion, originating from the proximal portion of the GPN, was excised. The post-operative course was satisfactory, except for a xerophtalmia, which was treated with artificial teardrops.

Conclusion: GPN schwannomas can originate anywhere alongside the course of the nerve, from its proximal segment near the facial hiatus to its distal segment near the foramen lacerum. For these reasons, it requires differential diagnosis with trigeminal nerve schwannomas or with injuries arising from the geniculate ganglion, because it can be easily confused with those lesions. However, in less severe cases, an early diagnosis can be able to preserve the function of the facial nerve by reducing iatrogenic injuries caused by surgical maneuvers.

Key Words: Facial nerve, greater petrosal nerve, schwannoma

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INTRODUCTION

Facial nerve schwannomas are rare and include only 0.8% of all intrapetrous mass lesions.[4,13,18-20,22] These particular tumors can arise from any segment of the nerve, but lesions originating from the greater petrosal nerve (GPN) are infrequently described.

In this study, our aim is to report a case of GPN schwannoma in a young woman by discussing clinical aspects, radiological features, surgical treatment and operative findings, together with reviewing the current literature.

CASE REPORT

A 23-year-old girl was referred to our department for an acute left facial palsy (Brackmann-House grade IV) associated with a disturbance in tear secretion of the

ipsilateral eye, as supported by Schirmer's test. An audiogram revealed a left-sided conductive hypoacusia, while Brainstem auditory evoked response (BAER) showed an increase in latency of waves III and IV on the left side.

A computed tomography (CT) scan revealed an isodense subtemporal mass, with a partial calcification of the rim and the erosion of the anterior aspect of the petrous bone, with a clear extension into the tympanic cavity [Figure 1]. Magnetic resonance imaging (MRI) confirmed the presence of an extradural mass (3.8 x 3.3 x 2.8 cm) in the middle cranial fossa, isointense on T1-weighted images and mildly hyperintense on T2-weighted images, causing a compression and dislocation of the temporal lobe [Figure 2].

The tumor was exposed by a left subtemporal extradural/interdural approach while continuously monitoring facial nerve electromyography; during the mobilization of the postero-inferior portion of the tumor, the GPN was identified [Figure 3]. The lesion was removed by cutting the nerve that could not be dissected from the lesion, thus minimizing traction on the geniculate ganglion; the tympanic opening and the mastoid air cells were filled with autologous abdominal subcutaneous fat tissue. Post-operative CT scan and MRI showed the complete resection of the lesion [Figure 4]. The histological evaluation revealed a typical biphasic schwannoma, containing both Antoni A and Antoni B tissue [Figure 5].

The post-operative course was satisfactory, except for a left xerophtalmia treated with artificial teardrops. After 6-month follow up, both hearing difficulties, supported by audiometric test, and facial palsy regressed (Brackmann-House grade I-II).

DISCUSSION

From 1936 to 2010, only 13 reports, accounting for a total of 22 GPN schwannomas, have been described in literature.[1-3,5,7,11-13,16,17,19-21] Both the average age of the patients (40 years) and the female prevalence (13:9) are consistent with the available data concerning the same lesions in other anatomical sides [Table 1].

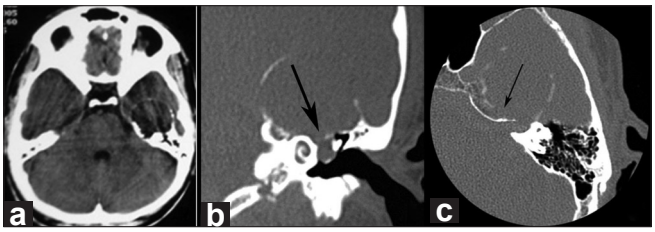

Figure 1: CT scan shows an isodense mass in the temporal fossa (a). The bone-window CT scan, with coronal image (b), shows a clear extension into the tympanic cavity; the axial view (c) reveals the erosion of the anterior aspect of the petrous bone (arrows)

