Case Report

Pleomorphic Adenoma mimicking Schwannoma-Like phenotype: A Case Report

Sajda Khan Gajdhar[1], Shaiq Gajdhar[2], Syed Mukith[1], Sabiha Mokashi Khan[3]

[1]Department of Oral and Maxillofacial Pathology, Ibn Sina National College for Medical College for Medical Studies, Jeddah, Kingdom of Saudi Arabia (KSA), [2]Department of Oral and Maxillofacial Rehabilitation, Ibn Sina National College for Medical College for Medical Studies, Jeddah, Kingdom of Saudi Arabia (KSA), [3]Medical Officer-Indian Cancer Society, Parel, Mumbai, India

ABSTRACT:

The schwannoma-like pleomorphic adenoma is a rare histopathological variant of the pleomorphic adenoma. It can occur in any salivary gland; however it is most commonly detected in the parotid. Pleomorphic adenomas are well-known for their cytomorphological and architectural variability, which is characterised by the presence of intermixed epithelial and mesenchymal-like components. We present a rare case of parotid gland pleomorphic adenoma with prominent schwannoma-like characteristics that mimicked a benign schwannoma. The tumour had a high prevalence (about 80%) of schwannoma-like regions with a localised (approximately 20%) epithelial component along with tubular organisation. Only 8 cases of schwannoma-like pleomorphic adenoma have been documented in the literature, to the best of our knowledge. The differential diagnosis between this entity and neurogenic and myogenic tumors is discussed.

KEYWORDS: Salivary gland tumor, Schwannoma-like pleomorphic adenoma, Parotid, Pleomorphic adenoma, Mixed tumors

Address for correspondence : Dr Sajda Khan Gajdhar, Lecturer, Department of Maxillofacial Pathology, Ibn Sina National College for Medical College for Medical Studies, Jeddah, Kingdom of Saudi Arabia (KSA), E-mail: drsajdagajdhar@gmail.com
Submitted: 16.09.2023, **Accepted:** 30.11.2023, **Published:** 26.12.2023

INTRODUCTION:

Salivary gland tumors account for about 3% of all head and neck neoplasia. The parotid gland is the main site for these tumors and about 95% of them are of epithelial origin[1]. Pleomorphic adenoma (PA) is the most common benign salivary gland tumor. The parotid gland is the main site of origin of this lesion, but minor salivary glands can also be affected.[1] PA represents about 66 % of these benign lesions. Although a large majority (95%) of PAs are benign, a small fraction may show an aggressive behaviour (carcinoma ex pleomorphic adenoma). This may occur after multiple local recurrences and/or radiotherapy[1-2]. Conventional PAs are characterized by a biphasic appearance, resulting from the intimate admixture of epithelium and mesenchymal-like components with low mitotic activity, lacking features of cellular atypia and nuclear hyperchromasia. Frequently a matrixrich in glycosaminoglycans is present. The proportion of benign versus malignant tumor varies greatly by site. In the major salivary glands, such as the parotid and the submandibular gland, the majority of the tumors are benign contrary to the minor salivary glands, such as the sublingual and the floor of the mouth, where most of them are malignant[1-2]. We present a rare case of parotid gland PA with prominent schwannoma-like characteristics.

CASE REPORT:

A 41-year-old woman presented for a

Access this article online

Quick Response Code:

Website: www.pjsr.org

DOI: doi.org/10.5281/zenodo.10432040

How to cite this article: Gajdhar SK, Gajdhar S, Mukith S, Khan SM. Pleomorphic Adenoma mimicking Schwannoma-Like phenotype: A Case Report. PJSR. 2023;16(2):22-25.

persistent asymptomatic swelling localized in the right retromandibular area since 8 months. A well-defined tumour measuring 1.8 cm, was found in the deep lobe of the right parotid gland, with parapharyngeal extension, according to a head and neck MRI. The radiologic findings pointed to the diagnosis of PA. The lesion was surgically removed and histopathology was performed. Macroscopically the tumor appeared as 1.8 cm brownish circumscribed nodule. The lesion was encapsulated and hypercellular under the microscope, and at greater magnification, the tumour showed high aggregates (about 80%) of schwannoma-like regions (Figure 1). The schwannoma-like areas showed tumor cells with spindle type morphology, small elongated and hyperchromatic nuclei arranged in palisades, with broad areas of Antoni A type (Figure 2). The sparse epithelial area was organised in short tubular structures and contained a few duct-like areas, sheets, and cords of tumour cells. There were also eosinophilic, acellular, hyalinized regions surrounded by cords or sheets of tumour cells (Figure 3). The overall features were suggestive of schwannoma-like pleomorphic adenoma. A regular follow-up showed no recurrence.

