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Case Report

Submandibular gland pleomorphic adenoma masquerading as a multinodular goitre: a case report

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ABSTRACT

Tumors of the salivary gland comprise of a small percentage of all head and neck tumors, of which nearly 80% are benign. The submandibular gland tumors constitute only 5-10% of this group, of which pleomorphic adenoma is the commonest type. They can weigh several kilograms and grow to ludicrous sizes. If left untreated, they may result in facial deformity and airway compromise. We describe a case of a big pleomorphic adenoma arising from submandibular salivary gland that developed in a 65 years old female presenting as a swelling in right side of neck since 15 years. The dimensions of the lesion were roughly around 8×12×11 cm. During surgical examination, a solid mass that originated from the right submandibular gland was discovered to be linked to both the skin and the soft tissue beneath it. Pleomorphic adenoma was confirmed by histopathological examination. Review and discussion of the relevant literature was conducted. This article's emphasis is on the fact that, although the pleomorphic adenoma is a well-reported entity, they can attain very large sizes involving the neck region and can misrepresent itself leading to diagnostic dilemma due to many possible differential diagnoses of neck swellings.

Keywords: Submandibular gland, Pleomorphic adenoma, Benign salivary gland tumors

INTRODUCTION

Pleomorphic adenoma is the most common major salivary gland neoplasm. Around 80% of them arise in the parotid glands, whereas 10% originate in the minor salivary glands. The pleomorphic adenoma of the extra-parotid site is defined by its location outside the primary salivary gland. The minor salivary gland adenomas occur at the hard and soft palate, lips, tongue, lacrimal glands, pharynx, larynx, paranasal sinus, and nasal cavity. Pleomorphic adenoma in parapharyngeal space may occur de novo or as an extension of the deep lobe of the parotid tumors.[1] It was previously known by other names such as mixed tumor, enclavoma, branchioma, endothelioma, and enchondroma de novo pleomorphic adenoma in parapharyngeal space is uncommon and develops from salivary gland tissue abutting a lymph node.[2] Other sites include the palate, larynx, submandibular gland, nasal cavity, etc. FNA can determine whether the tumor is malignant in nature with an approximate sensitivity of 90%.[3]

Core needle biopsy is more invasive but provides more accurate histological typing of the tumor with a diagnostic accuracy of around 97%. Predictors of malignant transformation include variation in consistency, rapid growth, pain and tenderness and regional lymphadenopathy, and facial nerve dysfunction.[2] Other risk factors for malignancy include advanced age, radiation therapy, large size, and recurrent tumors.[4] The tumors of the submandibular glands are treated with simple excision procedure with preservation of adjacent

nerve including the mandibular branch of the trigeminal nerve, the hypoglossal nerve, and the lingual nerve.[5]

In this report, we present the case of a 65-year-old female with a massive pleomorphic adenoma originating from the submandibular salivary gland, initially misdiagnosed as a multinodular goitre due to its location and size. This case underscores the diagnostic challenges posed by such tumors and highlights the importance of thorough investigation to avoid misdiagnosis.

CASE REPORT

A 65-year-old female presented with complaint of painless swelling across right side and front of her neck since 15 years. Over the course of 15 years, the swelling, which was initially pea-sized, steadily grew in size. The swelling was associated with dragging sensation, difficulty in breathing on lying down and restricted neck flexion. There was no history of fever, dysphagia, change in voice, or heat/ cold intolerance. The patient was otherwise healthy with no other significant medical history or findings.

On examination, a single approximately 9×12×10 cm sized multinodular, non-tender, hard, and mobile swelling which was present over anterolateral aspect of right side of neck extending from the floor of the mouth just below right mandibular ramus up to just above the right clavicle and 2 cm beyond the right sternocleidomastoid to 1cm across the midline over to the left side. The overlying skin was normal, without any dilated veins or ulceration. The swelling however did not move with deglutition or tongue protrusion.

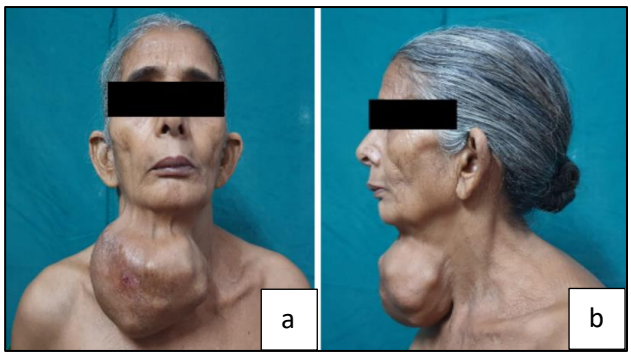

Figure 1: (a) Anterior view of neck swelling, (b) Lateral view of neck swelling.

