Case Report

Cystic Pleomorphic Adenoma: A Rare Occurrence and a Diagnostic Pitfall

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Abstract

Background: Cystic lesions of the salivary glands are a diagnostic dilemma. Pleomorphic adenoma, the most common neoplasm of the salivary gland, with cyst formation, is a rare event and can mimic various benign and malignant lesions.

Case report: Here, we report an interesting case of a 25-year-old male with cystic degeneration of pleomorphic adenoma.

Conclusion: Cyst formation in the salivary gland is not only seen in several cyst-associated lesions but can also be seen rarely in cyst-unassociated lesions like pleomorphic adenoma. Appropriate cytodiagnosis and optimal management of such cystic lesions require the application of the Milan System for Reporting Salivary Gland Cytopathology (MSRSGC).

Keywords:

Salivary gland cysts; Cystic pleomorphic adenoma; Diagnostic dilemma; The Milan System

Introduction

Cystic lesions of the salivary glands can be due to neoplastic and non-neoplastic pathology and account for almost 8% of salivary gland lesions [1]. Clinically, a broad spectrum of salivary gland lesions are of cystic nature [2].

Different imaging techniques can be used to assess salivary gland cystic lesions; however, the radiological features are nonspecific, and there is overlap between benign and malignant lesions. Additionally, the complex anatomy of the site, with the presence of non-salivary structures, makes interpretation a difficult task in radiology.

Fine-needle aspiration cytology (FNAC) is a well-recognized, minimally invasive procedure for the evaluation of salivary gland mass lesions. It is simple, cost-effective, rapid, and associated with low risk. It is highly specific, although relatively less sensitive, for the detection of neoplastic conditions in the salivary glands [3]. The cytology of salivary glands presents some diagnostic

difficulties due to heterogeneity, metaplastic changes, and morphological overlap between various lesions [4]. Although FNAC is a tool that accurately diagnoses pleomorphic adenoma, cystic degeneration or squamous metaplasia creates a diagnostic dilemma. The diagnosis of cystic lesions is tougher due to low cellularity with a nonspecific watery or mucoid background.

To address such difficulties, the effort for the Milan System for Reporting Salivary Gland Cytopathology (MSRSGC) was initiated in 2015 in Milan, and the atlas was published in 2018. The atlas consists of six diagnostic categories: Category I – "Non-Diagnostic," Category II – "Non-Neoplastic," Category III – "Atypia of Undetermined Significance (AUS)," Category IV A – "Neoplasm: Benign," Category IV B – "Neoplasm: Salivary Gland Neoplasm of Uncertain Malignant Potential (SUMP)," Category V – "Suspicious for Malignancy," and Category VI – "Malignant." MSRSGC is a risk-stratification six-tier classification scheme in which the risk of malignancy and appropriate management have been defined for each diagnostic category of salivary gland fine-needle aspiration cytology [5].

After surgical resection, histopathological examination (HPE) has the advantage of providing the whole tumor for evaluation, thereby eliminating issues related to morphological diversity. Almost 73% of salivary gland tumors arise in the parotid gland, of which around 63% are pleomorphic adenomas [6]. Pleomorphic adenomas (PA), the most common tumors of the salivary glands, are mass lesions characterized by a triphasic growth pattern of epithelial, myoepithelial, and stromal components [7].

Here, we report a case of benign pleomorphic adenoma of the parotid gland with cyst formation in a 25-year-old male patient and highlight the difficulty of making a correct diagnosis in such cystic lesions.

Case Report

A 25-year-old male presented with a gradually increasing swelling on the left side of his face (parotid region), associated with intermittent pain for about five months. On local examination, a 4 × 3 cm firm-to-hard swelling was seen. The overlying skin was non-tender and normal-looking. There was no suspicion of the swelling being cystic on clinical examination.

On a non-contrast computed tomography scan (NCCT), a well-defined, rounded, hypo-dense lesion in the superficial lobe of the left parotid gland, measuring approximately 3 × 2.2 × 2.6 cm, was seen. On performing magnetic resonance imaging (MRI), a well-marginated lesion measuring 2.8 × 2.2 × 2.0 cm was identified in the superficial lobe of the left parotid gland. A suggestion of a lesion likely to be pleomorphic adenoma or Warthin's tumor was given on radiology, and FNAC correlation was advised.

