Case Report

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Primary Pleomorphic Adenoma (Minor Salivary Gland) of Parapharyngeal Space

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Abstract

Parapharyngeal space is one of the potential facial planes of head and neck that may be involved by various pathological processes:infectious, inflammatory andneoplastic. Neoplastictumours seen at the parapharyngeal space represent less than 1% of all head and neck tumours. Both benign and malignant tumours may arise in this location, majority appears to be benign. Mostparapharyngealspace tumours are salivary gland tumours, neurogenictumours and lymphoreticular lesionswhich comprise nearly 75-80% of all parapharyngeal space tumours. We report a case of Pleomorphic adenoma arising de novo in the parapharyngeal spacewhich is a very rare site of its occurrence. World literature suggests parapharyngeal space lesions account for only 0.5% head and neck tumours and the majority of the minor salivary gland tumours are malignant. Pleomorphic adenoma arising de novo in the parapharyngeal space is of rare occurrence. High index of suspicion and an adequate clearance of the tumour with a cuff of surrounding dispensable normal tissues is the key to successful treatment of such tumours.

Keywords: Parapharyngeal; Salivary Gland Tumour; Pleomorphic; Adenoma.

Introduction

Parapharyngeal space (PPS) resembles an inverted triangular pyramid with concave faces. This space is located posterior to infratemporal fossa anteriorly, nasopharynx, the lateral pharyngeal wall medially, vertebral column posteriorly, and mandibular ramus laterally.

Parapharyngeal space tumours are not very frequent, accounting for some 0.5% of neoplasms of head and neck. Most of these tumours (70%-80%) are benign and 40-50% of these originate in the salivary glands, particularly the pleomorphic adenoma. Pleomorphic adenoma in the parapharyngeal space (PPS) can develop *de novo* or may arise from deep lobe of the parotid and extend through the stylomandibular tunnel into the PPS. The origin of *de novo* pleomorphic adenoma is probably from displaced or aberrant salivary gland tissue within a lymph node. Pleomorphic adenoma arising from the epithelial rests ofthe salivary gland tissue in the parapharyngeal space is very rare [1-3].

Very few cases of Pleomorphic adenoma arising in the parapharyngeal space has been reported so far in the English literature which makes this case report worth reporting.

Case Report

A 15year old male patient came to the ENT outpatient department with the complaints of progressively increasing painless swelling in the oral cavity, towards base of tongue since 8 months. He complained of difficulty in swallowing and change in quality of voice for 5 months. There was no history

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of weight loss, fever, bleeding from noseor symptoms like earache. On physical examination there was a smooth firm bulge of soft palate extended upto the right lateral pharyngeal wall. The swelling was bimanually palpable and clinically appear to be free from underlying structure.There was no significant cervical lymph node enlargement with intact cranial nerve functions.

Clinical diagnosis of benign parapharyngeal tumourwas suspected with the advice of CT scan to see the extent of tumour. CT scan of head and neck showed a large homogenously enhancing tumour of size 7.5x5.5x4.5 cm located in the prestyloid compartment of the right parapharyngeal space. (Figure 1) The whole tumorous mass was excised under general anaesthesiawith ease and sent for histopathology. Post- operative period was uneventful.

Grossly, the excised mass was well encapsulated, globular, smooth, greyish white cut surface measuring 7.5x5.5x4.5 cm. Cut section of the mass was homogenous, solid,gelatinous,greyish white lobulated appearancewith multiple tiny cysts filled with coagulated mucin (Figure 2).

Microscopy, of the resected mass showed biphasic appearance of intimate admixture of epithelial and stromal components. Epithelial components are arranged in acini, tubules and solid pattern. Neoplastic glands have a lining composed of two types of cells, basally located myoepithelial cells which may be cuboidal, flattened, clear or spindle shaped and luminally situated cells were cuboidal to columnar cell type.Stroma had characteristic chondromyxoid appearance (Figure 3).

Histopathological diagnosis of Pleomorphic adenoma was made. The patient postoperative course was uneventful. No recurrence was observed after follow- up of repeat dressing at 2 and 4 weeks duration.

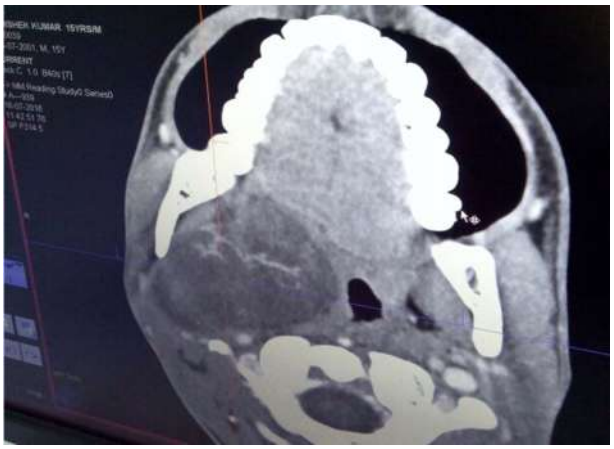

Fig. 1: CT scan of patient showing a large homogenously enhancing tumour in the prestyloid compartment of right parapharyngeal space