All routine blood tests were within the usual limits, additionally the thyroid profile also turned out to be normal (TSH 4.90 ng/ml, T3 1.1 ng/ml, T4 4.2 mcg/ml) Imaging studies were performed for further evaluation. An ultrasound of the neck revealed a mixed echogenic lesion on the right side of the neck that was about 7×11×12 cm in size and included interior cystic regions, vascularity, and calcification. Although still patent, the lesion abutted and compressed Internal jugular vein and Common carotid arteries. Postero-laterally, it abutted right sternocleidomastoid muscle. The epicenter of the tumor was found to be within the right submandibular salivary gland. The maxilla and mandible's bony structures were normal. MRI revealed approx. 7.7×11.0×10.5 cm sized large well-defined heterogeneous nodular lesion involving right side of neck spanning from floor of mouth to supra clavicular level with involvement of adjacent skin. It crossed midline on left side. There were few blooming areas with in suggesting calcification. It causes significant mass effect inform of compression over right sternocleidomastoid muscle, trachea, right thyroid submandibular gland and right Internal jugular vein with maintained fat planes, left sided tracheal deviation. Lesion on T1 weighted MR image appeared hypointense and appeared hyperintense on T2 weighted MR images No evidence of any cervical lymphadenopathy was found.

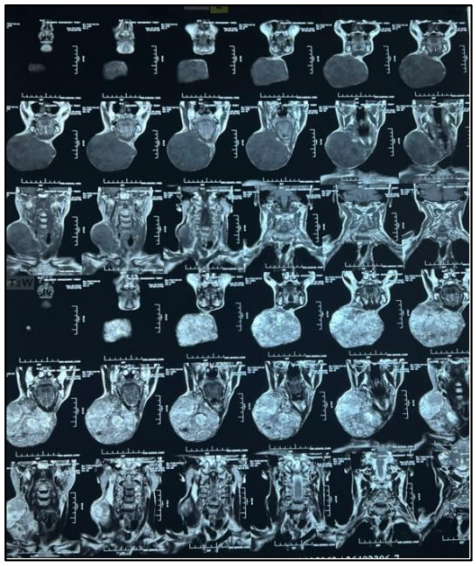

Figure 2: MRI T1W of neck-coronal section depicting right submandibular gland lesion.

Under local anaesthesia, fine needle aspiration cytology (FNAC) was carried out. The report was suggestive of benign salivary gland neoplasm containing admixed epithelial, myoepithelial and mesenchymal tissue elements with moderate cytological atypia. This raised the prospect of the mass being a pleomorphic adenoma of submandibular salivary gland. T1 weighted MR image appeared hypointense and appeared hyperintense on T2 weighted MR images no evidence of any cervical lymphadenopathy was found.

Patient was taken for surgery. A horizontal elliptical incision was kept over the centre of the swelling and an en bloc resection of the tumor was done with preservation of the surrounding tissues, major vessels and nerves. The swelling, which had a smooth surface and no signs of hypervascularity or haemorrhagic regions, was observed to be to be arising from the floor of the mouth over the lower aspect of right submandibular gland. The entire swelling along with majority of right submandibular

gland was excised and sent for histopathological examination. The excised tumor mass was 8×12×11 cm in dimension. The weight of the excised mass was 1.6 kg.

Wound was closed primarily with a negative suction drain placed in situ. The post-operative recovery was uneventful day and the patient was discharged on post-op.[5] The incision wound also healed well.

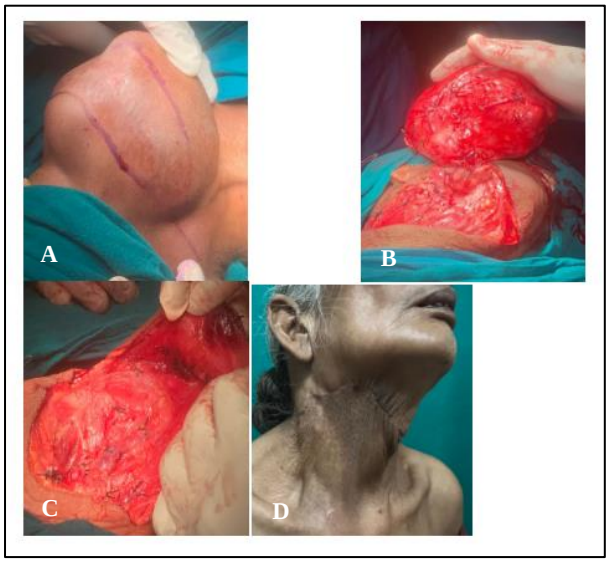

Figure 3 (A-D): Sequential surgical excision of mass and post operative scar mark of surgery.

