CASE REPORT

Giant Solitary Schwannoma of Submandibular Salivary Gland—A Rare Entity

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Abstract Submandibular gland tumours, especially mesenchymal tumours, are extremely rare. Schwannoma of the salivary gland is a particularly rare form of an extracranial neurogenic tumor, of which ancient schwannoma is one of five variants. Since the first description, only a few ancient schwannomas have been reported in different locations in the head and neck region. In the submandibular gland it is very rare. This report presents a case of 26 year old male patient with an ancient schwannoma of the submandibular gland seems to be arising from parasympathetic fibers.

Keywords Schwannoma · Neurilemmoma · Ancient · Submandibular gland

Introduction

Asymptomatic long standing large submandibular swellings are commonly diagnosed as benign salivary gland tumors like adenoma. Differential diagnosis for these lesions include cervical adenitis, granulomatous disorders like sarcoidosis or autoimmune disorders such as Sjogren's syndrome, Minkuliz's disease.

Schwannoma is not a common salivary gland tumor, which arises from nerve sheath cells. Schwannoma, also known as neurilemmoma or neurinomas was first described by Verocay in 1908 and in 1935 the term neurilemmoma was coined by Verocay and Stout [1, 2]. The term ancient neurilemmoma was first suggested by Ackermans and Taylor [3] in a review of 48 neurogenic tumors of thorax. They reported 10 cases which showed similar features of a typical neurilemmoma but distinctive because significant portion of tumors were composed of few schwann cells within hyalinised matrix. They classified that these features occurred in schwannomas of long standing origin. Schwannomas are characterized by diffuse areas of cellularity, focal accumulations of Hypocellular material, fatty degeneration, thick capsules, relative absence of necrosis and being usually infiltrated by large numbers of siderophages and histiocytes along with hyperchromatism are misleading in suggesting malignancy [2]. Schwannomas were first classified as benign salivary gland tumor by Rankow [4].

Nearly 45% of all schwannomas occur in the head and neck region, where they may originate from any of the peripheral, cranial or autonomic nerves [5]. A thorough search of literature revealed that there is an increase in the incidence of schwannoma in head and neck region varying from 25 to 45% [6]. However the intraoral lesions account for only 1% of all tumors and tongue is the most commonly

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affected organ followed by palate, buccal mucosa, lip and gingiva [7].

Salivary gland tumors are uncommon, representing 3–10% of all neoplasm of the head and neck region, and 70% of them involve the parotid gland. Sublingual gland tumors are very rare, accounting for only 0.3–1% of all salivary gland tumors, and most are malignant [8].

Sodagar and Daneshbod [9] published the first case report of a schwannoma of the submandibular gland in the year 1917. Since then only fifteen cases have been sporadically reported. Out of these cases only one was malignant schwannoma.

Case Report

A 26 years old male patient reported with a 18 month history of painless left upper neck swelling. Patient does not complain of pain, trismus, foul smell and altered sensations. On physical examination, a 7×4 cm swelling was noted extending from submental triangle to submandibular region over the left side of neck. On palpation swelling was firm to hard in consistency, with no extra-oral draining sinus, and skin was freely movable over the underlying pathology. On bimanual palpation a ballotable mass was palpable. Intraorally swelling was confined till midline and was firm to hard in consistency over the floor of the mouth on the left side (Fig. 1). Salivary flow from left Wharton's duct was normal. Patient did not have any sign and symptom of paraesthesia or anaesthesia over the tongue, floor of the mouth and no deviation of tongue on protrusion, which suggests normal function of lingual and hypoglossal nerve. Clinically provisional diagnosis of benign salivary gland tumour was made.

Contrast CT SCAN revealed a $59 \times 39 \times 58$ mm well circumscribed round to oval mass in left submandibular region with few foci of calcification. Large mass showing mild heterogeneous enhancement without involvement of mandible. This heterogeneous mass was primarily involving the deep lobe of submandibular gland, superficial lobe was normal, compressing the floor of the mouth and abutting the medial pterygoid muscle. Overall impression CT findings suggestive of solid lesion without cystic cavity which may be benign mixed tumor, lymph nodal mass or neurogenic tumor (Fig. 2).

FNAC was inconclusive and all laboratory investigations were within normal limit.

Clinical and radiographic findings were in favour of benign lesion of submandibular gland. Excision of the lesion along with the submandibular salivary gland was planned under general anaesthesia. Meticulous layer wise dissection and ligation of encountered facial vessels was performed to expose the submandibular gland via

Fig. 1 Pre-operative intra-oral photograph showing large mass occupying the floor of the mouth

submandibular incision. Intra operatively during excision of the gland it was noticed that multiple parasympathetic nerve fibres were entering the deep part of gland anchoring the lingual nerve to submandibular gland. These intra-operative findings suggested that the tumor may be arising from parasympathetic nerve fibres. On macroscopic examination tumor was entirely involving the deep part of submandibular gland whereas superficial part of submandibular gland was normal in appearance. Tumor was well encapsulated, firm to hard in consistency and the excised specimen was sent for histopathological examination (Fig. 3).

