Indian Journal of Surgical Oncology (March 2019) 10(1):101–106
https://doi.org/10.1007/s13193-018-0850-7

CASE REPORT

Management Dilemmas of Intraparotid Facial Nerve Schwannoma: Report of Four Cases and Review of Relevant Literature

Roshan K. Verma[1] · Neemu Hage[1] · Amit Bahl[2] · Amanjeet Bal[3] · Naresh K. Panda[1]

Received: 9 August 2018 / Accepted: 29 November 2018 / Published online: 3 January 2019

Introduction

Facial nerve schwannomas are benign, encapsulated tumors that arise from the facial nerve sheath [1–3]. Schwannomas of the facial nerve can arise from either the extratemporal or intratemporal course of this nerve [4]. Facial nerve schwannomas most commonly arise from intracranial and intratemporal part of the facial nerve. Facial nerve schwannoma arising from the extratemporal course of facial nerve accounts for only 9% of all facial nerve schwannomas [5]. Intraparotid facial nerve schwannoma is an even rarer entity and accounts for 0.5–1.5% of all parotid tumors [6]. Literature search has have described less than 100 cases of intraparotid facial nerve schwannomas.

Intraparotid facial nerve schwannoma is a diagnostic challenge due to its rarity and non-specific radiological features. It may often be confused with other benign tumors of parotid as was the case in one of our patients. Facial nerve schwannoma must be considered as a possibility in a slow-growing parotid mass with slowly progressive facial paresis. Fine needle aspiration cytology is not always reliable in making diagnosis of facial nerve schwannoma and may be reported as pleomorphic adenoma. Facial nerve schwannoma should be suspected when tumor is inseparable from the facial nerve trunk. Decision to operate or leave the tumor while preserving facial nerve function should be made judiciously and in consultation with the patient and his relatives.

The purpose of this study is to elicit the variation in presentations in patients with intraparotid facial nerve schwannomas and a literature review of the management protocols seen over the years.

Methods

Retrospective review of database of 200 parotid surgeries performed from January 2001 to June 2018 revealed four cases of intraparotid facial nerve schwannomas. Their clinical presentation, fine needle aspiration cytology, imaging findings and surgical details, and post-operative facial nerve function statuses were recorded.

Case 1

A 34-year-old female presented with a 1 ½-year history of a painless, gradually progressive swelling in the left parotid region. There was no history of facial weakness or asymmetry at presentation. Examination revealed a single, firm, non-tender swelling of approximately 2 × 2 cm. The facial nerve function was intact. Fine needle aspiration was suggestive of a nerve sheath tumor. Contrast-enhanced computed tomography showed an ill-defined heterogeneously enhancing mass lesion in the posterior pole of the superficial lobe of the parotid. Contrast-enhanced MRI showed a well-defined smooth marginated T1 isointense and T2 hyperintense mass lesion at subcutaneous plane in the left parotid region measuring 1.7 × 1.6 × 2.4 cm. The lesion showed diffuse restriction and homogenous contrast enhancement with central non-enhancing area suggestive of central necrosis

✉ Roshan K. Verma
roshanverma@hotmail.com

[1] Department of Otolaryngology and Head and Neck Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

[2] Department of Radiotherapy, Postgraduate Institute of Medical Education and Research, Chandigarh, India

[3] Department of Histopathology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

