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RESEARCH ARTICLE

SCHWANNOMA IN DISGUISE, THE NASOLABIAL STORY: REPORT OF A RARE CASE

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ABSTRACT

Schwannomas are rare kind of benign neural tumors. They arise from Schwann cells of the neural sheath of peripheral, cranial, or autonomic nerves. Of twenty five percent of all Schwannomas located in the head and neck, only 1% have intraoral presentation. Due to its abysmal rate of occurrence, it makes a less likely diagnosis in case of oropharyngeal masses, leading to delay in identification and treatment. The most definitive way to diagnose this tumor is by its histopathologic and immunohistochemical examination. We report an interesting case of a patient with a mass located in the nasolabial region, which initially had presentations similar to a nasolabial cyst, but finally turned out to be a Schwannoma. There has been no sign of recurrence almost two years after surgery. The importance of considering Schwannoma as a differential diagnosis can't be stressed enough since it is increasing in incidence of late.

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INTRODUCTION

Schwannoma, earlier known as neurilemoma or neurinoma, is a benignnerve sheath tumour of neuroectodermal origin. There is difference of opinion about their origin wherein some authors believe that they, arise from Schwann cells that comprise the myelin sheaths surrounding peripheral nerve or supporting fibroblast of peripheral, cranial or autonomic nerves. And then there are few others who think they originate directly from the neural tube or from the neural crest. (Parhar et al., 2014)Approximately 37–45 % of Schwannomas occur in the head and neck region, but only 1% shows an intraoral origin. These lesions commonly arise from the roots of cervical and cranial nerves in the parapharyngeal space, with a majority of them originating from the Vagus.

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Most common intraoral sites are the tongue, followed by palate, the floor of the mouth, buccal mucosa, gingiva, lips, and the vestibular mucosa. Although they may arise at any age, the peak incidence is between the third and sixth decades, and there is no gender predilection (Jadwani et al., 2012). Schwannomas remain asymptomatic unless they attain appreciable size. These tumors are usually solitary lesions but in unusual instance are multiple, or occur in the setting of von Recklinghausen's neurofibromatosis. In contrast to multiple neurofibromatosis, Schwannomas almost never undergo malignant transformation. Treatment of choice is surgical excision of the tumor. Schwannomas do not show recurrence if completely excised. Malignant transformation is rare. (Jadwani et al., 2012) An intraoral Schwannoma is a smooth, sub-mucosal swelling bearing an uncanny resemblance to mucocele, fibro-epithelial polyp, fibroma, lipoma ora benign salivary gland tumor.

Here, we report a case of a swelling in the right nasolabial region which had similar presentations of a nasolabial cyst, but turned out to be a Schwannoma on histopathologic examination.

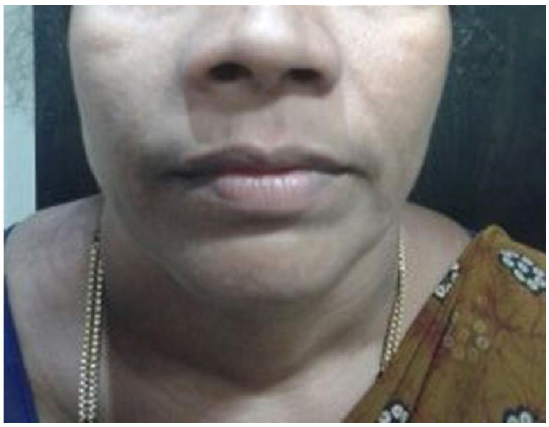

Figure 1. Front view of the pt. showing swelling below the rt. ala of the nose, obliterating the naso-labial fold

