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Case Series

Case Series-Intraparotid Facial Nerve Schwannoma: Our Experience in 5 Cases

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Abstract

Intraparotid Facial Nerve Schwannoma is an exceedingly rare tumor of head and neck region. It forms about 0.5% - 1.2% of all parotid tumors. It is neuroectodermal in origin. Facial nerve schwannomas are benign, encapsulated, slow growing tumours that arise from nerve sheath. Most common presenting complaint is unilateral facial swelling though facial paralysis is seen in few of the patients. Fine needle aspiration cytology (FNAC) and radiology play a key role in preoperative diagnosis. Treatment of choice for facial nerve schwannomas is complete surgical excision. Here we are reporting 5 cases of facial nerve schwannomas which lies within the parotid gland region. All the patients presented with unilateral facial swelling from the year 2017 to 2020. Clinical features, diagnosis and management of these cases are discussed here. We recommend that before going for excision, patient should be made aware of the possible functional facial deficit and as per the patient's priority, decision of excision should be made. After complete excision, recurrence rate was found to be extremely low. Usually, post-operative functional deficit recovers almost completely with physiotherapy.

Keywords: Facial Nerve; Neuroectoderm; Intraparotid Schwannoma; Parotidectomy; Facial Paralysis

Introduction

Schwannoma is a rare type of head and neck tumor. It is a benign nerve sheath tumor which can arise from any nerve covered with Schwann cell sheath. About 25 to 45% of all schwannomas originate from neural structures of head and neck region [1]. Facial nerve schwannomas are rare, benign, encapsulated, slow growing tumours that arise from facial nerve sheath. They are neuroectodermal in origin [2]. Most of facial nerve schwannomas are intratemporal, whereas 9% are located extracranially [3]. Facial nerve schwannomas account for 0.5% - 1.2% of all parotid gland tumours

[4]. Facial nerve dysfunction including paresis and paralysis may be present in only 20% of all patients, though tumour originates from facial nerve [5]. Radiological investigations mainly CT scan and MRI along with FNAC play a key role in establishing a preoperative diagnosis. Intra-operative identification of facial nerve and tumour delineation from the offending branch is the key to a good successful outcome. Here we are reporting 5 cases of intraparotid facial nerve schwannomas who presented with facial swellings with normal facial nerve function. Clinical features, diagnosis and management are discussed with review of literature. The purpose of this study is to demonstrate the variation in presentation and outcomes of the management.

Case 1

A 29 years old female presented to ENT OPD with chief complaints of swelling in front and below right ear for 3 years. The swelling was insidious in onset, gradually increasing in size, persistent, non-discharging, not relieved with medications and not associated with facial weakness. The examination of face revealed a single, spherical swelling of approximately 6 x 4 cm in right parotid area which was extending superiorly just in front of tragus, inferiorly till the level of angle of mandible, anteriorly about 3 cm anterior to tragus and posteriorly about 1cm behind pinna. Surface of swelling was smooth with regular margins.

Figure 1A-1D: A. Preoperative clinical image of case 1 showing 6 x 4 cm swelling in right infra-auricular region; B. Contrast enhanced CT of face and neck of the case 1 (axial view) depicting lesion; C. Magnetic resonance image of neck of the case 1 (axial view) depicting lesion; D. Intraoperative picture showing tumour mass was of approximately 6.5 x 4.5 cm size arising from zygomatic branch of facial nerve after flap elevation.

Palpation suggested normal local temperature, with a non-tender, firm, non-fluctuant, bimanually non-palpable swelling and normal facial nerve functions. CECT Face and Neck was suggestive of peripheral enhancing necrotic mass lesion 6 x 5.4 cm with thick shaggy margins involving deep part of right parotid region. MRI Neck was suggestive of a complex ovoid shaped 6.4 x 3.3 x 4.4 cm mass with cystic degeneration in right masseteric space extending superficial to parotid space. Fine needle aspiration cytology (FNAC) revealed a benign spindle cell tumour with closest to schwannoma. Patient was operated under general anaesthesia for right parotid (deep lobe) schwannoma excision after taking consent for facial nerve injury. Intra-operative findings revealed stretching of facial nerve trunk and its branches by tumour. Tumour mass was of approximately 6.5 x 4.5 cm size arising from zygomatic branch of facial nerve extending from deep lobe of parotid up to parapharyngeal space. Tumour had solid and cystic components.