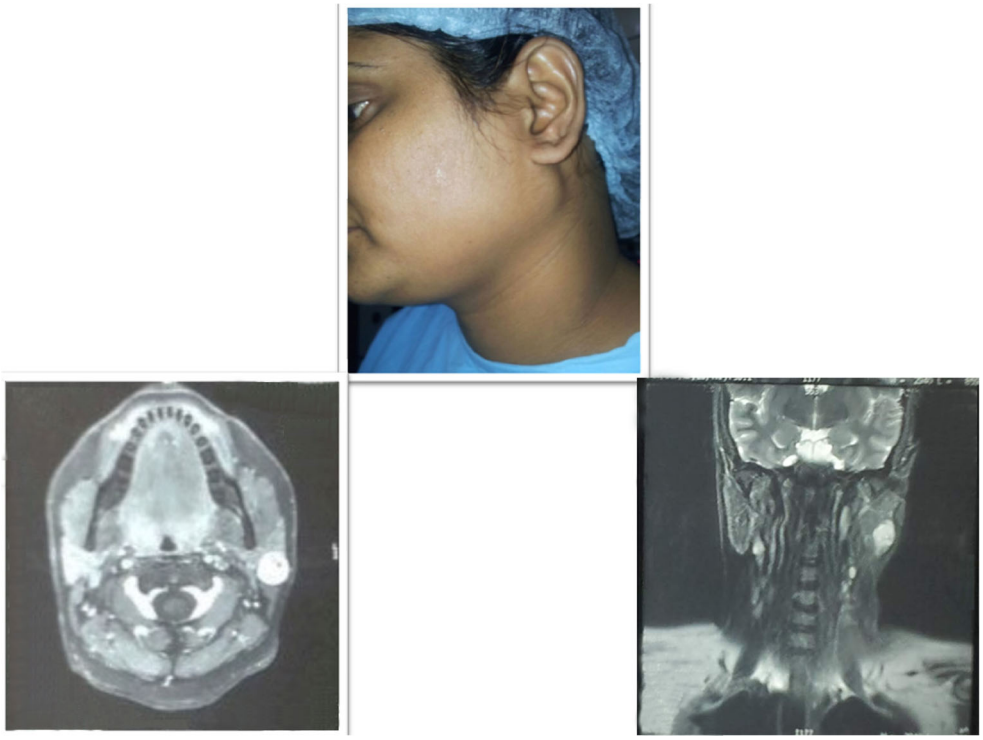

Fig. 1 **a** Preoperative clinical photo of case 1 showing 2 × 2-cm swelling in the left infra auricular region. **b** Contrast-enhanced MRI of case 1 showing a well-defined smooth marginated T1 isointense and T2 hyperintense mass lesion with homogenous contrast enhancement in the left parotid region

(Fig. 1). The patient was advised for regular follow-up but insisted on surgery.

A left superficial parotidectomy was performed. Facial nerve trunk was identified, and all the peripheral branches of the facial nerve were dissected. A 2 × 2-cm firm nodule was seen in the superficial lobe that appeared to be inseparable from the upper buccal branch of the left facial nerve which was sacrificed. The nodule was dissected out with a cuff of healthy tissue. Post-operatively, the patient had mild marginal mandibular nerve paresis which resolved over a period of 3 months with conservative management. Post-operatively, the diagnosis of facial nerve schwannoma was confirmed on histopathology (Fig. 1).

Case 2

A 19-year-old female presented with a 1-year history of gradually progressive swelling in the right parotid region. She complained of occasional dull pain over the swelling. There was no facial asymmetry at presentation. Clinical examination revealed a firm, mobile, tender swelling of about 1.5 × 1.5 cm in the right infra auricular region. Her facial nerve function was intact. Fine needle aspiration cytology was done which was reported as benign salivary aspirate. Contrast-enhanced computed tomography showed a round to oval, heterogeneously enhancing well-defined mass lesion measuring around 2 × 1.7 × 1.8 cm in the superficial lobe of the right parotid gland.

Right superficial parotidectomy was done. Intra-operatively, approximately 2.5 × 2-cm yellowish firm nodular lesion was seen arising from the upper buccal branch of the right facial nerve. The tumor was excised with a cuff of normal tissue while preserving the lower buccal branch of the facial nerve. All the branches of facial nerve were identified and preserved. Post-operatively, her facial nerve functions were preserved, and the House-Brackmann (HB) grading was I at 1-year follow-up. Histopathological examination of the specimen confirmed the diagnosis of a facial nerve schwannoma (Fig. 2).

Case 3

A 37-year-old male presented with a 4-year history of slowly progressive painless swelling in the right parotid region with facial asymmetry. He had a history of surgical intervention 3 years ago in another institution after which he developed facial asymmetry. The histological report was that of a schwannoma.

Examination revealed a non-tender, soft to firm 7 × 8-cm swelling involving the right parotid region with a hypertrophic scar over the previous surgical incision line. There was a pre-existing grade III (House-Brackmann) facial nerve paresis on the right side at presentation.

