Preauricular Intraparotid Schwannoma: A Rare Presentation with Literature Review

Abstract

Schwannoma is a benign tumor rarely found in the intraparotid facial nerve region. It clinically presents as a slow-growing, asymptomatic mass. Due to its rare presentation, preoperative diagnosis is often unclear before surgical removal and histopathological examination. Imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) play an important role in suggesting the nature of mass and narrowing down the differentials. The CT scan offers the advantage to detect the relationship of the facial nerve and osseous changes within the bone, however MRI shows a mass relative to brain type of tissue. We report a rare case of intraparotid facial nerve schwannoma in a 17-year-old female who had sustained swelling in her left preauricular area for 5 years. Ultrasonography and CT findings revealed the impression of pleomorphic adenoma. However, MRI and histopathological findings were characteristic of schwannoma. Thus, this article provides an insight into a rare presentation of schwannoma with literature review.

Keywords: *Schwann cells, soft-tissue tumors, schwannoma*

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Introduction

The parotid gland is a rare location for schwannoma which arises from the Schwann cells of the nerve sheath. The intraparotid facial nerve schwannoma accounts for 9% of schwannoma arising from the facial nerve and its incidence is 0.2%–1.5% of all schwannomas.[1] Clinically, it presents as an asymptomatic slow-growing parotid mass without any distinctive pathognomonic findings mimicking with most common benign tumors such as pleomorphic adenoma.[2] The preoperative diagnosis of schwannoma is a diagnostic dilemma as fine-needle aspiration cytology (FNAC) is inaccurate mostly, and it is difficult to distinguish it using imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) from pleomorphic adenoma.[3] We report a similar rare case of schwannoma diagnosed by histopathology.

Case Report

A 17-year-old female patient presented in the department of oral medicine and radiology with the chief complaint of swelling in the upper left front region of the ear for 5 years. The swelling developed spontaneously which gradually increased in size and attained the present size. It was not associated with pain, fever, loss of weight, loss of appetite, or related to meals. Past medical history and dental history were uneventful, and the patient was apparently healthy on general physical examination.

On extraoral examination, a solitary ovoid swelling was present in the left preauricular region. The swelling was lobulated, same as the color of the facial skin measuring about 2 cm × 1.5 cm in size. The swelling was nonpulsatile with a smooth surface and well-defined edges. On palpation, the swelling was firm, lobulated, nontender with well-defined margins, and was not attached to underlying structures [Figure 1]. The swelling was pinchable, nonfluctuant, and noncompressible with no signs of inflammation and regional lymphadenopathy. No abnormality was detected on the facial nerve and temporomandibular joint examination.

On intraoral examination, there was no evidence of any swelling, with normal opening of Stensen's duct. On the basis of history and clinical examination, a provisional diagnosis of benign neoplastic lesion (pleomorphic adenoma of the left side) was proposed. Sialadenosis, fibroma, preauricular

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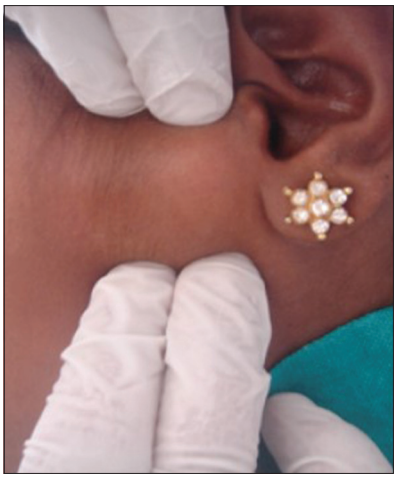

Figure 1: Examination of swelling

lymphadenopathy, lipoma, leiomyoma, schwannoma, and Warthin's tumor were given as differential diagnosis [Table 1]. Orthopantomograph and complete hemogram were normal. FNAC was nondiagnostic as it revealed cohesive sheets of epithelial cells with scattered lymphocytes.

On ultrasound, tumor mass was delineated as hypoechoic masses with multiple internal cystic areas in the soft tissue overlying the left ramus with a sharply delineated contour and posterior echo enhancement suggestive of benign neoplastic lesion [Figure 2a]. CT scan was carried out which revealed an ill-defined multiloculated cystic lesion with internal particulates are noted in the soft tissue overlying the ramus of the left mandible approximating the facial nerve. The lesion measured approximately 4.04 cm × 1.87 cm and no involvement of underlying

