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SUBMANDIBULAR REGION SCHWANNOMA - A RARE CASE REPORT

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ABSTRACT

Schwannomas are solitary, homogenous, mostly benign, encapsulated, slowly progressive tumours that originates from nerve sheath layer of soft tissues, internal organs, glands, cranial nerves or peripheral nerve roots. They are associated with neurofibromatosis type1.9 Many schwannomas are symptomless and discovered incidentally on routine imaging. Non-vestibular extracranial head and neck schwannomas most frequently present parapharyngeal neck mass. In the submandibular region, schwannoma can arise from the hypoglossal nerve, the lingual nerve or the submandibular gland, each one being a rare entity.8 In this study, we feature a case of a 12 year old male presenting with a progressive, painless left submandibular swelling. FNAC came out to be salivary gland neoplasm, ultrasound suggested pleomorphic adenoma. Complete exicision of the mass along with left submandibular glad was performed. HPE reported as schwannoma of submandibular region which was confirmed by Immunohistochemistry. Post operative period was uneventful.

INTRODUCTION

Schwannomas, also known as neurilemmoma or neurinoma, are solitary, homogenous, mostly benign,encapsulated, slowly progressive tumours that originates from nerve sheath layer in soft tissues, internal organs, glands, cranial nerves or peripheral nerve roots . Schwannomas of the head and neck are a fairly common occurrence and is reported in approximately 25-40% of total schwannoma cases.[3] It can also be found incidentally in 3–4% of patients at autopsy[2]. It involves the cranial nerves V, VI,VII, X, XI, and XII, the sympathetic and peripheral nerves.[4] The most commonly affected cranial nerve is the vestibular portion of the vestibulocochlear nerve[1]. Verocay bodies with Antoni A and Antoni B areas are histologically seen in schwannomas. Schwannoma of submandibular salivary gland is a rare form of head and neck schwammomas. We present a rare case of a left submandibular region schwannoma of a 12 year old child who presented with a painless, progressive swelling in left submandibular region. FNAC suggested salivary gland neoplasm. Complete excision of the gland was done and HPE reports came out to be schwannoma of left Submandibular region. Patient was relieved of previous symptoms post operatively.

CASE REPORT

A 12 year old Indian male hailing from Guwahati, Assam reported of a swelling in the left submandibular region of 4 years duration, which was painless and was gradually progressive in size. No recent history of fever or prior history of URTI, nor there was pain or increase in size of swelling during eating food. No history of difficulty in respiration. No similar past or family history. On examination, a 3.5cm×3cm singular swelling in the left submandibular area could be palpated which was non-tender, firm, non-fluctuant, non-pulsatile, had smooth surface, didn't move with deglutition with well defined lower border & ill defined upper border and having a normal looking skin overlying it. The swelling was bimanually ballotable and no calculi was palpable over the submandibular gland duct. No neck nodes were palpable. High resolution USG of the submandibular region showed a well circumscribed lobulated hypoechoic and exophytic mass of approx 4x3cm size in left submandibular fossa displacing and compressing the submandibular gland laterally. The mass was showing internal vascularity suggesting pleomorphic adenoma of left submandibular region. USG guided FNAC of the gland showed loose clusters of epithelial cells embedded in hyalinised fibrocollagenous stroma. These cells had moderate amount of cytoplasm and round to oval nuclei with mild anisonucleosis suggesting salivary gland neoplasm which needed confirmation by HPE. After evaluating routine blood and radiological investigations, the patient was prepared for left submandibular gland excision under general anaesthesia after obtaining proper consent. After giving an incision of size 10cm over the left submandibular area, subplatysmal flap was raised upto the lower margin of mandible. The left marginal mandibular nerve was identified and secured. Facial artery identified and ligated. The gland was then separated from anterior belly of digastric and mylohyoid. It was noticed that the growth extended deep and medial over the mylohyoid muscle, which was then partially divided to free the gland. Mylohyoid was retracted superolaterally. The mass was observed to be adhered to and engulfing both lingual and hypoglossal nerve. The mass and submandibular gland was then carefully excised out preserving hypoglossal and lingual nerve. Submandibular ganglion was divided to free the lingual nerve. Wharton's duct was then ligated and divided. Repair of skin done in layers and a drain was kept. Post operative period was uneventful[Figure1]. HPE was consistent with submandibular region schwannoma showing verocay bodies with antony A and antony B areas [figure2]. It was also positive for S-100[Figure 3] and NSE[Figure 4] Immunohistochemistry markers, thus confirming the diagnosis.