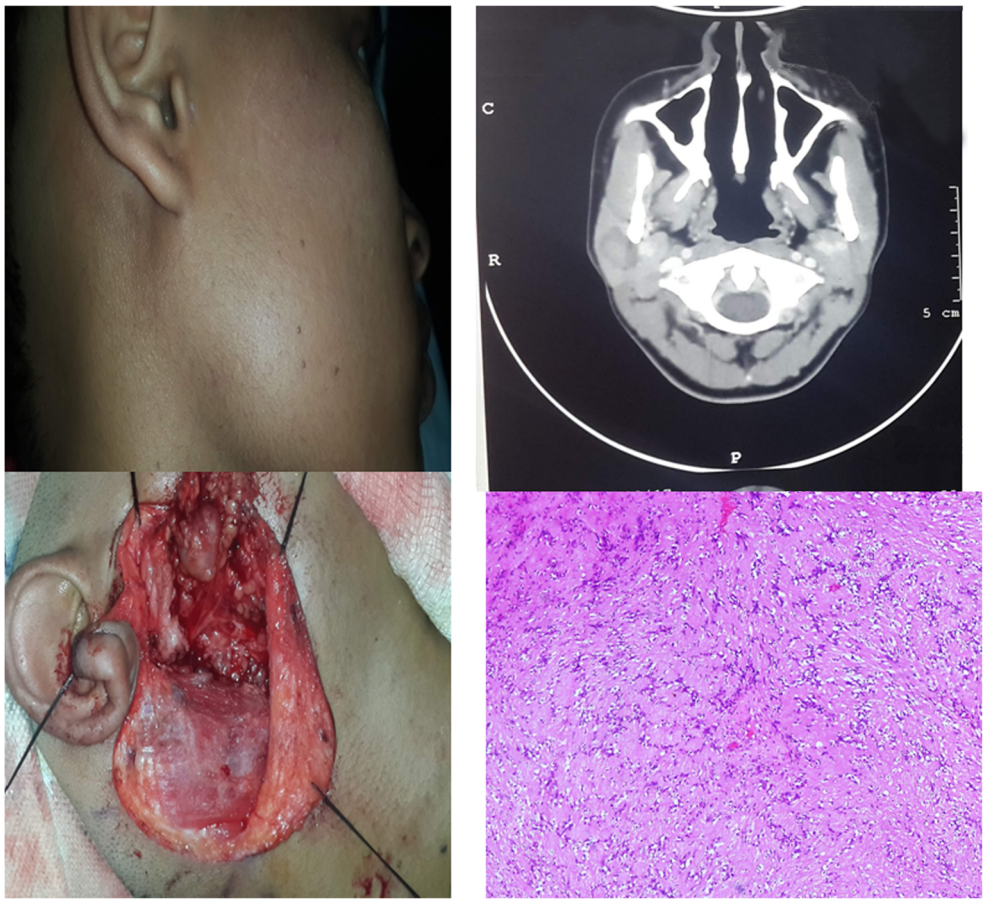

Fig. 2 **a** Preoperative clinical photo of the case. **b** Contrast-enhanced CT coronal cut of case 2 showing heterogeneously enhancing, well-defined mass lesion in the superficial lobe of the right parotid gland. **c** Intraoperative photo of case 2 showing 2.5 × 2-cm firm nodular lesion arising from the lower buccal branch of the right facial nerve. **d** Hematoxylin and Eosin–stained section shows a well mass composed of densely packed spindle cell with characteristic nuclear palisading

Contrast-enhanced MRI revealed right parotid enlargement with focal lobulated and well-defined heterogeneous lesion measuring 7.6 × 3.9 × 4.4 cm. Heterogeneous signals with mixed iso- and hyperintense in T1 with subtle T2 intense cystic areas were noted at the periphery of the lesion. Post-contrast heterogeneous enhancement with few enhancing septae and non-enhancing cystic areas was noted.

Right superficial parotidectomy was done. Intraoperatively, there was a firm encapsulated tumor measuring 7 × 5× 3 cm situated deep into the superficial lobe, excised in along with the superficial lobe of the parotid. Facial nerve trunk was inseparable from the tumor and had to be sacrificed, and cable nerve grafting with great auricular nerve was done. Histopathological examination confirmed the diagnosis of schwannoma. Post-operatively, his facial palsy was HB IV (Fig. 3).