Grade IV facial nerve function paresis was present postoperatively. Patient was started on facial physiotherapy, and there was complete recovery of facial nerve function after 6 months. The Histopathology report revealed facial nerve sheath schwannoma.

Figure 2: Histopathological examination suggestive of schwannoma (red arrows showing verocay bodies).

Case 2

A 10 years old male presented to ENT OPD with chief complaints of swelling in front of left ear for 3.5 years. Patient underwent left superficial parotidectomy in 2016 after 6 months of development of swelling. Patient remain asymptomatic for 6 months. Patient again presented swelling in front of left ear for 2.5 years. On examination of face, there is a single, spherical swelling of approximately

6 x 4 cm present on left parotid area. Scar mark of approximately 0.5 x 3 cm present, 1cm in front of tragus vertically downwards. Facial nerve function was normal.

Figure 3A-3C: A. Preoperative clinical image of case 2 showing 6 x 4 cm swelling in right infra-auricular region; B: Magnetic resonance image of neck of the case 2 (axial view) depicting lesion; C: Intra-op findings depicting a single 7 x 5 cm firm, well-encapsulated tumour mass seems to be arising from nerve sheath of left zygomatic branch.

FNAC was suggesting a benign nerve sheath tumour. MRI Neck was suggestive of a well-defined lobulated homogenous intense enhancing left parotid mass lesion 24.3 x 21.6 x 23.3 mm with string sign. Patient underwent left parotid recurrent benign nerve sheath tumour excision. Intra-op findings revealed single 7 x 5 cm firm, well-encapsulated tumour mass seems to be arising from nerve sheath of left zygomatic branch which was sacrificed. Left facial nerve trunk and its branches were pushed inferiorly by tumour. Divisions and branches of facial nerve were going over the tumour. Tumour was delineated- projection going up to left zygoma which was thinned out and posteriorly going up to infratemporal region. Periosteum was drilled and zygoma was drilled at the point of attachment of tumour. Grade 4 facial weakness present on left side in postoperative period, which improved up to grade 2 after 3 months of physiotherapy. The histopathology report revealed schwannoma.

Case 3

A 13 years old male presented to ENT OPD with chief complaints of swelling in front of right ear for 4 years. Facial nerve function was normal.

Figure 4A-4D: A. Preoperative clinical image of case 3 showing 3 x 2 cm swelling in right infra-auricular region; B and C: Magnetic resonance image of neck of the case 3 (axial view) depicting lesion pre and post contrast respectively; D: Intra-op findings showing a single 2.5 x 1.5 cm, firm and well-encapsulated tumour mass separated from twigs of facial nerve upper trunk branches.

FNAC was suggesting a benign nerve sheath tumour. MRI Neck was suggestive of vivid peripheral enhancement within the lesion ~(2.5 x 1.7cm) on post contrast images. Patient underwent right parotid benign nerve sheath tumour excision. Postoperatively, facial nerve function was normal. The Histopathology report revealed schwannoma.

Case 4

A 26 years old female presented to ENT OPD with chief complaints of swelling left side of face below pinna for 4 years, which was not associated with facial weakness.

Figure 5A-5C: A. Preoperative clinical image of case 4 showing 4 x 3 cm swelling in right infra-auricular region; B: Contrast enhanced CT of face and neck of the case 4 (axial view) depicting lesion; C: Intra op findings showing a single well-defined cystic lesion in left parotid gland with enhancing walls, approximately 3.2 x 3 cm in superficial part of parotid.

FNAC was suggestive of benign salivary aspirate. CECT (Base of Skull to T4 level) was suggestive of a single well-defined cystic lesion in left parotid gland with enhancing walls, approximately 3.2 x 3 cm in superficial part of parotid. Patient underwent left superficial parotidectomy under general anaesthesia. Facial nerve function was preserved postoperatively. The Histopathology report revealed schwannoma.

Case Report 5

A 52 years old man presented with chief complaints of swelling right side of face for 3 years. Facial nerve function was normal.

Figure 6A-6C: Preoperative clinical image of case 5 showing 3 x 3 cm swelling in right infra-auricular region; B: Contrast enhanced CT of face and neck of the case 5 (axial view) depicting lesion; C: Magnetic resonance image of neck of the case 5 (axial view) depicting lesion.