Histopathologic Features

Macroscopically, excised specimen of submandibular gland showed superficial lobe measuring approximately $3.0 \times 2.5 \times 2.0$ cm, lobulated surface, greyish white in colour and deep lobe measuring approximately $6 \times 4.5 \times 3.0$ cm, smooth shiny surface, yellowish white in colour. The cut surface of deep lobe showed centre dark brown cystic area with papillation.

Fig. 2 Contrast computed tomogram showing a well defined lesion in left deep lobe of submandibular gland with few foci of calcification

Fig. 3 Intra-operative excised specimen showing normal superficial lobe and tumor only involving deep lobe of submandibular gland

Microscopic examination revealed a well encapsulated tumor composed of two patterns, areas of organized spindle-shaped cells in a palisading arrangement around acellular, eosinophilic areas, forming Verocay bodies giving an Antoni type A pattern and loose textured myxoid areas Antoni type B with microcyst formation and hyalinization. The cells in the compact areas had oval, smooth nuclei and poorly defined eosinophilic cytoplasm. There were no mitotic figures. Antoni type A areas predominantly made up the tumor volume. These findings were suggestive of Ancient schwannoma.

Discussion

Schwannomas, that involve or encroach upon the major salivary glands is a rare entity and has been classified by Auclair et al. [10], as benign non epithelial neoplasm of major salivary glands of mesenchymal origin. After Attie et al. [11] described a case of schwannoma arising from lingual nerve and Sutay et al. [12] reported a schwannoma originating from hypoglossal nerve. Sato et al. [13] published first case report of parasympathetic schwannoma of submandibular gland. In the year 2003, Arde et al. [14] first reported an unusual case of intra-oral schwannoma of sublingual gland. Schwannoma are often difficult to diagnose as they do not have any characteristic identifying features. The features are comparable to any benign tumor arising from salivary gland parenchyma or ducts. A tumor of mesenchymal origin usually presents as an asymptomatic, slow growing, well-circumscribed mass in the preauricular, submandibular, or less frequently in submental region. The preoperative evaluation of such tumors often is perplexing if based primarily on the clinical features and medical history [15]. Pre-op diagnosis of schwannoma is difficult except MRI. A characteristic target sign is seen as an increased peripheral signal intensity and decreased central signal intensity on T2 weighted images. Pre-op MRI was not advised as it is not a routine diagnostic aid however, MRI with gadolinium is the better choice because it can diagnose even small intra glandular nerve cell tumor [16].

Head and neck schwannomas are frequently misdiagnosed and preoperative investigations are often fruitless [15]. Although aspirates of ancient schwannoma may show some features such as nuclear pleomorphism, nuclear inclusions, perivascular sclerosis, xanthomatous changes, or nuclear atypia [17], but fine needle aspiration biopsy has questionable value [18]. It may show unclear histopathological results from these cases, which have the potential to confuse this lesion with a more serious one such as sarcoma [15, 17]. Even in the present case FNAC was non conclusive so intra-operative frozen section of excised pathology may be helpful in diagnosis of benign or malignant, but due to non functioning of cryostat could not

be performed in this case. Mostaan et al. [19] reported that frozen section's identification of major salivary gland tumor mass as either benign or malignant has 98% sensitivity, 100% specificity, 99.2% accuracy, 100% positive predictive value, 99% negative predictive value and concluded that frozen section is considerably accurate in diagnosis of malignant versus benign pathology, regardless of exact histopathological variety. Post operatively patient can be managed by chemotherapy, radiotherapy or both according to the pathology if turned out to be malignant.

The size of Schwannoma arising in the submandibular glands range from 1.0 to 6.0 cm in diameter and our case measured approximately $6 \times 4.5 \times 3.0$ cm [10]. They may appear fusiform in shape along the nerve of origin or may occur as a well defined eccentric nodule or mass. Tissue sections of tumor appear gray-white with fibrillary shiny areas, variably sized cystic spaces, or hemorrhagic foci. Histologically five variants of schwannoma have been described namely common, plexiform, cellular, epithelioid and ancient schwannoma [20]. Schwannoma shows characteristic histologic appearance, dominated by an encapsulated lesion arising from a nerve and composed of an intimate mixture of spindle cells forming highly cellular so called Antoni type A areas and less cellular, myxoid Antoni B areas. Between palisades of Antoni type A cells are regions that are devoid of nuclei termed Verocay bodies, after the person who first described schwannomas. The present case showed predominantly Antoni type A areas. However, the Antoni A and B areas are used strictly for pathologic determination of the tumor and have no clinical significance. However, histopathologic features of schwannoma are quite characteristics and can be diagnosed light microscopically as in the present case. But, In diagnostically problematic cases, immuno-peroxidase and ultrastructural studies should provide incontrovertible support. Immunohistochemical evaluation of neurilemmomas for S-100 protein provides consistently positive results, especially in Antoni type-A areas. Additionally, tumour cells also show positivity for neuron specific enolase, vimentin and glial fibrillary acidic protein. Electron micrographs show characteristic layers of electron-dense basal lamina and long-spacing collagen stacked between Schwann cells [11, 16].

Schwannoma are best managed by complete surgical excision and radical treatment is not advisable due to rare recurrence after surgery.

Conclusion

Rare benign tumors like schwannoma should also be included in the differential diagnosis of salivary gland tumors and the same should be added to the classification of salivary gland tumors by WHO.

Compliance with Ethical Standards

Conflict of interest No conflict of interest.

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