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Schwannoma-like Pleomorphic Adenoma: A Rare Variant

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Abstract: Pleomorphic Adenoma is the commonest neoplasm of the salivary gland. It has a characteristic biphasic pattern with epithelial-myoepithelial component and a mesenchymal component. This variably capsulated tumor is characterised by its architectural pleomorphism. Myoepithelial component varies from spindle to plasmacytoid to oncocytic where as the mesenchymal component is mucoid, myxoid or chondroid. We report a rare case or pleomorphic adenoma with palisading schwannoma like areas.
Keywords: Myoepithelial component, Pleomorphic Adenoma, Salivary gland, Schwannoma-like.

INTRODUCTION

Pleomorphic adenoma is the most common salivary gland tumour and accounts for about 60% of all salivary neoplasms. About 80% of pleomorphic adenomas arise in the parotid, the lower pole being the most common location [1]. Despite their protean histopathology, each tumor shares with others the essential diagnostic features of being composed of epithelial and mesenchymal components [2]. The remarkable degree of morphological diversity is what earns pleomorphic adenoma a special distinction. The modified myoepithelial component giving rise to palisading spindle celled areas resembling a schwannoma is extremely rare.

CASE REPORT

A 53 year old male presented with a swelling over the left parotid since 2 years. The swelling was insidious in onset with gradual increase in size. On examination the swelling was soft to firm in consistency and non tender. Head and neck 3D volumetric CT scan revealed a 3x2x2.4 cms well defined heterogeneously enhancing soft tissue density mass lesion in the superior lobe of left parotid- possibly benign pleomorphic adenoma with cystic change.

FNAC of the same done outside yielded clusters of plump spindle cells, benign epithelial cells in sheets, foamy macrophages and lymphocytes against a myxomatous background. A possibility of pleomorphic adenoma was considered and excision biopsy was advised. With this report the patient attended the Surgical Outpatient Department at Osmania General Hospital (OGH).

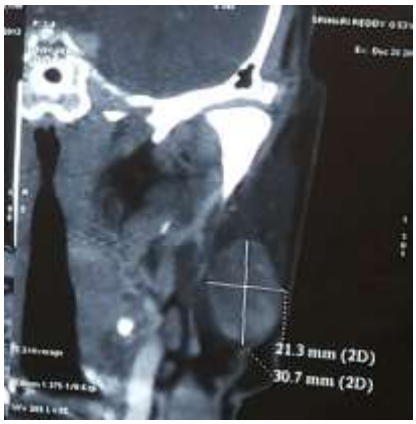

Fig. 1: CT Imageology showing a heterogeneously enhancing soft tissue mass in the left parotid

A left superficial parotidectomy was done taking care to preserve the facial nerve and the

specimen was sent in formalin to Department of Pathology, Osmania General Hospital. Grossly we received two grey white to grey yellow irregular soft tissue masses, larger measuring 4x4x2 cms and the smaller measuring 3x2x1 cm. Cut section was grey white with a nodular area.

H& E stained histology slides revealed a hypercellular tumor with epithelial component represented by cells in sheets and tubules intermingled with chondriod and myxoid areas. Adjacent to it are seen extensive schwannoma like areas. These areas are composed of elongated spindle shaped tumor cells with slender elongated hyperchromatic nuclei arranged in palisades forming Antoni A type areas. A focus of normal salivary gland tissue is also seen.

Immunohistochemistry demonstrated the expression of p63 and CD10 confirming the myoepithelial origin of the cells. Further S-100 positivity confirmed the neurogenic nature of the spindle component. A final histopathological diagnosis of Schwannoma-like pleomorphic adenoma of the left parotid gland was offered.