Table 1: Clinical differential diagnosis of preauricular swelling

Differential Diagnosis	Favourable features	Unfavourable features
Pleomorphic adenoma	Slow growing, painless swelling	4-6 decade of life span
	Firm	Local discomfort
	Long duration history	Difficulty in mastication and talking
	Female predilection	
	Mostly present in Front of ear or ascending ramus	
	Mostly do not show fixation to deep tissues or underlying skin	
Sialadenosis	Slow growing swelling	Usually bilateral
	Painless	Older age group affected
		Decreased salivary secretions
		Associated systemic history of diabetes
Fibroma	Most common tumor	Extraoral presentation is rare.
	Long duration history	Most common location is the buccal mucosa along the bite line
	Painless	
	Gradual progression	
	Firm consistency	
Preauricular lymphadenopathy	Swelling in front of ear	Usually painful
	Common presentation	Redness on affected side
		History of fever and Ear infection
Lipoma	Sessile	Very rarely it occurs in head and neck region.
	Smooth surfaced lobulated swelling	Most commonly it occurs in trunk and extremeties
	Long duration history	Soft in consistency
	Painless	Positive slip sign
	Gradual progression	Buccal mucosa and buccal vestibule are the most common sites
Lieomyoma	Slow growing	Very rare finding in the oral cavity
	Firm, mucosal lesion	Usually pedunculated
	Asymptomatic	
Schwannoma	Slow growing lesion	Rare entity
	Painless	Facial twitching
	Long duration history	Facial nerve paralysis
	Single circumscribed nodule	Hearing loss & tinnitus
Warthin tumour	Slow growing painless swelling	Commonly occur beneath ear lobe near tail of gland
	Firm non tender mass	Mostly bilateral and more common in males.

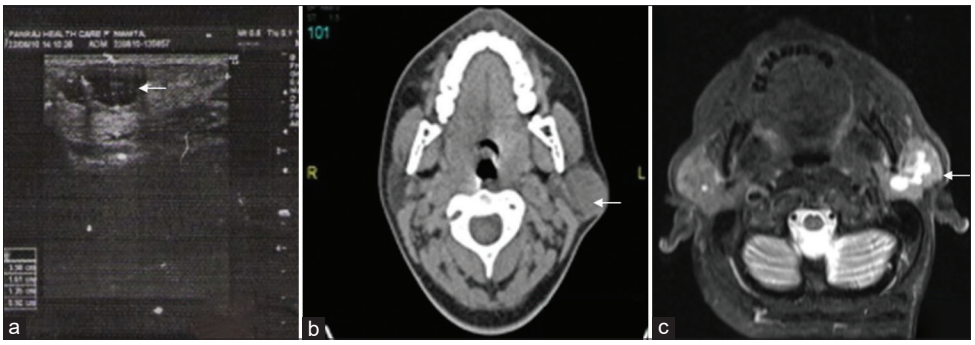

Figure 2: Radiographic imaging of lesion (a) Ultrasound imaging revealed markedly hypoechoic mass (b) computed tomography showed multiloculated cystic lesion approximating the left facial nerve (c) Axial T1 magnetic resonance imaging presented isodense dumbbell lesion approximating the left facial nerve

Table 2: Diffe Differential Diagnosis on Basis of MRI rential Diagnosis on Basis of MRI

Differential Diagnosis	Favourable Features	Unfavourable features
Schwannoma	Isotense T1 MRI signal	String sign absent
	Dumbbell shaped	Target sign absent
	Approximating Facial nerve	
Benign Salivary tumour	Well encapsulated	High intensity T2W1 Signal suggestive of myxoid appearance not present
	Intra-tumoural cystic change	No Peripheral enhancement of fibrous capsule
Neurofibromatosis	Well defined margins	Homogenous T1 Signal not present
	Peripheral enhancement absent	Heterogenous T2 Signal not present
		Intra-tumoral cystic change
Nerve Sheath myxoma	Well demarcated lesion	Absence of heterogenous high intensity when compared with muscle tissue on T1W image.
	Absence of Specific signs	Deep seated lesion
		Peripheral enhancement on T2W images absent

Table 3: Swellings in Pre-auricular Region

Etiology	Disease	Prevalence
Due to Parotid gland	Parotitis	27-30%
	Sailadenitis	10-24%
	Sialadenosis	26-30%
	Pleomorphic adenoma	60-70%
	Warthin's tumour	8-10%
Due to Lymph node	Preauricular lymphadenopathy	20-52%
	Lymphoepithelial cyst	3-5%
Skin	Sebaceous cyst	3-5%
	Dermoid cyst	1-3%
Muscle	Leiomyoma	1-2%
Connective tissue	Lipoma	0.6-4.4%
	Fibroma	2-4%
Nerve	Schwannoma	0.2-1.5%

ramus. A benign tumor mass was evident on the CT report [Figure 2b]. Since the lesion was approximating facial nerve for further characterization, MRI was advised. In axial T1MRI, a homogenous, isodense dumbbell lesion was seen approximating the left facial nerve relative to brain tissue [Figure 2c]. The radiographic Differential Diagnosis (d/d) included were benign cystic tumor, nerve sheath myxoma, neurofibromatosis, and schwannoma [Table 2].