Histopathology revealed a well capsulated, highly cellular mass with interspersed epithelial cells and myoepithelial cells containing eosinophilic cytoplasm along with salivary gland. The cut surface was pinkish, solid, gelatinous and shiny. Thus, the diagnosis of benign mixed submandibular salivary gland tumor-Pleomorphic adenoma was confirmed.

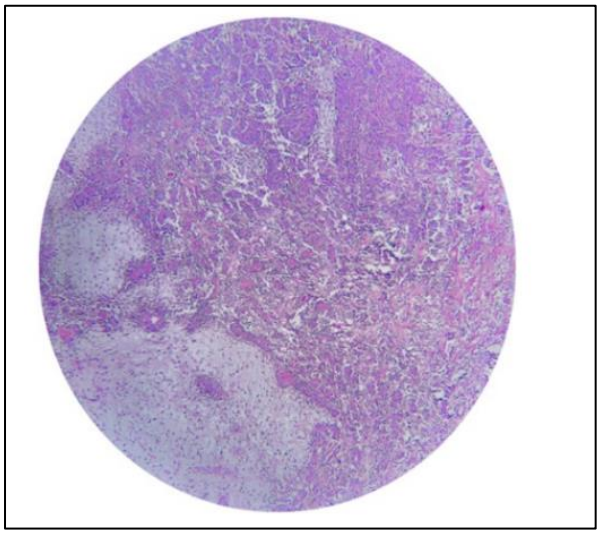

Figure 4: Histopathological photograph of H and E-stained specimen showing features of pleomorphic adenoma of salivary gland.

DISCUSSION

This clinical case describes an intriguing instance of a pleomorphic adenoma misrepresenting as a multinodular goitre owing to its large size, nodularity, and ectopic location. However, after further examination, it was discovered that the mass was actually a pleomorphic adenoma of the submandibular salivary gland. Thyroid swellings also have the tendency to attain large sizes thus occupying the whole neck region however they can be differentiated from other neck swellings by the following findings i.e. they move with deglutition/protrusion of tongue, there is associated cervical lymphadenopathy and nerve impairment leading to hoarseness of voice. The thyroid function tests may also be altered in some cases while in cases of pleomorphic adenoma of the salivary gland these features are not present. Salivary gland tumors are the rare tumors (comprising of less than 5% of all head and neck tumors), commonly affecting the parotid gland (80%) followed by 10% each of submandibular and sublingual salivary glands 6. Although less frequently, they have also been noted to develop in the main bronchus, columella, larynx, pharynx, trachea, lacrimal gland, sinonasal tract, and maxillary sinus. It affects all age groups, however, it peaks in 6th decade with a female preponderance.[7]

The most common neoplasms in the submandibular glands are pleomorphic adenoma (36%), adenoid cystic carcinoma (25%), mucoepidermoid carcinoma (12%) and malignant mixed tumor (10%). Pleomorphic adenoma of the submandibular salivary gland presents as a slow-growing, painless, smooth, firm, mobile mass in the submandibular region without any fixity to underlying structures. The tumors do not produce any symptoms other than pressure symptoms due to attaining a large size. There is usually no cervical lymphadenopathy or features of nerve involvement.[8] Pleomorphic adenomas do not invade the underlying structures owing to their benign nature. The malignant transformation rate is about 8.5 % however only 0.15% of pleomorphic adenomas undergo malignant change.[7,8]

Carcinoma ex pleomorphic adenoma and metastasizing benign mixed tumor are the two variants that undergo malignant transformation.[10] Radiotherapy is not indicated due to the radio resistant behaviors of the tumor. Prognosis is excellent (95 %) after complete excision.[11] Willis proposed three main hypotheses to explain this kind of heterotopias. These were an abnormal persistence and development of vestigial structures, dislocation of portion of a deficient rudiment during mass movement and development, and abnormal differentiation of the local tissues (heteroplasia). This may happen due to metaplasia, neoplastic degeneration of ectopic salivary gland tissue or due to the implantation after surgical excision of the salivary gland tumor.[12]