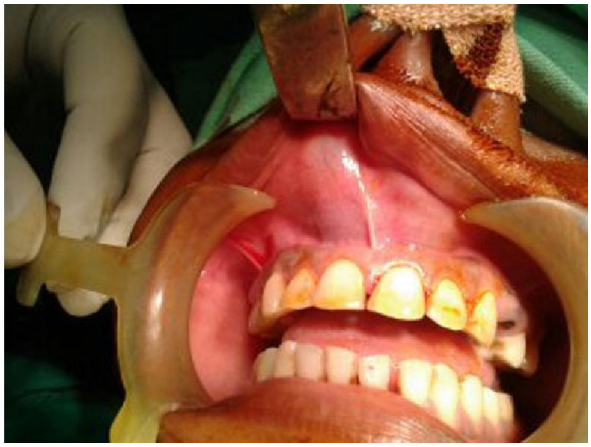

Figure 2. Intra oral view of the lesion, showing obliteration of the vestibule

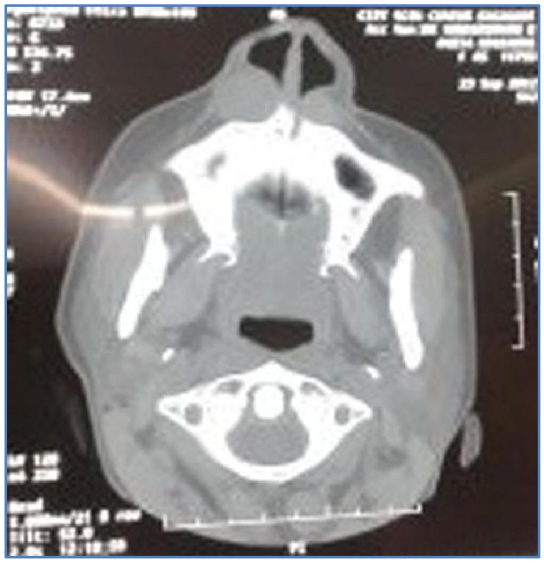

Figure 3. CT Slice showing the lesion below the ala on RT side

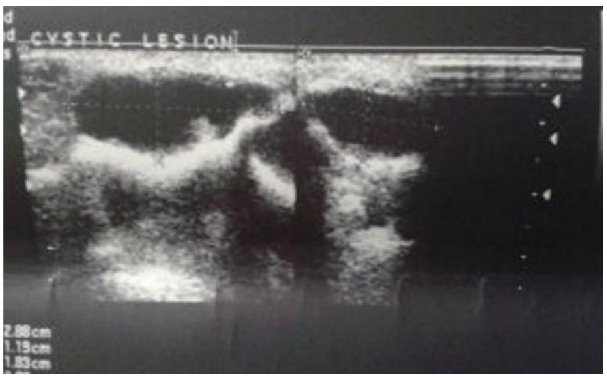

Figure 4. USG showing the Cystic Lesion

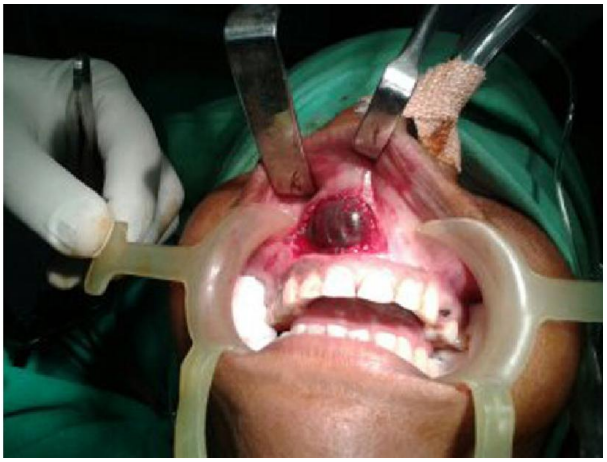

Figure 5. Exposure of the lesion through a vestibular incision

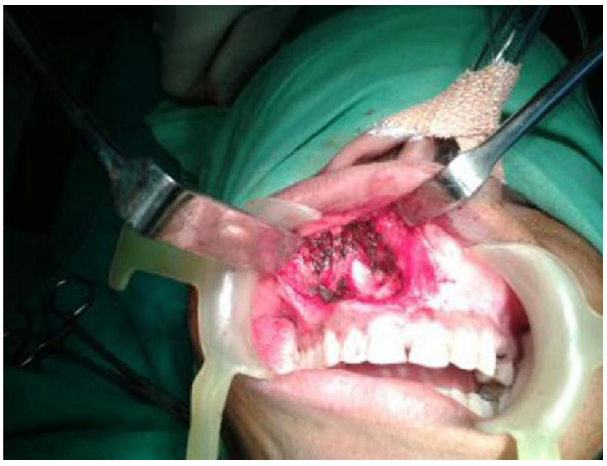

Figure 6. Surgical Defect after tumor excision

Case report

A 24 years old woman reported to our clinic with complaint of a swelling on the right side of face near the naso-labial fold since one year. The swelling progressed to recorded size gradually and was not associated with any pain. She gave a history of paresthesia on the right side of the lip. Physical examination was carried to rule out any more lesions elsewhere on the body as in cases with multiple tumors which turned out to be negative.

Her medical history was unremarkable. On extra-oral examination, the swelling measured about 2.0 X 2.0cms in dimensions, lateral to the right ala of the nose causing obliteration of the nasolabial fold (Fig 1). It had a smooth surface with ill-defined margin, and there was no breach or discoloration of the overlying skin. The swelling was not warm to the touch but was firm in consistency. Skinover the swelling was not freely movable. Intra-oral examination revealed that thes welling had obliterated the maxillary vestibule on the same side 1.0 X 1.5cms in size, solitary with smooth surface, non-compressible with no discoloration of the overlying mucosa (Fig 2). Paresthesia was elicited over right lateral aspect of nose and lip.