Excisional biopsy was planned, and a complete encapsulated lesion was excised. The facial nerve was preserved; sutures were placed and the sample was sent for histopathological examination. The histopathological report revealed tumor parenchyma consisted of hypercellular spindle cells. These spindle cells showed areas of nuclear and cytoplasmic pleomorphism and nuclear palisading resembling Antoni B-type tissue consisting of streaming fascicles of spindle-shaped cells confirming the diagnosis of schwannoma [Figure 3]. Postoperative follow-up could not be carried out as the patient did not turn up after suture removal.

Discussion

Verocay first described the lesion in 1910 and named it neurinoma. In 1935, stout coined the term neurilemmoma.[4] Intraparotid schwannomas are solitary, painless, slow-growing masses where the function of the facial nerve is generally unaffected.[5] Among schwannomas, the preauricular region is rare and poses a difficulty in preoperative diagnosis since it is the most common site for the salivary gland neoplasms[6] [Table 3]. Gallo *et al*.

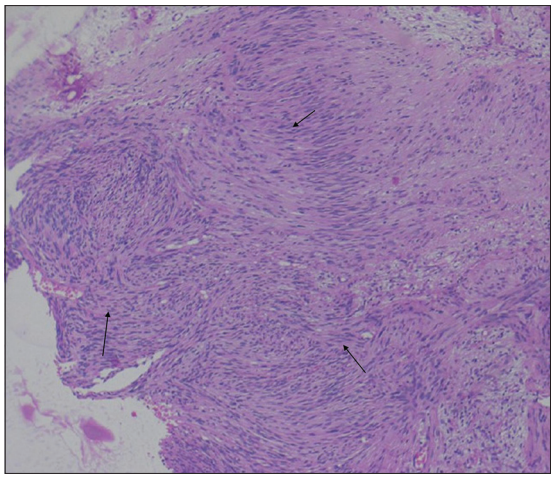

Figure 3: Histopathological examination revealed streaming fascicles of spindle cells with nuclear palisading resembling Antoni B-type tissue

reported 157 cases of schwannoma involving head and neck region and found that 45.2% occur on the tongue, 13.3% in the cheek, and 10% in the parotid region.[7] Schwannoma can occur at any age with no significant gender bias.

FNAC is still debatable as it is not diagnostic always. A7 Although some authors advocated the role of cytology as schwannoma may reveal a sheet of spindle cells exhibiting nuclear palisading within a hypocellular stromal background.[8]

Various imaging modalities are available to delineate the tumor mass such as ultrasonography, CT, and MRI. Sonographically, it appears as a marked hypoechoic well-circumscribed mass with no demonstrable internal vascularity representing benign neoplasms.[2] On a CT scan, the appearance of a schwannoma is generally characteristic of well-circumscribed encapsulated mass.[3] CT offers advantage to delineate the osseous structures and calcifications, however MRI is the best investigative modality of choice because it demonstrates isodense signal on T1W1 image, hyperdense on T2W1 suggestive of neurogenic tissue with any of the two signs, i.e., string sign (mass with beak-like protrusion near stylomastoid foramen) and target sign (hyperdense signal at periphery of tumor).[3,9] In the present case, FNAC was not conclusive, however ultrasound and CT report gave the perception of pleomorphic adenoma, but on MRI, neural tumor was evident, although specific signs could not be observed.

Treatment of intraparotid schwannoma is surgical excision. Marchioni *et al*. suggested a classification of intraparotid schwannoma according to its position in the facial nerve pathway. In this classification, there are four types of tumor resection: Type A (facial nerve is preserved), Type B (partial sacrifice of the facial nerve), Type C (the main trunk of the facial nerve is sacrificed), and Type D (facial nerve trunk and its main divisions are sacrificed).[10] Our present case was considered as Type A.

A definitive diagnosis should be based on the histological findings. Histologically, the tumor is characterized by a stream of elongated spindle cells often arranged in a palisaded pattern. Areas consisting of thick concentration of cells are Antoni A and loosely irregularly placed cells are Antoni B cells.[4]

Conclusion

Intraparotid schwannomas are a rare entity, but it should not be discarded when observing a submucosal soft-tissue swelling of the parotid gland region, as in the present case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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