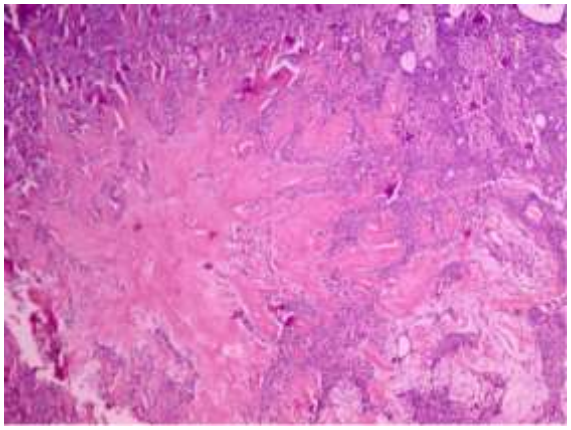

Fig-2: H&E ,10X Showing classical chondroid and myxoid areas with inteveining myoepithelial cells

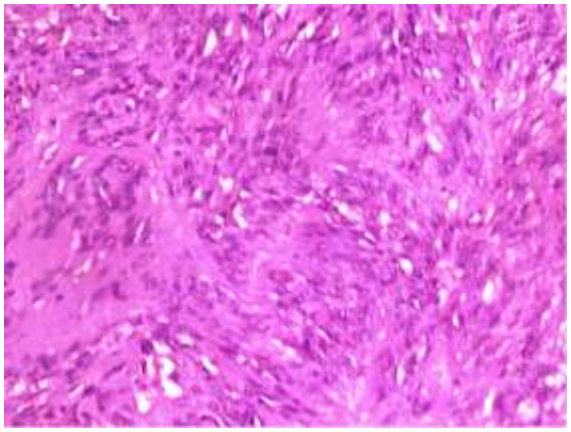

Fig-4: H&E,40X showing palisading Antoni's Type A areas as in schwannoma

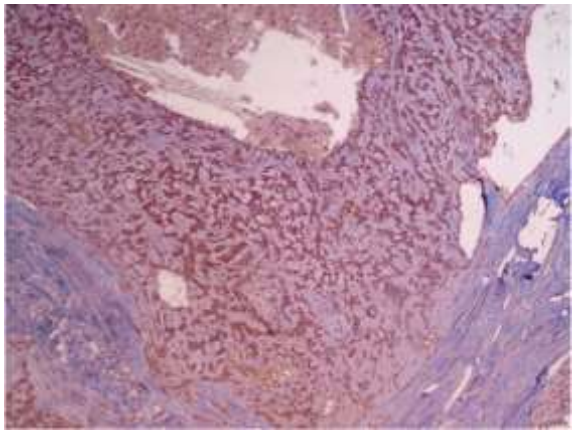

Fig-5: Immunoreactivity for p63 in Schwannian like areas and adjacent chondroid seen, 10X

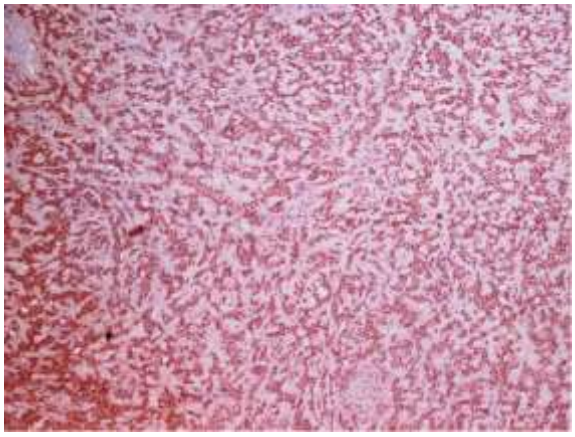

Fig-6: Immunoreactivity for S-100, 10X

DISCUSSION

Pleomorphic adenoma (PA) is the most common salivary gland neoplasm. About 80% of pleomorphic adenomas arise in the parotid, 10% in the submandibular gland and 10% in the minor salivary glands [1]. PA usually appears as a solitary, slowly growing, painless rubbery mass [3]. It is usually solitary but may show synchronous or metachronous association with other tumors [1].