A pathological confirmation is necessary since the ectopic development of a pleomorphic adenoma in an

uncommon site can present surgeons with a diagnostic challenge. Ultrasound is the initial modality of investigation however, CT scan or MRI are the gold standard for radiological investigation.[13] Furthermore, the diagnosis is aided by FNAC from the swelling. The use of fine needle aspiration cytology (FNAC) is debatable. As its diagnostic accuracy ranges from 80 to 95 %, some writers prefer it and believe it to be a commonly used preoperative diagnostic tool.[14] However, some people believe that pre-operative needle aspiration biopsy is inappropriate in any benign mixed tumor because it increases the risk of tumor cell implantation and may also increase the likelihood of recurrence.[15] The final diagnosis is always histopathological after excision of the lesion. The choice of surgery is to excise the entire tumor with the involved submandibular salivary gland.[16] This is significant because, given that these tumors frequently have microscopic pseudopod-like extensions into the surrounding tissues through the capsule, incomplete excision, rupture of the capsule, or tumor spillage during excision could seed tumor cells into the surrounding tissues and cause recurrence.[17] The commonest complication observed in this type of surgery is injury to marginal mandibular nerve leading to temporary or permanent paralysis.[18]

Recurrence rates have reportedly decreased. 7% of 1,342 patients with benign parotid neoplasms and 6% of patients with benign small salivary-gland tumors experienced a recurrence, according to Spiro et al.[19] Pleomorphic adenomas have been known to come back over time, though. According to Verma et al 20 follow-up exams should be conducted for up to 10 years. Follow-up of our case after surgery showed smooth healing of the wound with no evidence of recurrence after one year. In our case histopathology report confirmed the diagnosis of pleomorphic adenoma. Histopathologically, the myxoid stroma is one of its most characteristic features, 2 admixed with mucoid, fibro hyaline or chondroid stroma.[21] Three histological subtypes of the pleomorphic adenoma exist: myxoid (80% stroma), cellular (predominantly myoepithelial cells), and mixed (classic). With areas of epidermoid metaplasia, epithelial cells are organized in cord-like and duct-like cell patterns.

There are fibrous, hyaline, myxoid, cartilaginous, and osseous regions seen in the intercellular matrix. Such pleomorphic extracellular matrix is produced by myoepithelial cells. Lesions in minor glands are frequently more cellular or solid than those in major glands, and myoepithelial cells in these lesions are frequently polygonal with a pale eosinophilic cytoplasm, giving them an epithelioid or plasmacytoid character. Acini, the typical form of salivary architecture, were also seen.[10,13] The firmness of these tumors varies with the nature and amount of the stromal component the tumors are encapsulated with varying thickness of the pseudo capsule due to compression of surrounding structures and fibrosis.

Reporting a case of this kind is important because delaying treatment can lead to rapid tumor growth (which some believe is caused by mutated RAS genes), poor aesthetics, psychological trauma, and a potential threat to life, particularly when the tumor presses on the parapharyngeal space and oropharynx and is made worse by infection, which could compromise the airway.[14] The growing mass could further deform the anatomy, displacing important components and complicating the dissection. The presentation of scenarios like the one being described underscores the plight of many patients, particularly in underdeveloped countries. Whatever the cause, such illness progression should never be deliberately ignored, especially in the twenty-first century.

CONCLUSION

This case report highlights a rare instance of pleomorphic adenoma presenting as a large, multinodular mass in the submandibular region, initially misdiagnosed as a multinodular goitre in 65-year-old female. Pleomorphic adenomas, though predominantly occurring in the parotid glands, can present in atypical locations such as the submandibular gland, where they pose diagnostic challenges due to their potential similarity to other neck masses. Thorough diagnostic workup including imaging and fine needle aspiration cytology ultimately revealed the presence of a pleomorphic adenoma. The successful surgical excision of the tumor, which weighed 1.6 kilograms, confirmed the diagnosis and allowed for complete histopathological evaluation. For distinguishing of such case from usual cases is the importance of distinguishing between thyroid and salivary gland tumors through detailed examination and imaging and the necessity of histopathological confirmation post-surgery for accurate diagnosis and treatment.

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