FNAC was inconclusive as the smears were paucicellular with a nonspecific, mucoid, and hemorrhagic background, thus falling under Category III of the Milan system, for which the management is repeat FNA or surgery. In due course, a superficial parotidectomy was performed, and the sample was sent for histopathology.

On gross examination, a grey-brown, irregular, soft tissue mass measuring about 4 × 3.5 × 3 cm was seen. On the cut section, a well-circumscribed lesion with a growth and a large cystic cavity having a smooth lining and partially filled with mucus-like material, altered hemorrhage, and necrotic material were seen (see Figure 1).

Histopathology from the solid cystic lesion showed hemosiderin-laden macrophages, eosinophilic granular necrotic material, and mucus from the cystic portion (see Figure 2a) and a triphasic growth pattern of epithelial, myoepithelial, and stromal components from the solid portion (see Figure 2b). Initial sections did not show any chondroid areas; however, deeper sections were valuable as they showed chondroid areas (see Figure 3a). Occasional areas of squamous metaplasia were also noticed (see Figure 3b). Unremarkable salivary gland tissue was identified surrounding the lesion.

A diagnosis of pleomorphic adenoma with cystic degeneration was made on histopathology, and the lesion healed uneventfully after surgery.

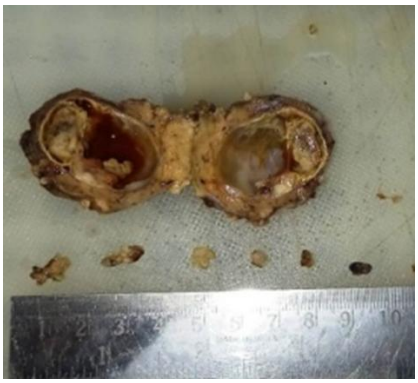

Figure 1: Cut section through the grey brown tissue mass revealed a well-circumscribed lesion with a growth and a large cystic cavity having smooth lining and partially filled with mucus-like material, altered hemorrhage and necrotic material.

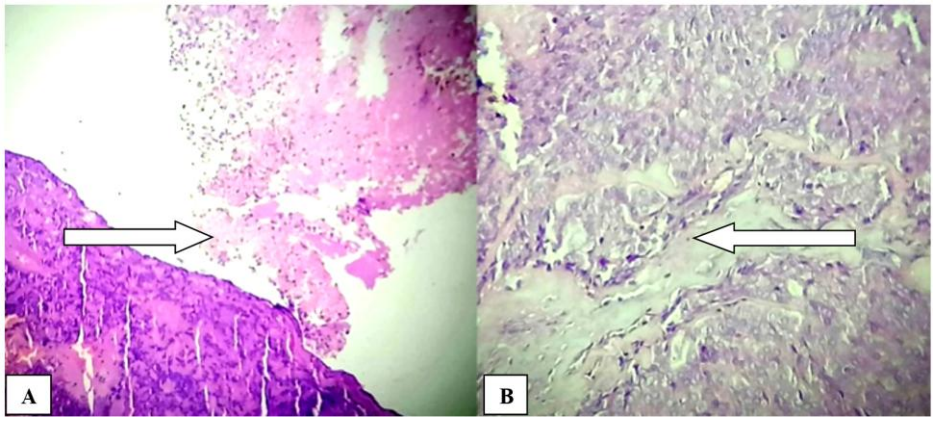

Figure 2: Histopathological sections with Hematoxylin and Eosin (H&E, 10 x) staining showing hemosiderin-laden macrophages, eosinophilic granular necrotic material and mucus from the cystic portion (A) and epithelial cells, myoepithelial cells and myxoid stroma from the solid portion (B).

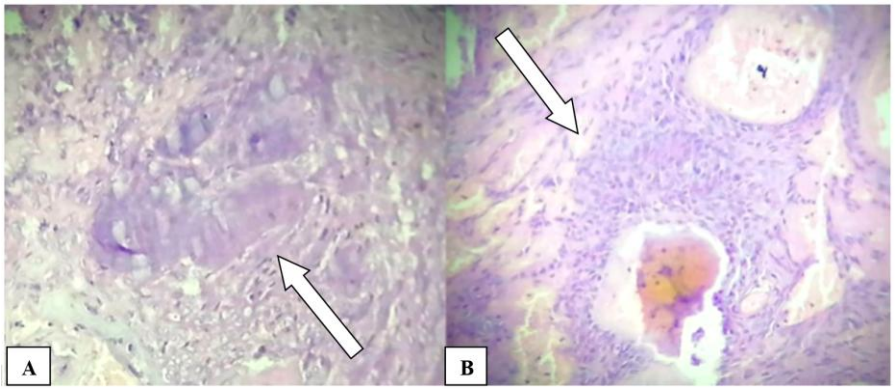

Figure 3: Histopathological sections with Hematoxylin and Eosin (H&E, 40 x) staining from deeper sections showing chondroid areas (A) and occasional areas of squamous metaplasia (B).