FNAC was suggestive of benign salivary aspirate. CECT (Base of Skull to T4 level) was suggestive of a single well-defined cystic lesion in right parotid gland with enhancing walls, approximately 3.1 x 3 cm in superficial part of parotid. MRI face was suggestive of a well- defined round to oval lesion approximately 3.1 x 3 x 2 cm in size seen in superficial lobe of right parotid gland. It appears iso to hypointense on T1 and heterogeneously hyperintense on T2/STIR images. Patient underwent right superficial parotidectomy under general anaesthesia. Facial nerve function was preserved postoperatively. The Histopathology report revealed schwannoma.

Discussion

Facial nerve schwannomas (FNS) also termed as neurilemmomas were 1st reported by Virchow in 1908 [6]. They can originate from any site along the nerve, from glial-Schwann cell transition site at the cerebello-pontine angle to the terminal branches of facial nerve [7]. Caughey., *et al*. reported a retrospective study of 3,722 patients diagnosed with schwannoma, in which 29 cases were associated with facial nerve and 8 cases involving intraparotid region of facial nerve [8]. Duration of symptoms varied from 3 to 4 years in our study. In a study reported by Caughey., *et al*. age distribution was from 7 to 78 years with average age at diagnosis was 44 years [8]. Our study comprised patients of age ranging from 10 to 52 years with average age at diagnosis was 26 years. As the intraparotid FNS is a rare entity, gender preponderance is still to be established. In our study, it was 3:2 and in congruence with available literature [9,10]. Two of our patients were of pediatric age group, which is a rare occurrence. The difference in average age at diagnosis is mainly due to small sample size. Intraparotid FNS often remain asymptomatic for longer duration till swelling becomes apparent, then only the patient seeks medical advice. When it is associated with pain and facial paraesis, it may create diagnostic confusion with parotid malignancy. Rarity of facial palsy in intraparotid FNS is due to propensity of tumor for eccentric growth, pushing nerve fibers away [11]. FNAC is not reliable in accurately diagnosing the intraparotid FNS [9,10]. In our study, FNAC was inconclusive in one case. CT scan, although not ideal in this case, shows smooth circumscribed lesion in parotid. Gadolinium enhanced MRI being preferred modality shows 'Target sign' characterized by hyperintensity in periphery while hypointensity in the centre on T2 weighted images and 'String sign' due to mass being situated just below the stylomastoid foramen with breaking into foramen [12]. String sign was seen in one of the cases in our study. On macroscopic examination, schwannoma appears as a well-defined cystic mass with smooth surface and variegated colours. Histopathologically, arrangements of Antoni A with Verocay bodies and arrangements of Antoni B. Immunostaining for S-100 for neural origin of tumour and smooth muscle actin to rule out leiomyoma [13].

Intraparotid FNS, hence, becomes difficult to diagnose based on clinical findings, FNAC and radiological examinations. Thus, final diagnosis is only possible after surgical excision and histopathological evaluation of microscopic findings. Alicandri-Ciufelli., *et al*. have implied that the location, adherence of intraparotid FNS and preoperative facial nerve function status are the important factors in determining the functional outcomes of the surgery [14].

Marchioni., *et al*. described IPFNS into 4 types which aid clinician in evaluating therapeutic and prognostic factors [15]. Two of our cases belonged to type A with normal facial nerve function postoperatively. While two cases belonged to types B with grade 4 postoperative facial paraesis in one and normal facial nerve function in other. There was one case of type C with grade 4 postoperative facial palsy. Surgeon must weigh the detrimental effect of postoperative facial palsy after excision against the risk of leaving schwannoma as it is. However, as the final diagnosis can only be made after excision, it is imperative to go for in depth discussion with the patient about possible facial palsy and its management. Also, recurrence rate is usually low after complete excision. At the same time, patient should also be counselled if wait and watch approach is considered in smaller lesions, then regular follow up with imaging and facial nerve function monitoring is mandatory.

Conclusion

Intraparotid FNS are rare benign tumors with slow indolent course. Preoperative FNAC and radiological diagnosis are often present us difficulty in making definitive diagnosis. We recommend that before going for excision, patient should be made aware of the possible functional facial deficit and as per the patient's priority, decision of excision should be made. However, after complete excision, recurrence rate was found to be extremely low. Facial nerve function usually returns to grade 1 or 2 in cases with postoperative weakness of grade 3 or 4 with vigorous physiotherapy and oral steroids.

Declaration of Conflict of Interest

None.

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