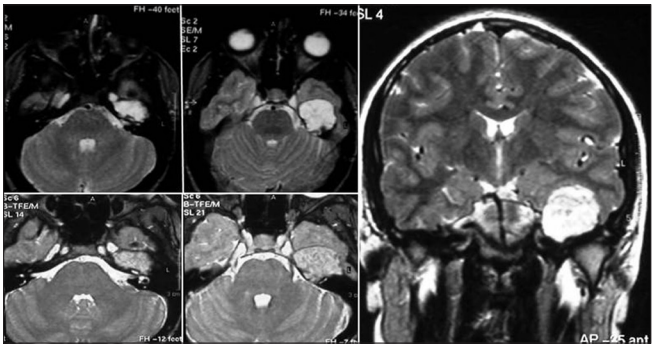

Figure 2: Pre-operative MR images shows a mass hypo- or isointense relative to the brain on T1-weighted and heterogeneously hyperintense on T2-weighted images with enhanced after administration of gadolinium developing from the facial hiatus to the foramen lacerum of 3.8 × 3.3 × 2.8 cm

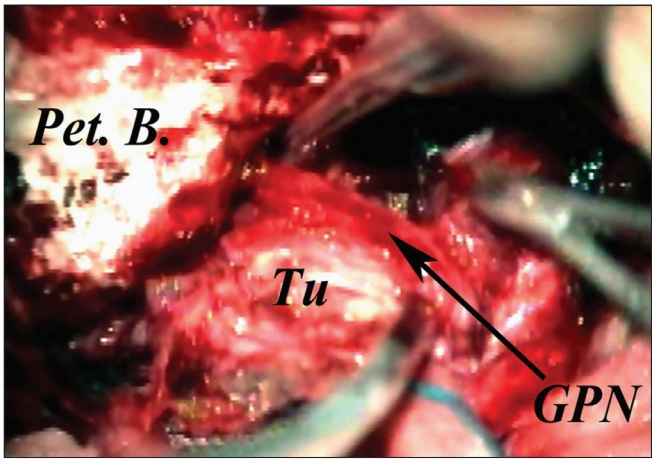

Figure 3: Intraoperative image showing the lesion arising from the greater petrosal nerve. GPN: greater petrosal nerve; Pet. B.: petrous bone; Tu: tumor

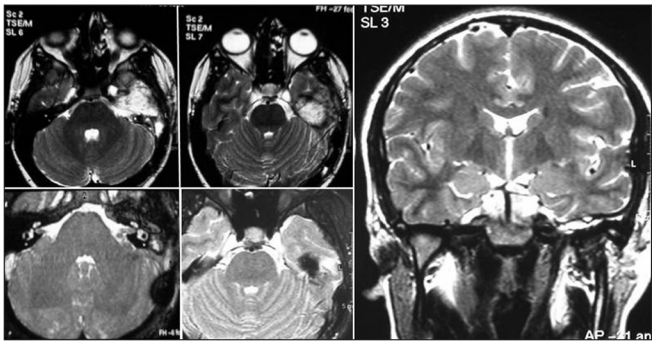

Figure 4: Post-operative MR images demonstrate the complete removal of the tumor. The tympanic opening was filled with autologous abdominal subcutaneous fat tissue

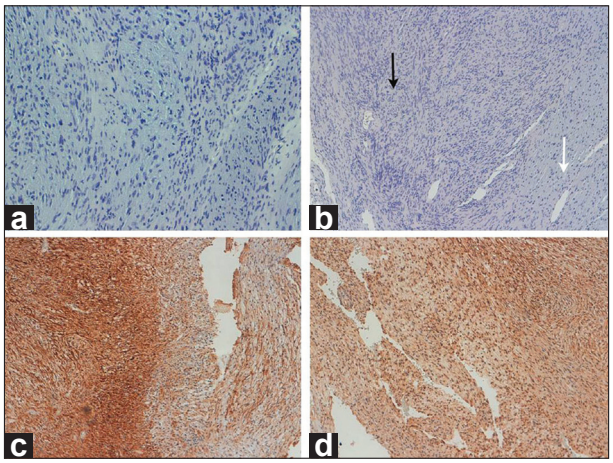

Figure 5: Microscopic and immunohistochemical examination showing nuclear hyperchromasia and pleomorphism of the schwannoma (a). The cellularity of the tumor, positive for S-100 (c and d) and Vimentin, is biphasic containing both Antoni A (black arrow) and Antoni B (white arrow) tissue (b)