CT examination along with ultrasonography were advised. The axial section revealedlesion lateral to the right nasal region obliterating the nasal space (Fig 3).USG revealed a well encapsulated hypoechoic oval lesion measuring 1.2 X 2.2cm's (Fig 4). An excisional biopsy was performed under general anesthesia. To avoid extra oral scar, excision of the lesion through intraoral approach (vestibular incision) was planned and executed (Fig.5 & 6). Clean cleavage plane around the lesion and presence of a thick capsule were the significant surgical findings. The lesion was dissected out from the subcutaneous plane *in-toto* and submitted for histopathologic examination. Based on the clinical findings like location of the swelling and clinical presentation, we assumed the clinical diagnosis to be a nasolabial cyst, but due to the presence of paresthesia Schwannoma was considered for a differential diagnosis. The histopathologic picture showed a well capsulated lesion with mixture of Antoni type A forming Verocay bodies and Antoni type B pattern. Immunohistochemical examination of S-100 protein, the *sine qua non* for Schwannoma was carried out and positive reactivity was observed in both the cytoplasm and nuclei of the neoplastic cells. Post-surgical wound healing was uneventful but, paresthesia was a persistent finding over the right lateral nasal region and over the labial skin and mucosa on the right side after a follow-up of 1year and 10months with no signs of recurrence.

DISCUSSION

Schwannoma is a benign nerve sheath tumor arising from perineural Schwann cells which develop during the 4th week of gestation, when the ectomesenchymal detachment of the neural crest occurs. Formerly known as neurilemoma, it is slow-growing and nearly always benign.(Jadwani et al., 2012) In addition to a typical Schwannoma with classic Antoni A and Antoni B areas, many variant types have been described, such as plexiform, cellular, epithelioid, and ancient Schwannomas. (Wiess et al., 2001) The tumor is frequently located in the head and neck region (about 25–45 %), but only few (1 %) have an intraoral origin (Williams et al., 1993) showing a predilection for the mobile portion of the tongue; however, Schwannoma of the gingival mucosa is extremely rare (Parhar et al., 2014) Schwannomas are known to occur as solitary massesand they occur with a wide age range of first to eighth decades of life (average age 34 years) and with a definite female predilection (Parhar et al., 2014). Schwann cell is a type of glial cell of the peripheral nervous system that helps separate and insulate nerve cells, while a Schwannoma being tumor of these cells that form the nerve sheath. Our patient presented with characteristic clinical features of Schwannoma: slow-growing, solitary, well-encapsulated and smooth-surfaced. But these features are also the presentations of lipoma, fibroma, nasolabial cyst etc. therefore not pathognomonic of Schwannoma, making it a difficult to be diagnosed. Our patient presented with paraesthesia over the right lateral nasal region as well as the labial region, due to which possibility of compression of the nasolabial cyst over the infra-orbital nerve was suspected with neurilemoma being a differential diagnosis. The diagnosis of Schwannoma presenting in this unusual location in our patient would have been difficult to make based upon routine haematoxylin and eosin stains alone. Immunohistochemistry is a simple, accurate, and costeffective technique used in the diagnosis of Schwannoma. S-100 protein is a diagnostic immunohistochemical marker for Schwannoma. (Schnitt et al., 1986)The peri-capsular region of aSchwannoma may contain EMA- and CD34- positive cells. Avery small minority of Schwannomas may show rather extensive immunoreactivity with cytokeratin antibodies, which is thought torepresent cross-reactivity with glial fibrillary acidic protein rather thantrue expression of cytokeratin proteins. (Parhar et al., 2014)

Treatment should ideally be complete excision, wide excision is not being advocated. Neck dissection is not necessary, because lymphatic spread and metastases are rare if at all. (Conley et al., 1975) The paresthesia which was seen in our patient existed pre-op as well as post-op during the follow up period of 22 months, which could possibly be due to inadvertent damage to the terminal branches of the infra-orbital nerve during surgical excision. Generally cure entails a complete resection but this conflicts with the surgical instinct to preserve the nerve of origin. The role of radiosurgery as an alternative for surgical resection is reserved for the cases originating from definitive nerve trunks, e.g. trigeminal, laryngeal, vagus and the vestibular nerve. (Huang et al., 1999) The relevance of this technique in our case would have served little purpose since the nerves involved are extremely small in size. Schwannomas aren't radiosensitive, so radiotherapy plays no role in the treatment. (Jornet et al., 2005) Prognosis for benign Schwannoma of the oral cavity is excellent; rarely does a Schwannoma recur after complete surgical excision. On a follow-up of 22 months post surgically, no signs of recurrence were seen. Although paresthesia was persistent, it did not cause the patient any inconvenience or distress.

Conclusion

Schwannomas should be reckoned with, when examining a well-circumscribed mucosal tumor in the oral cavity or on the face especially when the patients present with paresthesia as a symptom. Clinical examination is a key for the suspected tumor. Histopathologic examination, specifically immunohistochemistry should be done on a routine basis so as to avoid misdiagnosis, and more importantly, prevent incorrect or delayed treatment.

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