Histological diversities are the hallmark of PA as its name implies. The epithelial, myoepithelial and mesenchymal components are found in close apposition, with the latter being represented by mucoid, myxoid or chondroid material. The proportion between epithelial and chondromyxoid elements are variable.

The epithelial component originating from the duct epithelial-myoepithelial cells shows a wide variety of cell types including cuboidal, basiloid, squamous, spindle, plasmacytoid and clear cells. The epithelium usually forms sheets or duct like structures [1].

Modified myoepithelial cells are thought to play an important role in the histopathological changes of the stroma . Modified myoepithelial cells in pleomorphic adenoma form interlacing fascicles, mostly composed of spindle cells mimicking neurogenic, myogenic or fibroblastic differentiation [2, 4]. A very distinctive appearance is seen when the myoepithelial cells are plasmacytoid or oncocytic [1]. Very rarely spindle cells showing nuclear palisading as in palisading leiomyoma or Antoni's A type neurilemmoma [2, 4].

The stromal component of these tumours is most often predominantly myxoid with focal chondroid or fibrous areas. Sometimes squamous or osseous metaplasia is found in pleomorphic adenoma but is encountered less frequently [5].

Here, ours is a case of pleomorphic adenoma with extrensive schwannoma like areas constituting about 50% of the tumor.The modified myoepithelial cells represented by spindle cells showing nuclear palisading showed immunoreactivity with p63, cd10 and S-100 further confirming the neurogenic nature of the myoepithelial cells.

Literature search revealed only 7 cases of pleomorphic adenoma with schwannoma like areas, ours being the 8th .Of the 7 cases 6 were females and only one male. Ours is the second case of a male patient. Age ranged between 39 to 75 years. All the lesions except one were localized in the parotid gland. One was rarely localized in the hard palate.

Authors	Age	Sex	Location
Marino *et al*. (1977) [8]	74	Female	parotid
	39	Female	parotid
Takeda *et al*. (1999) [2]	62	Male	Parotid
	48	Female	Hard palate
Kajor *et al*. (2006) [6]	75	Female	Parotid
JC Tille *et al*. (2011) [5]	47	Female	Parotid
M Lombardi *et al*. (2013) [14]	45	Female	Parotid
Present case (2014)	**53**	**Male**	**parotid**

Due to the diverse architectural pattern and abundance of spindle cell component, a spindle cell tumour of the salivary glands has to be ruled out. Spindle cell tumours are rare in the salivary glands, representing 1.9% to 5% of parotid neoplasms. Differential diagnoses of benign spindle cell tumours in salivary glands include neurogenic tumours: schwannoma, composed of areas with Verocay bodies and neurofibroma, as well as smooth muscle proliferations: leiomyoma [5] .

Pleomorphic adenomas typically exhibit high signal intensity on T2-weighted images, lack of homogeneity, and sharp demarcation from the adjacent parotid gland. These findings are similar to those of intraparotid facial nerve schwannoma; however, if the tumor has a growth toward the facial canal, facial nerve schwannoma may be reasonably suspected [7].

Schwannoma-like pleomorphic adenomas are composed of modified myoepithelial cells expressing p63, CD10 and S-100 by IHC. Merino *et al*. [8] demonstrated the presence of desmosomes in these spindle-shaped cells by electron microscopy confirming a myoepithelial origin. The pathogenesis of this phenomenon could be a result of plasticity of myoepithelial cells.

FNAC which is considered as a first line investigation in pre operative diagnosis of salivary gland tumors may be misleading in cases with abundant spindle cell component. Immunocytochemistry can come to rescue in such instances in confirming the myoepithelial nature of the cells.

CONCLUSION

True to its nomenclature pleomorhic adenoma exhibits marked diversity in morphology. Therefore it is essential for a pathologist to be aware of all the possible differentiations and metaplasias that can occur in the tumor in order to avoid misdiagnosis.

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