Case 4

A 45-year-old female presented with a 3-month history of painless, gradually progressive swelling of the left parotid region. There was no history of facial weakness, facial twitching, or numbness. On examination, there was a single 4 × 3-cm firm, non-tender, mobile swelling over the lower part of the left parotid gland. Facial nerve function was normal. Contrast-enhanced computed tomographic scan showed lobular, well-defined homogenous enhancing mass involving the deep lobe of the parotid gland (Fig. 4). Fine needle aspiration cytology of the lesion showed salivary aspirate.

Total conservative parotidectomy was done. The branches of the facial nerve were identified. Intraoperatively, about 4 × 3-cm cystic mass with solid component was found involving deep lobe of the parotid gland. The lower buccal branch was found adhered to the tumor and was dissected with great difficulty. The upper buccal and the platysmal branches were lifted with the help of nerve retractor, and the tumor was removed. Post-operatively, the patient developed deviation of the angle of the mouth which improved after 6 months. Histopathology of the excised mass showed spindle-shaped cells arranged in characteristic Verocay bodies. The neoplastic cell showed nuclear and cytoplasmic immunopositivity with S-100 and negative for smooth muscle actin. A diagnosis of schwannoma was confirmed. Her post-operative facial nerve function was HB grade II at 1-year follow-up (Fig. 4).