Most studies dealing with lesions arising from GPN described facial palsy and hearing difficulties,[5,7,12,13,16,20,21] while Mori *et al*.[17] reported a rare case without facial palsy, but with only hearing loss. The latter is mostly conductive secondary to tympanic cavity extension of the tumor,[5,7,12,13,16,17,21] but if the inner ear or cochlear nerve are involved, there may be also a sensorineural hearing loss.[4,20,13,19] In fact, Sade *et al*.[19] described a case with only low-frequency hearing loss because of a tumor which, growing from GPN, involved the cochlear apex. The decrease of tear secretion with xerophthalmia is considered pathognomonic for GPN schwannoma,[3] but Schirmer's test was not always performed [Table 1].[2,3] In few cases, right 6th nerve palsy,[3] trigeminal affection,[11] headaches, generalized seizures, eye pain and abnormal hysterical behavior were observed [Table 1].[2,7] This range of minimal symptoms is highly correlated to the location, dimension and development of the lesion in the temporal bone[2,22] although such an uniform clinical presentation seems to be unlikely [Table 1]. Nevertheless, a close neurological examination is always mandatory to reveal the delicate points denoting the origin of the lesion from the petrosal branch of the facial nerve.

If properly interpreted, the pre-operative imaging can be quite diagnostic in performing differential diagnosis of the lesions. A GPN schwannoma shows an epidural mass in the middle cranial fossa developing from the facial hiatus to the foramen lacerum,[1,2,6,9-11,13-15,20,22] thus compressing both the temporal base[13] and the temporal lobe.[20,22] It is possible to misunderstand the origin of the tumor from the geniculate fossa or from the GPN.[13] As a rule, lesions arising from the geniculate ganglion have a "bulbous" enlargement at the geniculate fossa, while lesions arising from the GPN scallop the anterior margin of the geniculate fossa and the adjacent bony petrous apex.[2,9,13,22] The tumor can produce the erosion of the temporal bone, thus inducing a characteristically smooth defect on top of the petrous bone and eroding the anterior aspect of the petrous bone, affecting more its midportion than the apex, which is commonly eroded by trigeminal schwannomas.[3,13,20,22] The eventual erosion near the facial hiatus can produce the expansion into the tympanic cavity and to the mastoid air cells;[20] however, if the mass is big enough[3] or if it arises from the distal segment of the GPN,[11] apex and midportion of the petrous bone, foramen lacerum and carotid canal may be eroded by the tumor.[3,11,13] In these cases, an angiography is indicated to evaluate the contact and eventual displacement of the internal carotid artery by the tumor, and to exclude the origin of the lesion from the deep petrosal nerve. Whenever an exploration near the carotid artery is needed during the surgical procedure,[2,3,5,11,13,16,20,21] the execution of a balloon occlusion test can be considered as well.[11]

As shown in the existing literature, there may be different approaches to a GPN schwannoma [Table 1]. Most authors have performed a middle fossa approach,[1-3,7,11,13,16,17,19,20] but three studies reported a middle fossa exploration and/or mastoidectomy.[5,12,21] The GPN normally runs in the interdural space at the base of the middle fossa, therefore a subtemporal epi- and inter-dural approach is considered an ideal option to remove GPN schwannoma.[7] When the lesion is more than 3-4 cm in diameter and it is mostly located within the temporal fossa rather than within the petrous bone, a sub-temporal approach with a combined extra/intradural avenue could be preferred, with particular concern about the main trunk of the facial nerve and the small branches coming from the internal carotid artery toward the capsule of the tumor. Both the size of the tumor and its main symptomatology would indicate whether an inter- or extradural approach should be undertaken as the first step.[2] Normally, if the lesion arises from the GPN, it does not show more proximal involvement than the facial hiatus, and it occupies the Glasscock triangle of the middle fossa floor.[19] Ahihara *et al*.[1] suggested that drilling to the middle fossa is mandatory to facilitate tumor removal, but in our experience it was not necessary because the lesion itself eroded the bone plane. Nevertheless, if the schwannoma originates from the distal portion of the GPN, the execution of the Kawase's petrosectomy[8] is helpful to preserve the internal carotid artery by surgical procedure following the intimate relationship of the tumor with the petrous and cavernous segments of the artery itself.[11] However, the definite sealing of the mastoid air cells is mandatory to prevent post-operative cerebrospinal fluid leakage.

