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Pathology

CASE REPORT ON RARE TUMOR- CARCINOMA EX PLEOMORPHIC ADENOMA OF PAROTID GLAND

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ABSTRACT Carcinoma ex pleomorphic adenoma is a rare tumor of salivary gland. CXPA is a malignant transformation of a primary or recurrent PA. It is aggressive tumor with poor prognosis. The diagnosis of this tumor is problematic for the pathologosist and clinician because of component of residual benign pleomorphic adenoma. It may be invasive and non invasive tumor. Accurate diagnosis is important for management and the most important prognostic factor is histopathological classification of tumor. We describe a rare case report of 54 year old male with CXPA of parotid gland. The patient had previously undergone two surgeries for pleomorphic adenoma.

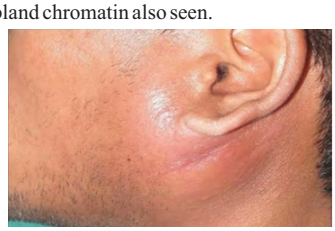
KEYWORDS : Carcinoma ex pleomorphic adenoma , parotid gland

INTRODUCTION

Pleomorphic adenoma (PA) is the most common neoplasm of salivary glands and may sometimes undergo malignant transformation in its natural course in about 2-25% of cases. There are three subtypes of malignant pleomorphic adenoma. Carcinoma ex pleomorphic adenoma , carcinosarcoma and metastasizing Pleomorphic adenoma. Carcinoma ex pleomorphic adenoma (CXPA) is considered to be a malignant transformation product of a pre-existing pleomorphic adenoma [1]. Ca ex PA accounts for approximately 3.6% of all salivary gland neoplasms and typically develops in the sixth and seventh decades of life.[2] Carcinoma ex pleomorphic adenoma was first described by Beahrs et al. in 1957. . The most common clinical presentation of carcinoma ex pleomorphic adenoma is a firm mass in the parotid gland. Patients become aware of the cancer only after experiencing sudden enlargement. Facial pain, paralysis, and dental pain may occur in tumor with local extension into surrounding structures including soft tissues, facial nerve, and jaw. Surgery, radiotherapy, chemotherapy is the treatment of choice. We present here a case of Carcinoma ex pleomorphic adenoma affecting a 54 year-old male patient.

CASE REPORT

A 54 year old male reported to our deptartment with chief complain of a smalll swelling in left parotid region for 20 years, the swelling initially small in size and slow growing but there was a sudden enlargement of swelling over 1 month and pain over the swelling from 15 days. On examination-swelling measuring 6*6cm in parotid region , firm- hard, non mobile and non tender and non ulcerated fig 1. No lymph node palpated in cervical region. Oral cavity and oropharynx were within normal limits. USG of head and neck revealed well defined encapsulated mass of mixed echogenicity in left parotid region. Fnac was performed by 22 gauge needle and diagnosis of carcinoma ex pleomorphic adenoma was rendered. Cytological smear show sheets and clusters of pleomorphic cells having round to spindloid cells and plasmacytoid cells .These cells are showing atypical changes in the form of high nucleo-ctytoplasmic ratio, irregular nuclear membrane,hyperchromatic nucleus and abundant eosinophilic cytoplasm. Focal area of pleomorphic adenoma with benign looking nucleus and bland chromatin also seen.



The clinical photograph of the patient exhibiting the huge parotid mass

DISCUSSION

Carcinoma ex pleomorphic adenoma is a tumor in which the neoplasm develops from the epithelial component that fulfils the criteria for malignancy [3]. In most instances, 75% of the luminal epithelial cells undergo malignant change. In 19% of cases, a dual epithelial/myoepithelial differentiated carcinoma is seen. Pure myoepithelial malignant change is seen in only 6% of cases [2]. The scarcity of well documented cases and lack of generally accepted histopathologic criteria has led to poor understanding of this form of salivary gland cancer [3]. To satisfy the definition of CXPA, at least a focus of benign pleomorphic adenoma must be identified or a previous benign pleomorphic adenoma must have been excised from a site in which recurrent tumor is carcinomatous [4]. The proposed criteria for defining carcinoma ex pleomorphic adenoma by Nagao et al. were used to select and reclassify our case of carcinoma ex pleomorphic adenoma. The use of strict pathological criteria may underestimate the frequency of carcinoma ex pleomorphic adenoma because the malignant cells in some cases may obliterate the original pleomorphic adenoma [5].

CXPA usually presents clinically with a history of a slowly growing, painless mass that suddenly or over a short period enlarges rapidly. It presents with signs and symptoms suggesting malignancy (e. g. Fixation to surrounding structures, occasional pain, skin infiltration, trismus, facial nerve weakness, or palsy). Facial nerve weakness or palsy has been detected in approximately 23-40% of cases [6]. In present case patient complain of facial pain and difficulty in mastication. These are clinically aggressive lesions often leading to metastasis and a tumor related death [5]. Beahrs et al. reported the predominance of males whereas Evans and Cruickshank showed that CXPA are more common in the females. Chen HH et al. showed that CXPA is the most common malignant change in PA. Nagao et al showed that 50% of the CXPA of the parotid gland were greater than 5 cm in size. However the tumor size in the present case was large measuring 6*6 cms in dimension and affecting a female patient.

According to the diagnostic criterias proposed by Gerughty et al. [3], we think that our case is a carcinomatous transformation in pleomorphic adenoma. Our case had all the features of carcinomatous transformation along with local metastasis. Immunohistochemistry was believed to be of potential assistance in the diagnosis of salivary gland tumors and in the prediction of histogenesis.

Possible treatments of carcinoma ex pleomorphic adenoma include four different modalities; surgical therapy, radiotherapy, chemotherapy and combined therapy. According to Chen et al., surgery followed by postoperative radiation should be considered the standard of care for patients with CXPA [8]. Gerughty et al. suggested that the histological evidence of an invasive growth pattern, neural or vascular invasion, necrosis and focal calcification implied a poor prognosis [9].

CONCLUSION

This case illustrates the classic presentation of carcinoma ex-pleomorphic adenoma with regards to clinical, pathological, and radiographic diagnosis. This case highlights the diagnostic dilemma of these heterogeneous malignancies. Surgical decision making is largely dependent on pathological and radiographic findings. Carcinoma ex pleomorphic adenoma is a rare and fascinating tumor that requires a multidisciplinary approach for optimal patient outcome. Finally, more research is needed in regards to tumor markers, prognostication and adjuvant therapy in the more aggressive cancer types.