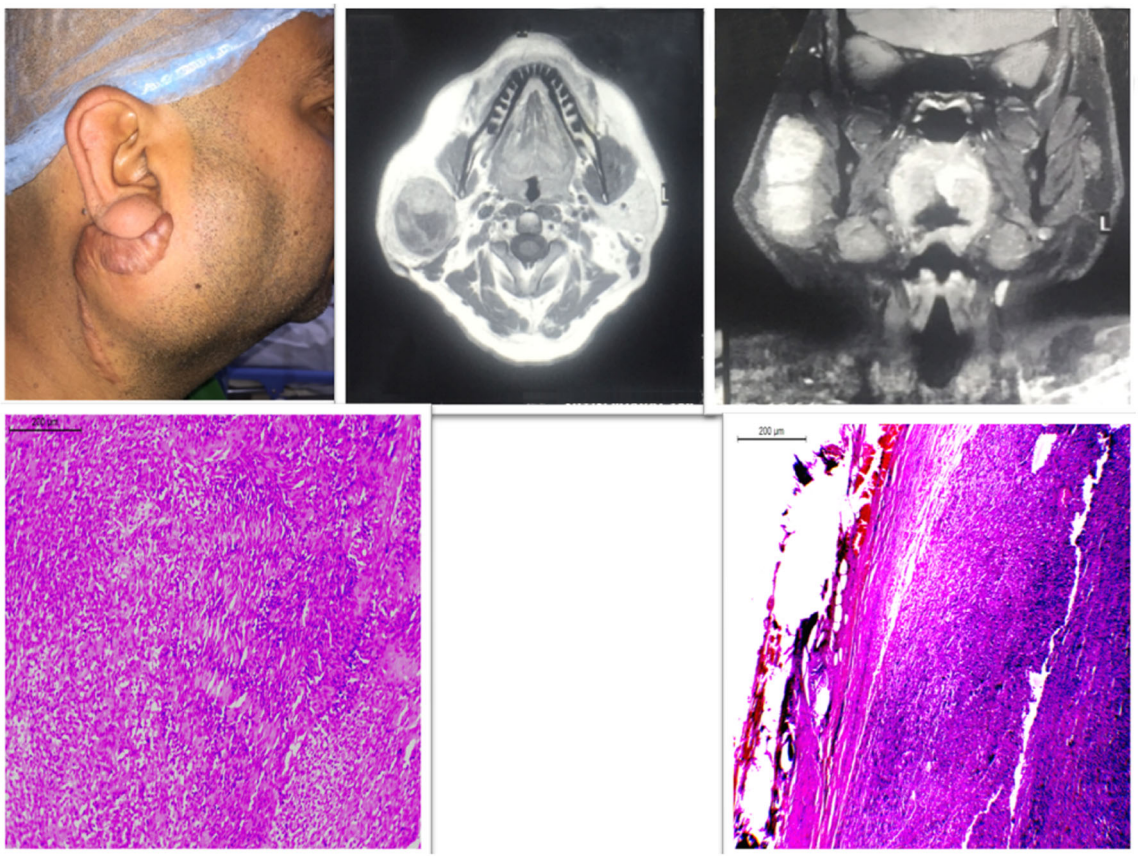

Fig. 3 **a** Preoperative clinical photos of case 3 showing around 7 × 8-cm swelling involving the right parotid region with a hypertrophic scar over the previous surgical incision line. **b** Contrast-enhanced MRI of case 3 showing both axial and coronal cuts showing heterogeneous mass lesion in the right parotid area with post-contrast heterogeneous enhancement. **c**, **d** Hematoxylin and eosin–stained section (magnification × 40) shows a well-encapsulated mass composed of densely packed spindle cells with characteristic nuclear palisading

Discussion

Facial nerve schwannomas have been well documented in the medical literature since its first description by Schmidt in 1930. Most of them arise from the intra-cranial course of the facial nerve. Intraparotid facial nerve schwannomas are a rarer entity and account 0.5–1.5% of all facial nerve schwannomas [6]. One study quotes incidence of less than 70 cases of parotid involvement in around 400 documented cases of facial nerve schwannomas over 38 years [7].

The duration of symptoms in our series was between 3 months and 4 years (average 1.6 years). This refutes the notion put forward by Caughey that the prolonged duration of symptom would best indicate a schwannoma [8]. Due to the rarity of this entity, the gender preponderance has not been established yet, albeit the available literature all point towards an equal incidence amongst both sexes [6, 9]. In our series, three patients were female and one was male.

Intraparotid facial nerve schwannomas are largely asymptomatic, and it is only when the swelling is noticeable to the patient that he/she seeks medical attention. In all our cases, the presenting symptom was that of a painless, slow-growing swelling in the parotid region. In case 2, there was occasional pain on presentation, but on further speculation, it was established that the pain developed over the swelling after she underwent fine needle aspiration cytology (FNAC) for the same. This is in agreement with other studies with larger samples such as that quoted by Marchioni D (91.1%). Hence, it is a commonplace to confuse it with other benign tumors of the parotid such as pleomorphic adenoma. Intraparotid facial nerve schwannoma presenting with facial paresis is infrequent, and reported incidence is 20–27% in various studies [6, 10, 11]. However, when it presents with pain and facial paresis, it may be confused with malignant parotid tumor. Low incidence of facial palsy at presentation is due to the ability of the parotid gland to accommodate tumor expansion and propensity of tumor to grow eccentrically and push nerve fibers away [10]. In our series, one of the patients developed facial palsy at presentation which was due to prior surgical intervention.

FNAC can accurately distinguish benign and malignant parotid lesion, and its sensitivity is 90%. However it is not reliable to accurately clinch the diagnosis of facial nerve schwannomas in all the cases [6, 9]. In our series, FNAC