Discussion

Pleomorphic adenomas are benign tumours that may rarely present with cystic degeneration and squamous metaplasia, making the cytodiagnosis a problem, especially in the absence of chondromyxoid stroma, as was seen in the present case [6].

Clinically, a number of benign and malignant neoplasms, such as pleomorphic adenoma, Warthin's tumor, mucoepidermoid carcinoma, acinic cell carcinoma, and lesions such as mucocele, may be cystic [8].

In addition to careful clinical evaluation, imaging modalities like ultrasonography (USG), computed tomography (CT) scan, and magnetic resonance imaging (MRI) help determine whether the lesion is intra-glandular or extra-glandular, whether there is any local invasion and extension or perineural spread (a common feature of adenoid cystic carcinoma), and if there is any nodal metastasis and systemic involvement. USG should be the investigation of first choice since it is a quick and economical technique with the advantage of a thorough assessment of the salivary gland without exposing the patient to radiation.

FNAC can be used along with imaging for safe and accurate sampling of the lesion. The advantages of FNAC are many, as it is an economical, fast technique that helps the clinician understand the nature of the lesion, decide the appropriate treatment strategy, and provide proper patient counseling.

However, the cytological diagnosis has some challenges: FNAC only samples a small portion of the lesion. False-positive diagnosis due to reactive changes associated with inflammation or false-negative diagnosis of hypo-cellular cystic lesions and low-grade tumors (due to their bland cytological features) can occur at times. Similarly, the less common high-grade malignancies, although readily recognizable as malignant, are difficult to distinguish from one another. Some benign tumours (e.g., basal cell adenoma) have a malignant counterpart (e.g., basal cell adenocarcinoma) that is morphologically identical except that it has an infiltrative growth pattern—something that cannot be assessed cytologically. Similarly, differentiating mucus-containing cystic lesions from mucoepidermoid carcinoma or mucocele is also difficult on cytology. Oncocytic lesions, clear cell neoplasms, and spindle cell lesions are other diagnostic dilemmas in salivary gland cytology [9].

The Milan system aimed at standardizing the reporting terminology so that the conventional descriptive interpretation of salivary gland FNAC could be replaced, achieving better communication between clinicians and institutions. The Milan system is useful for making clinical decisions by providing the risk of malignancy (ROM) and is useful for lab quality control by providing the frequency of each diagnostic category. The MSRSGC consists of six diagnostic categories, and each category has certain diagnostic criteria, ROM, incidence frequency range, and management protocol. Therefore, this guides cytologists in reporting salivary gland FNAC, as well as surgeons in determining the most appropriate treatment strategy [10].

Cystic change in pleomorphic adenomas is a diagnostic dilemma and can mimic mucoepidermoid carcinoma, mucocele, carcinoma ex pleomorphic adenoma, and squamous cell carcinoma [8]. Cytology reporting and management of such lesions should also be done as per the criteria in the Milan system. Although the usefulness of the Milan system for salivary gland lesions as a whole is well established, recent studies, such as that by Maleki et al. [11] on the application of the Milan system to cystic lesions of the salivary glands, have shown that the Milan system is also beneficial in the case of cystic salivary gland lesions. In the present case of Milan category III, surgical resection was thus performed, which proved to be useful.

After reviewing the literature, we found that cyst formation in the most common salivary gland tumour—pleomorphic adenoma—is a very rare event, and only a few studies have reported such cystic pleomorphic adenomas [8, 11, 12]. In this article, we have thus reported such a case of cystic salivary gland pleomorphic adenoma that was a diagnostic dilemma on cytology and discussed

the cytological challenges and the importance of the Milan system in reporting such cystic lesions.

Conclusion

Cyst formation in salivary glands is seen in several benign and malignant cyst-associated and, rarely, non-associated lesions like pleomorphic adenomas. The diagnosis of cystic lesions on cytology is difficult because of low cellularity with a nonspecific watery or mucoid background. Application of MSRSGC is required for appropriate cytodiagnosis and optimal management of such cystic salivary gland lesions.

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