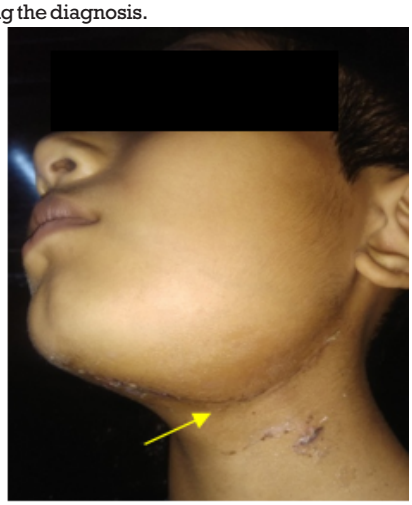

Figure 1: Post-operative submandibular scar(yellow arrow) showing normal healing of wound.

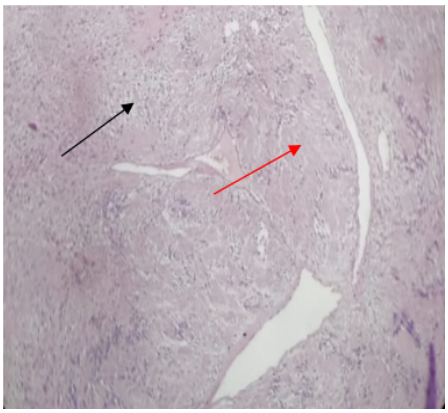

Figure 2: Histopathology showing Verocay bodies and Antoni A and Antoni B areas. Antoni A are densely cellular areas (black arrow), Antoni B in contrary are sparsely cellular areas(red arrow).

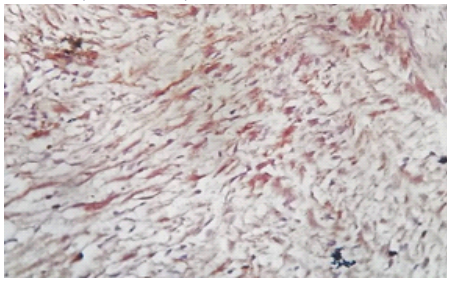

Figure 3: Immunohistochemistry showing S100 protein positivity.

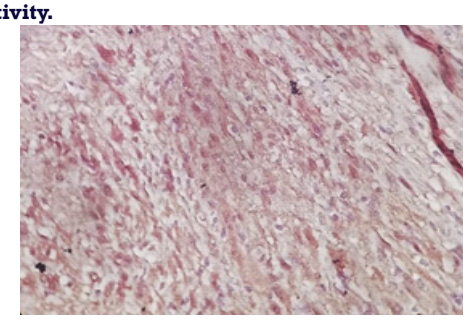

Figure 4: Immunohistochemistry showing NSE (nerve specific enolase) positivity.

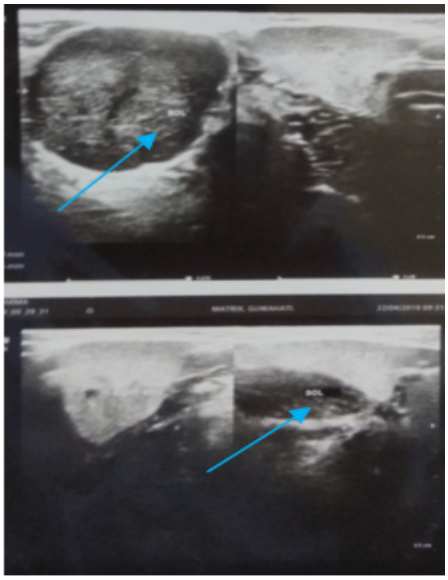

Figure 5: High resolution ultrasonography showing hypoechoic and exophytic Space occupying leshion(blue arrow) in left submandibular gland.