Fig. 2: Gross image of cut section of the tumour mass which was well encapsulated, solid, greyish white, homogenous and gelationous having lobulated appearance with multiple tiny cysts filled with coagulated mucin.Inset shows the entire encapsulated globular tumour mass

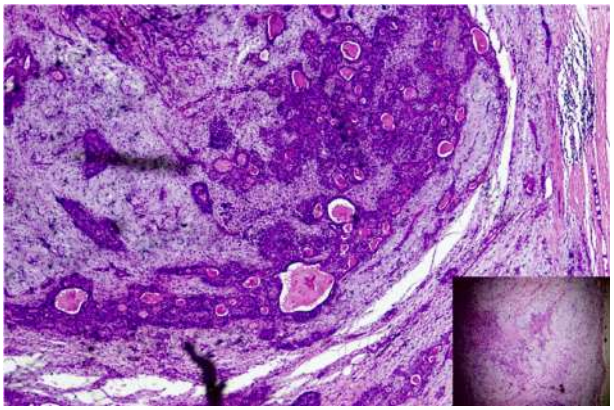

Fig. 3: Microphotograph showing biphasic appearance of intimate admixture of epithelial and stromal components with epithelial components are arranged in acini, tubules and solid pattern against a chondromyxoid background. (H& E;400 X) Inset shows a tumour mass with a well defined fibrous capsule and bhipahsic appearance of epithelial and stromal components. (H&E;Scanner view)

Discussion

Parapharyngeal space masses account for 0.5% of all headand neck tumours and the majority are histopathologicallybenign (76%) [4]. Neoplasms of salivary gland origin arelocated in the prestyloidparapharyngeal space andaccount for 40–50% of parapharyngeal space lesions [5]. Salivarygland neoplasmsmay arise from the deep lobe of the parotid gland, ectopic salivary rests, or minor salivary glands of the lateral pharyngeal wall. Minor salivary gland tumours constitute 22% of all thesalivary gland tumours and among them only 18% ofthem are histopathologically benign, the rest being malignant. Among the benign salivary gland tumours seen inthe parapharyngeal space the most common prestyloid lesion is pleomorphicadenoma, which

represents 80–90% of salivary neoplasms intheparapharyngeal space [6,7]. According to a study the most common site of pleomorphic adenoma of the minor salivary gland is the palate followed by lip, buccalmucosa, floor of mouth, tongue, tonsil,pharynx, retromolar area, and nasal cavity. Unlike mostmixed tumors, those in the palate and lip frequently lack acapsule. Pleomorphic adenoma arising from the epithelial rests ofthe salivary tissue in the lymph nodes of the parapharyngeal space is very rare. According to the literature, there are fewcases reported up till now that have taken the origin fromthe minor salivary gland tissue of the parapharyngeal space [1,2].18% of thetumours arising in the minor salivary glands are benign andpleomorphic adenoma is the commonest of all [8]. In two of the cases reported in this study, the tumoursoriginated from the minor salivary gland tissue of the parapharyngealspace.Pleomorphic adenoma may also invade parapharyngeal space as an extensionof the deep lobe of the parotid gland [9]. Parotid glandtissue can herniate through a weakness in the stylomandibularmembrane and lie in the lateral pharyngeal wall. Forthis reason, tumors deep in the parotid gland can presentas parapharyngeal masses [10]. Almost10–12% of pleomorphicadenomas of the parotid are thought to arise from the deeplobe of parotid and parapharyngeal extensions of the mass inthe deep lobemay remain asymptomatic until reaching a verylarge size [11].

Histopathologically, pleomorphic adenoma is thinly capsulated with few small smooth contoured buds that may protrude through the fibrous capsule. The prototypic histologic appearance consists of tubular structures enveloped by myoepithelial mantles submerging in chondromyxoidstroma.Theluminal epithelial component takes the form of anastomosing tubules, cysts, ribbons and solid sheets.The cells are columnar, cuboidal or flat. The duct lumen may be empty or filled with PAS positive diastase resistant eosinophilic material.Myoepithelial cells appear as cuboidal, spindle, stellate,plasmacytoid hyaline and hydropic clear cells [12,13].Plasmacytoid hyaline cells represent the most distinctive form of modified myoepithelial cells,they are oval shaped with homogenous eosinophilic hyaline cytoplasm. The nucleus is round,eccentrically located with a tendency of peripheral margination of the dense chromatin.The extracellular stroma takes the form of a mixture of chondroid,myxoid,chondromyxoid, hyaline and very rarely osseous and adipose tissues.The presence of chondromyxoidstroma in a salivary gland tumour is practically pathognomonic of pleomorphic adenoma [14].

Complete surgical enucleation is the treatment of choice. Rate of recurrence is high in surgical exposure of tumour or tumour capsule risk spillage, but pleomorphic adenoma of the minor salivary glands have little propensity for recurrence. The most frequent surgical issue are pseudopodia, capsular penetration and tumour rupture.

Conclusion

Pleomorphic adenoma arising de novo in the parapharyngeal space is of rare occurrence, if adequate clearance of tumour with a cuff of surrounding normal tissues is excised, is the key to successful treatment of such tumour.

A histopathological biopsy should be routinely taken after the excision of the neoplastic lesion. Wide excision with negative margins is the optimal surgical approach to avoid recurrence and the key to successful treatment of such tumour.

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