



CARCINOMA EX PLEOMORPHIC ADENOMA: A RARE ENTITY TO BE DIAGNOSED ON CYTOLOGY

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ABSTRACT A 65 year old male patient presented with a painless swelling in front of the left ear for last 20 years. It started initially as a painless nodule and increased gradually to the present size. Patient also gave history of a rapid increase in size during the last 2 months associated with mild pain. FNAC of the swelling was done and a diagnosis of carcinoma ex pleomorphic adenoma was rendered.

KEYWORDS : Carcinoma ex pleomorphic adenoma, parotid gland, salivary gland carcinoma

INTRODUCTION

Carcinoma ex pleomorphic adenoma is defined as a carcinoma arising from epithelial or myoepithelial or both the components of a primary or recurrent benign PA [1]. Malignant changes in PA have been associated with long duration, tumor size, tumor recurrence, radiation therapy and advanced age [2]. In majority of the cases (75%), luminal epithelial cells undergo malignant transformation. CXPA more commonly occurs in the major salivary glands than in the minor salivary glands. Further in major salivary glands, parotid (67%) is the most frequently affected gland and sublingual is the least affected one (<1%). CXPA has the highest false-negative rate of 35.3% of all malignant salivary gland tumor[3]. Fine needle aspiration cytology (FNAC) has been a widely accepted diagnostic tool for palpable salivary gland lesions. Diagnostic accuracy of FNAC of salivary gland lesions ranges from 80-95% in adequately sampled specimens [4, 5]. One of the entities rarely diagnosed on cytology is carcinoma ex pleomorphic adenoma [5]. We present a case report of this uncommon entity diagnosed on cytology.

CASE HISTORY

A 65 year old male patient presented with a painless swelling in front of the left ear for last 20 years. It started initially as a painless nodule and increased gradually to the present size. Patient also gave history of a rapid increase in size during the last 2 months associated with mild pain. There was no skin discoloration, ear discharge or fever. General and systemic examinations were within normal limits.

Local examination revealed a 2.5cm x 2.5cm firm to hard, non mobile and non tender swelling in parotid region. Signs of inflammation were absent. FNAC was performed with a 22 gauge needle. Smears were prepared, fixed and stained with hematoxylin and eosin stain.

FNAC FINDINGS

Microscopic Examination of the smears revealed highly cellular smears showing mixed population of benign and malignant cells. Sheets and clusters of pleomorphic cells having round to spindle cells and plasmacytoid cells were seen (Fig.1). These cells were showing cytological atypia in the form of high nucleocytoplasmic ratio, irregular nuclear membrane, hyperchromatic nucleus and abundant eosinophilic cytoplasm (Fig.2). The benign epithelial component appeared as small round to oval cells with bland nuclear features. Malignant changes occurring in a background of pleomorphic adenoma was suggested based on the above mentioned cytological features.

DISCUSSION

Carcinoma ex pleomorphic adenomas (Ca ex PA) are a diverse group of malignant tumors arising from pleomorphic adenomas and one of the rare entities to be diagnosed on cytology. The most common clinical presentation of carcinoma ex pleomorphic adenoma is a firm

mass in the parotid gland. Ca ex PA accounts for approximately 3.6% of all salivary gland neoplasms and 11.6% of all malignant neoplasms and typically develops in the sixth and seventh decades of life.[6] Patients become aware of the cancer only after experiencing sudden enlargement. Facial pain, paralysis, and dental pain may occur in tumors with local extension into surrounding structures including soft tissues, facial nerve, and jaw respectively. Increased preoperative duration of pleomorphic adenoma increases risk of malignant transformation with a mean lead up time before malignant transformation of 9 years.[7]. The possibility of malignant transformation in a long standing pleomorphic adenoma increases from 1.6% in < 5 years to 9.5% in a tumor present for 15 years.[8] The common malignancies occurring in a background of pleomorphic adenoma are adenocarcinoma not otherwise specified (42.4%) and salivary duct carcinoma[9]. The other uncommon malignancies that can arise in a setting of pleomorphic adenoma are adenosquamous carcinoma, adenoid cystic carcinoma, undifferentiated carcinoma, myoepithelial carcinoma, epithelial – myoepithelial carcinoma and sarcomatoid carcinoma.

CONCLUSION

A case reported here is a rare carcinoma ex pleomorphic adenoma of the salivary gland. It is a dreadful disease affecting salivary glands and can cause death if not diagnosed early. It is difficult to diagnose this rare entity on cytology. It has a high false negative rate of 35.3% on FNAC. In our case, since we made multiple needle passes from different areas of the swelling, we were able to sample the areas of malignant transformation. This rare case report leads emphasis on multiple areas to be sampled in salivary gland lesions so that this entity is not missed out on FNAC and an early diagnosis can be rendered.

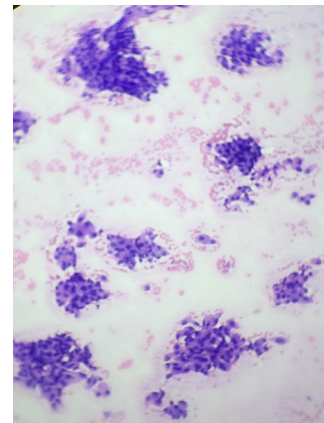


Fig. 1

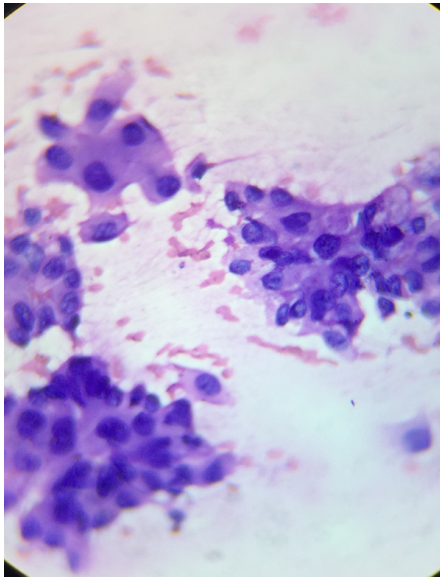


Fig. 2

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