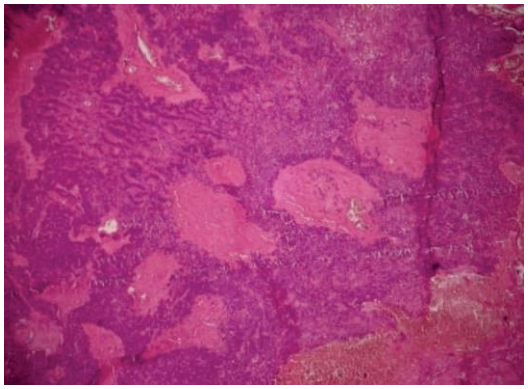

Figure 1: Low magnification of schwannoma-like pleomorphic adenoma with both components (hematoxylin-eosin, magnification 4X).

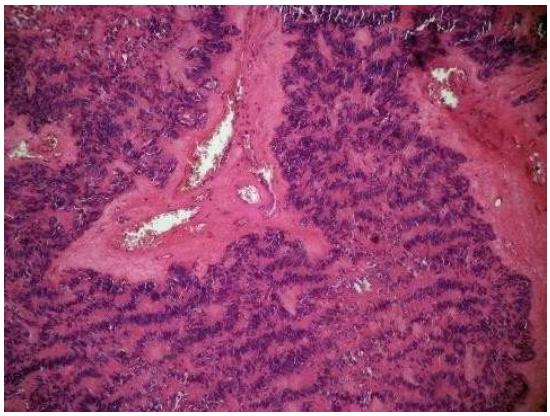

Figure 2: The tumor is composed by spindle-shaped cells with elongated nuclei and ill- defined cell contours, with nuclear palisading resembling Antoni-A areas of schwannoma (hematoxylin and eosin, 10X magnification).

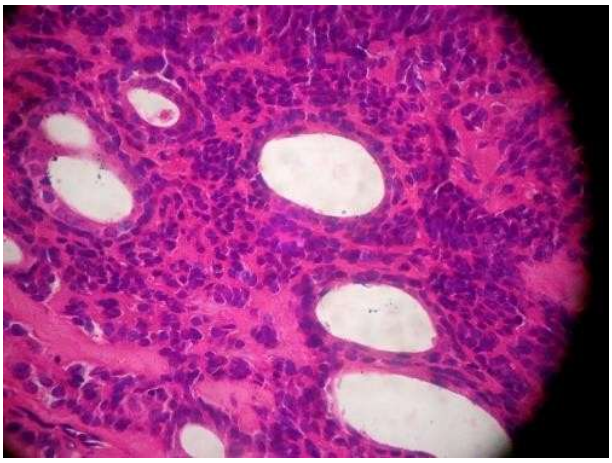

Figure 3: Under high power view the duct like areas are surrounded by double row of cells and hyaline-like collagen (hematoxylin and eosin, 40X magnification).

DISCUSSION:

PAs are well-known for their cytomorphological and architectural diversity. These tumours are made up of both epithelial and stromal components. Although PAs are usually neatly contained, tiny expansions protrude into the adjacent normal tissue in a pseudo-infiltrative pattern. The epithelial cells are derived from ductal epithelial and myoepithelial cells[1-3]. Tumour cells in the epithelial layer are cuboidal in shape and line duct-like structures that vary in size and shape. Tumour cells of myoepithelial origin (MMCs) contain polygonal, spindle, or plasma cell-like shapes, form sheet, clump, or strand-like structures, and are admixed with a myxoid or myxo-chondroid component[1-3]. MMCs in PA occasionally form interlacing fascicles, which are usually composed of spindle-shaped cells reflecting neurogenic or myogenic differentiation[1]. Rarely, small areas of MMCs may exhibit schwannoma-like palisading of nuclei, however this is usually only a localised aspect of the overall tumour mass[3]. Our case showed features of PA with widespread schwannoma-like characteristics extending in almost the whole lesion. Schwannoma-like PAs are composed of modified myoepithelial cells[4-5]. PA resembling schwannoma is an extremely unusual lesion; only 8 occurrences of this nosological entity have been documented in the literature (Table 1)[3-11]. There is a strong predilection for females. 6 of the cases included females and 2 involved males, with ages ranging from 39 to 90 years (mean age 64.5). Our case was seen in female. The lesions were primarily found in the parotid gland (n=6), with the exception of 1 unusual case found in the hard palate and 1 case in submandibular gland. Our case also involved parotid gland. The lesions ranged in size from 1.5 to 3.5 cm

Table: 1 Previously reported case of schwannoma-like pleomorphic adenoma.