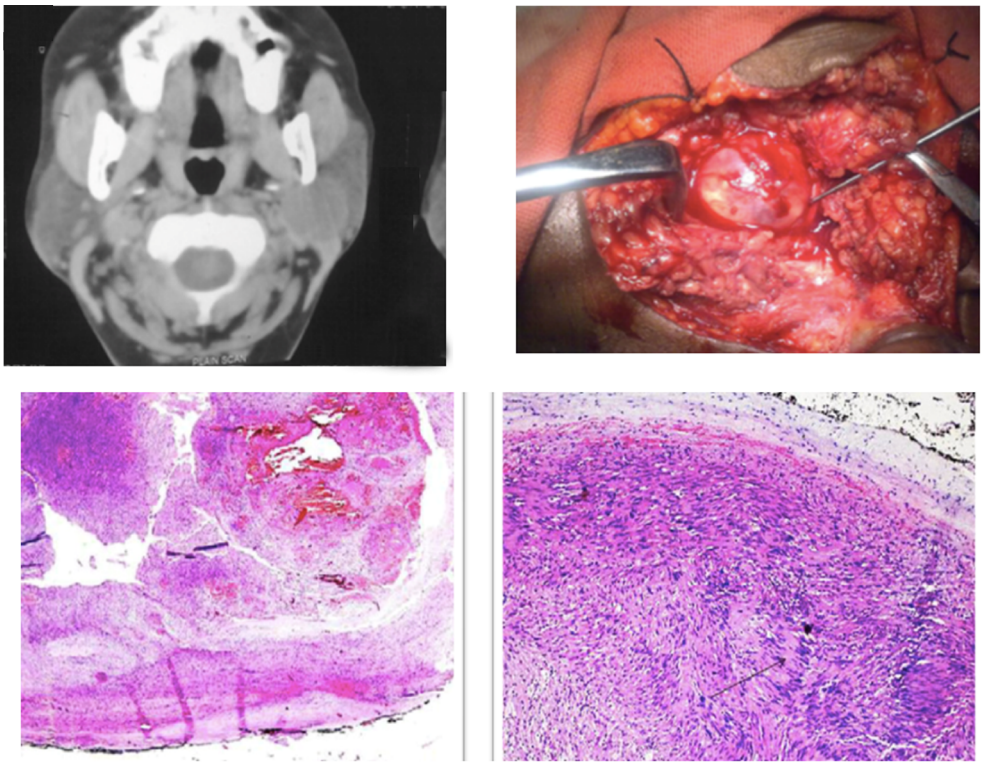

Fig. 4 **a** Contrast-enhanced computed tomography scan (axial cuts): dumbbell-shaped homogenous enhancing tumor involving deep lobe of the parotid gland. **b** Intraoperative photograph showing 4 × 3-cm cystic mass with solid component was found involving deep lobe of the parotid gland. **c**, **d** Fibrous capsule with the underlying tumor exhibiting cellular and hypocellular areas. The cellular area shows nuclear palisading around the fibrillar cellular processes, so called Verocay bodies (arrow) and myxoid hypocellular area

was suggestive of schwannoma in only one out of four cases. Cytological features are quite characteristic and include fragments of spindle-shaped neoplastic cells forming Verocay bodies [12]. A preoperative diagnosis of a facial nerve schwannoma reassures the surgeon and helps in counseling the patient with regard to the facial nerve and helps the patient in decision making.

CT scan is not the ideal imaging modality for evaluation of parotid masses. The best investigative modality of choice is the gadolinium-enhanced MRI [13]. Martin et al. [14] described in their series the lesions in MRI to be well defined with signal intensities isointense to the muscle on T1-weighted images and hyperintense to the muscle on T2-weighted images. Three of the five facial nerve schwannomas in their study exhibited a "target sign" characterized by increased peripheral signal intensity and decreased central signal intensity on T2-weighted images.

Surgical excision is the treatment of choice. Two important characteristics of facial nerve schwannoma are: some tumors can be dissected facial nerve preserving its nerve integrity while in some cases the tumor may be tightly adherent to the nerve, requiring the surgeon to remove the tumor along with a variable length of facial nerve tract, followed by reconstruction [7]. Various options include observation, partial resection of tumor with preservation of nerve function, and resection of tumor with sacrifice of the facial nerve and cable grafting [6].

There remains a controversy regarding the management of an intraparotid facial nerve schwannoma. Alicandri-Ciufelli et al. suggested that the localization and adherences of intraparotid facial nerve schwannomas, the relationship with the facial nerve, and the preoperative facial nerve function are important factors in the decision-making process in order to optimize the functional outcomes [15]. If a "wait and watch" approach is considered, patients should undergo regular follow-up with imaging and close monitoring of the facial nerve function. However, if the size of the tumor is considerable, it may compromise the facial nerve function; therefore, it is rational to surgically resect such masses.

Two important characteristics of intraparotid facial nerve schwannoma are some tumors can be dissected from the facial nerve while preserving nerve integrity and some of the tumors may be tightly adherent to the nerve, requiring sacrifice of the nerve [16]. Marchioni et al. [6] classified peripheral facial nerve schwannomas into four types [6]. Type A comprised of tumors which could be resected without any facial nerve injury. Types B, C, and D required sacrifice of the facial nerve and reconstructive measures thereafter. Based on the classification system proposed by Marchioni et al., three of our cases belonged to type A tumors as evident by the post-operative intactness of the facial nerve in all three patients, and one belonged to Type B tumors (Table 1).