DISCUSSIONS

Schwannoma was first described in 1908 by Verocay, which commonly occurs between 30 and 50 years of age[6] but in our case it is a 12 year old child.

The head and neck region is a common location for benign peripheral nerve sheath tumors (PNST) and a rare site for malignant PNST.[5] According to *Biswas et al*, commonest anatomical location of extracranial head and neck schwannomas was in the neck (42%) and an isolated neck lump was the commonest presentation (77%).[7] It is observed that these schwannomas have a female preponderance over males with the mean age of presentation being 48 years.[8] It can involve cranial nerves such as abducens, trigeminal, facial, vagus, cranial accessory, hypoglossal nerves or the sympathetic and peripheral nerves.[4][9] The most commonly affected cranial nerve is the vestibular portion of the vestibulocochlear nerve.[1] The most common nerves of origin for extracranial non-vestibulocochlear schwannoma were the vagus and the cervical sympathetic chain.[8]

The most common pathology presenting with a submandibular swelling is a pleomorphic adenoma of the gland. Others being reactive submandibular gland swelling or lymphadenopathies.

Schwannoma originating from the submandibular gland is extremely rare and only a few cases have been reported so far.[10] Our case presented with a slowly progressing, solitary and painless, left submandibular swelling with a good post operative prognosis which is clinically comparable with the works done by *Ho CF et al*[10], *Satish Kumar Ranjan et al*[11] and *Gaffar Aslan et al*.[12]

HO CF et al, in their studies showed that computed tomography confirmed a well-defined cystic lesion in the left submandibular space, which caused superior and posterior displacement of the left submandibular gland. It is more or less similar to our study where USG of submandibular area showed circumscribed lobulated hypoechoic and exophytic mass displacing and compressing the submandibular gland laterally.

FNAC is not always conclusive as erroneous results indicating pleomorphic adenoma might be reported. In our case, it could not specify schwannoma but reported as submandibular gland neoplasm.

Intraoperatively, the tumour was found to be engulfing the lingual and hypoglossal nerve, so particular nerve of origin could not be specified. It might have originated from either of the two nerves.

Diagnosis is confirmed by histopathology showing the presence of Antoni A and Antoni B areas, nuclear palisading, whirling of cells and Verocay bodies.[14] Our HPE study also showed elongated spindle shaped Schwann cells arranged in pallisading manner with eosinophilic verocay bodies suggesting schwannoma.

Immunohistochemistry is a useful tool in the diagnosis of these tumours and magnetic resonance imaging is the preferred imaging technique to delineate their extent[7].S-100 IHC marker is universally positive in all cases. They are also positive for collagen-IV[13]. Immunohistochemistry is essential to differentiate benign schwannoma from its malignant forms. In our study IHC markers were positive for S-100 and NSE, confirming the diagnosis.

Surgical excision is the only option in these cases as schwannomas are radio and chemo resistant[9]. We had excised the tumour along with the left submandibular gland preserving both lingual and hypoglossal nerves. More than half of the surgically treated cases exhibited postoperative

neural deficits that were primarily caused by iatrogenic injury to either the nerve of origin or adjacent neural ending.[8] However, postoperatively we had not encountered any such neural deficits.

CONCLUSIONS

History and clinical examination of the pleomorphic adenoma may be as similar as schwannoma, so clinical diagnosis is almost impossible. FNAC is also many a times inconclusive. It is HPE and IHC that confirms the diagnosis. Submandibular gland schwannomas run a benign course and excision is the definitive treatment. Immediate and remote post operative complications are rare. There was no recurrence of disease during follow up.

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