S.No.	Authors	Age	Gender	Site	Duration	Size(cm)	Symptoms
1	Merino and Livolsi	74	F	Parotid gland	5 years	2X3	Asymptomatic
2	Takeda and Shimono	62	M	Hard palate	10 months	NA	Asymptomatic
3	Kajor et al.	75	F	Parotid gland	NA	3	Asymptomatic
4	Tille et al.	47	F	Parotid gland	2months	3.2	Pain
5	Lombardi et al.	44	F	Parotid gland	3 months	1.5	NA
6	Chandra et al.	60	M	Parotid gland	12 years	3.5	Asymptomatic
7	Roth SH et al	90	F	Submandibular gland	NA	3.22	Asymptomatic
8	Roth SH et al	40	F	Parotid gland	1.5 years	2	Asymptomatic
9	Case	41	F	Parotid gland	8 months	1.8	Asymptomatic

(mean value 2.6 cm)[3-11]. Areas of spindle cells grouped in palisades resembling a benign schwannoma were noted in all of the reported cases. Fine-needle aspiration biopsy (FNAB) is a widely used diagnostic technique. However, in the presence of lesions with prevalent schwannoma-like architecture this method may be inadequate to distinguish among PA and neurogenic or myogenic tumors[4-7]. Schwannomas associated with cranial and peripheral nerves have been reported within salivary glands and there are published cases that have been initially misdiagnosed as schwannoma-like pleomorphic adenoma[7]. In addition, a glandular variant of schwannoma has been also reported in the literature[8]. This lesion was characterized by entrapped or metaplastic glands in an otherwise pure schwannoma. This extremely rare schwannoma type can add to the uncertainty in the differential diagnosis of schwannoma-like pleomorphic adenoma. An additional technique, such as immunohistochemistry, can be used to alleviate this diagnostic problem: a positive staining for cytokeratin in both the mesenchymal-like and epithelial portions allows the diagnosis of schwannoma-like pleomorphic adenoma[1].

In conclusion schwannoma-like pleomorphic adenoma is a rare variant of PA that, although biologically benign, deserves to be considered in the differential diagnosis with neurogenic and myogenic tumors of the parotid gland.

CONCLUSION:

Schwannoma-like pleomorphic adenoma is a rare entity that is mostly seen in females and affects parotid gland. This entity should be distinguished from the other neurogenic and myogenic tumors. Prognosis of this lesion is good.

Financial Support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES:

1. Ellis GL. What's new in the AFIP fascicle on salivary gland tumors: a few highlights from the 4th Series Atlas. Head Neck Pathol. 2009 Sep;3(3):225-30. doi: 10.1007/s12105-009-0128-z. Epub 2009 Jul 21. PMID: 20596976; PMCID: PMC2811626.
2. Coli A, Bigotti G, Bartolazzi A. Malignant oncocytoma of major salivary glands. Report of a post-irradiation case. J Exp Clin Cancer Res. 1998 Mar;17(1):65-70. PMID: 9646235.
3 Kajor M, Gierek T, Markowski J, Pajak J. Schwannomalike mixed tumor of the parotid gland: a case report. Acta Cytol. 2006 Sep-Oct;50(5):529-30. doi: 10.1159/000326007. PMID: 17017438.
4. Tille JC, Reychler H, Hamoir M, Schmitz S, Weynand B. Schwannoma-like pleomorphic adenoma of the parotid. Rare Tumors. 2011 Oct 21;3(4):e40. doi: 10.4081/rt.2011.e40. Epub 2011 Sep 16. PMID: 22355495; PMCID: PMC3282445.
5. Merino MJ, LiVolsi VA. Pleomorphic adenomas of the parotid gland resembling mesenchymal tumors. Oral Surg Oral Med Oral Pathol. 1977 Sep;44(3):405-10. doi: 10.1016/0030-4220(77)90410-8. PMID: 198719.
6. Kapila K, Mathur S, Verma K. Schwannomas: a pitfall in the diagnosis of pleomorphic adenomas on fine-needle aspiration cytology. Diagn Cytopathol. 2002 Jul;27(1):53-9. doi: 10.1002/dc.10121. PMID:

12112817.
7. Kim YC, Park HJ, Cinn YW, Vandersteen DP. Benign glandular schwannoma. Br J Dermatol. 2001 Nov;145(5):834-7. doi: 10.1046/j.1365-2133.2001.04476.x. PMID: 11736913.
8. Lombardi M, Socciarelli F, Fini G, Leonardi A, Bartolazzi A. Schwannoma-like pleomorphic adenoma: a case report with review of the literature. Head Neck Pathol. 2014 Jun;8(2):178-81. doi: 10.1007/s12105-013-0473-9. Epub 2013 Jul 10. PMID: 23838857; PMCID: PMC4022934.
9. Chandra SR, Karim F, Rawal YB. Divergent Schwannoma-Like Phenotype in a Pleomorphic Adenoma. Head Neck Pathol. 2017 Dec;11(4):567-574. doi: 10.1007/s12105-017-0817-y. Epub 2017 Apr 24. PMID: 28439689; PMCID: PMC5677055.
10. Roth SH, Faquin WC, Gimenez C, Vadalia B, Frank DK, Li JY. Schwannoma-Like Pleomorphic Adenoma: Two Cases and a Review of the Literature. Head Neck Pathol. 2020 Mar;14(1):166-172. doi: 10.1007/s12105-019-01036-y. Epub 2019 Apr 9. PMID: 30968284; PMCID: PMC7021839.