Conclusion

Because intraparotid facial nerve schwannomas are benign, indolent growing tumors with exceptionally low malignant

Table 1 Tabulated summary of the cases

S. no.	Age/sex	Presentation	Duration of symptoms	Pre-operative facial nerve grade (HBG)	Pre-operative work-up FNAC	Surgical procedure	Origin of tumor	Post-operative facial nerve grade (HBG)	Size of tumor
1	34/f	Painless parotid swelling	1 year	I	Conclusive	Superficial parotidectomy	Upper buccal branch	I	2 × 2 cm
2	19/f	-do-	1.5 years	I	Inconclusive	Superficial parotidectomy	Upper buccal branch	I	2.5 × 2 cm
3	37/m	Facial deviation, parotid swelling	4 years	III	Inconclusive	Tumor resection with facial nerve sacrifice and cable grafting	Not identified	IV	7 × 5 × 3 cm
4	45/m	Painless parotid swelling	3 months	I	Inconclusive	Total conservative parotidectomy	Lower buccal branch	I	3 × 4 cm

potential, pre-operative FNAC is usually in conclusive. Priority should be placed on preserving facial nerve function and facial cosmesis.

References

1. Pessey JJ, Serrano E, Percodani J. (1994) Nervous tumors of the neck [in French]. Editions Techniques-Encycl Med Chir(Paris-France). Otorhinolaryngologie 20–875-C-10
2. O'Keefe JJ (1949) Neurinoma of the facial nerve in the parotid gland. Ann Otol Rhinol Laryngol 58:220–225
3. Verocay J (1910) Information on the neurofibrome [in German]. Beitr Path Anat 48:1–68
4. Balle VH, Greisen O (1984) Neurilemomas of the facial nerve presenting as parotid tumors. Ann Otol Rhinol Laryngol 93:70–72
5. Forton GE, Moeneclaey LL, Offeciers FE (1994) Facial nerve neuroma: report of two cases including histological and radiological imaging studies. Eur Arch Otorhinolaryngol 251:17–22
6. Marchioni D, Alicandri-Ciufelli M, Presutti L (2007) Intraparotid facial nerve schwannoma: literature review and classification proposal. J Laryngol Otol 121(8):707–712
7. Elahi MM, Audet N, Rochan L et al (1995) Intraparotid facial nerve schwannoma. J Otolaryngol 24:364–367
8. Caughey RJ, May M, Schaitkin BM (2004) Intraparotid facial nerve schwannoma: diagnosis and management. Otolaryngol Head Neck Surg 130(5):586–592
9. Gross BC, Carlson ML, Moore EJ, Driscoll CL, Olsen KD (2012) The intraparotid facial nerve schwannoma: a diagnostic and management conundrum. Am J Otolaryngol–Head and Neck Med Surg 33:497–504
10. O'Donoghue GM, Brackmann DE, House JW, Jackler RK (1989) Neuromas of the facial nerve. Am J Otolaryngol 10(1):49–54
11. Guzzo M, Ferraro L, Ibba T, Quattrone P, Bianchi R, Rezzonico S, Scaramellini G (2009) Schwannoma in the parotid gland. Experience at our institute and review of literature. Tumori 95: 846–851
12. Salemis NS, Karameris A, Gourgiotis S, Stavrinou P, Nazos K, Vlastarakos P, Tsiambas E, Tsohataridis E (2008) Large intraparotid facial nerve schwannoma: case report and review of the literature. Int J Oral Maxillofac Surg 37:679–681
13. Thompson AL, Aviv RI, Chen JM, Nedzelski JM, Yuen HW, Fox AJ, Bharatha A, Bartlett ES, Symons SP (2009) Magnetic resonance imaging of facial nerve schwannoma. Laryngoscope 119: 2428–2436
14. Martin N, Sterkers O, Mompoint D, Nahum H (1992) Facial nerve neuromas: MR imaging—report of four cases. Neuroradiology 34: 62–67
15. Alicandri-Ciufelli M, Marchioni D, Mattioli F, Trani M, Presutti L (2009) Critical literature review on the management of intraparotid facial nerve schwannoma and proposed decision-making algorithm. Eur Arch Otorhinolaryngol 266(4):475–479
16. Fyrmpas G, Konstantinidis I, Hatzibougias D, Vital V, Constantinidis J (2008) Intraparotid facial nerve schwannoma: management options. Eur Arch Otorhinolayngol 265:699–703