CONCLUSIONS

GPN schwannoma, although rare, can originate anywhere

Table 1: Reviewing of the literature of the greater petrosal nerve schwannomas

Authors and year	No. of cases	Pt. age (years) and sex	Symptoms at diagnosis	Schirmer's test	Operation	Size	Postoperative status
Tremble and Penfield 1936	1	42, M	Facial palsy Hearing difficulties Tinnitus	Not reported	Radical mastoidectomy	Not reported	Not reported
Kleinasser and Friedman 1959	1	19, F	Facial palsy Hearing difficulties	Not reported	Middle fossa exploration	Egg sized	Not change
Furlow 1960	2	48, M 44, M	Facial palsy Hearing difficulties Generalized convulsion (in both cases)	Not reported	Middle fossa exploration (1st case) Temporal craniotomy and mastoidectomy (2nd case)	Tennis-ball sized (1st case) Large (2nd case)	Not change
Kumon 1999	1	21, F	Facial palsy Hearing difficulties Decrease of the tear secretion	Not reported	Extradural middle fossa approach	2.5 × 2.5 × 2 cm	Not facial palsy Useful hearing
Michel 2000	1	20, F	Vertigo Facial palsy Hearing difficulties	Not reported	Extradural middle fossa approach	Not reported	Not facial palsy
Kinouchi 2001	2	58, F 49, F	Severe vertigo (case 1) Trigeminal affection (case 2)	Not reported	Extradural subtemporal approach (In both cases)	Not reported	Lacrimation deficit (in case 1) Not deficit (in case 2)
Ahihara 2001	1	65, M	Acute facial palsy	Not reported	Extradural middle fossa approach	1.5 × 1.5 × 2 cm	Facial palsy improving Hearing improving Decrease of the tear secretion
Schmidinger 2005	1	65, F	Facial palsy Hearing difficulties Occasional tinnitus	Not reported	Extradural subtemporal approach	3.5 × 3.0 × 2.5 cm	Facial palsy increased Hearing difficulties not changed
Mori 2007	1	50, M	Hearing loss	Not reported	Extradural middle fossa approach	Not reported	Not deficit
Sade 2007	1	63, F	Facial palsy Hearing difficulties	Not reported	Extradural middle fossa approach	2.4 cm	Facial palsy improving Useful hearing
Ayberk 2008	1	16, F	Diplopia Headache Decrease of the tear secretion	Reported	Extradural subtemporal approach	2.5 × 2.0 × 3.0 cm	Not deficit
Amirjamshidi 2009	5	22, F 25, M 28, M 28, M 54, F	Chronic headaches Generalized seizures (case 1) Eye pain (case 2) Headache and Eye pain (case 3) Eye pain (case 4) Hysterical behavior generalized headache (case 5)	Reported in all cases	Extra/intradural subtemporal approach (in cases 1 and 4) Extradural subtemporal approach (in cases 2, 3 and 5)	Not reported	Facial palsy improving after 1 year (in cases 1 and 2) Not palsy (in case 4) Satisfactory, except for a xerophthalmia treated with artificial teardrops (in cases 3 and 5)

Table 1: Contd....

Authors and year	No. of cases	Pt. age (years) and sex	Symptoms at diagnosis	Schirmer's test	Operation	Size	Postoperative status
Ichimura 2010	4	25, F 27, M 35, F 49, F	Xerophthalmia (case 1) Convulsions and facial palsy (case 2) Xerophthalmia and facial palsy (case 3) Xerophthalmia and hearing difficulties (case 4)	Not reported in all cases	Extra/Interdural subtemporal approach in all cases	Not reported	Transient facial palsy with xerophthalmia (cases 1 and 3) Facial palsy (case 2) Not change after surgery (case 4)
Present case	1	23, F	Facial palsy Hearing difficulties Decrease of the tear secretion	Reported	Extra/Interdural subtemporal approach	3.8 × 3.3 × 2.8 cm	Xerophthalmia treated with artificial teardrops, ipsylaterally to the lesion

alongside the course of the nerve from its proximal segment near the facial hiatus to its distal segment near the foramen lacerum. For these reasons, it requires differential diagnosis in order to be distinguished from trigeminal nerve schwannomas or from injuries arising from the geniculate ganglion, because it can be easily confused with those lesions. However, when the lesion is small, an early diagnosis can be able to preserve the function of the facial nerve by reducing iatrogenic injuries of the surgical maneuvers.

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