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Carcinosarcoma of parotid gland - A rare case report

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ABSTRACT

Carcinosarcoma (true malignant mixed tumor) is an extremely rare tumor of the salivary gland. It is a biphasic tumor and is composed of both malignant epithelial and malignant mesenchymal component. Commonly, squamous cell carcinoma and adenocarcinoma forms the epithelial component and chondrosarcoma forms the mesenchymal component. We, hereby, report a parotid lesion in a 47 year old female that contained adenocarcinoma as epithelial component and chondrosarcoma as mesenchymal component. We report this unusual case of carcinosarcoma of parotid gland due to its rarity. It mimics benign lesions on ultrasonographic examination. On fine needle aspiration, the diagnosis of carcinosarcoma can be missed, as was in our case where it was reported as pleomorphic adenoma with atypical features. Histopathology only gives confirmatory diagnosis. In addition, a short review of literature along with possible pathogenesis of malignant mixed tumor of salivary gland is also presented.

Keywords: Adenocarcinoma, Carcinosarcoma, Chondrosarcoma, Malignant mixed tumor, Salivary gland

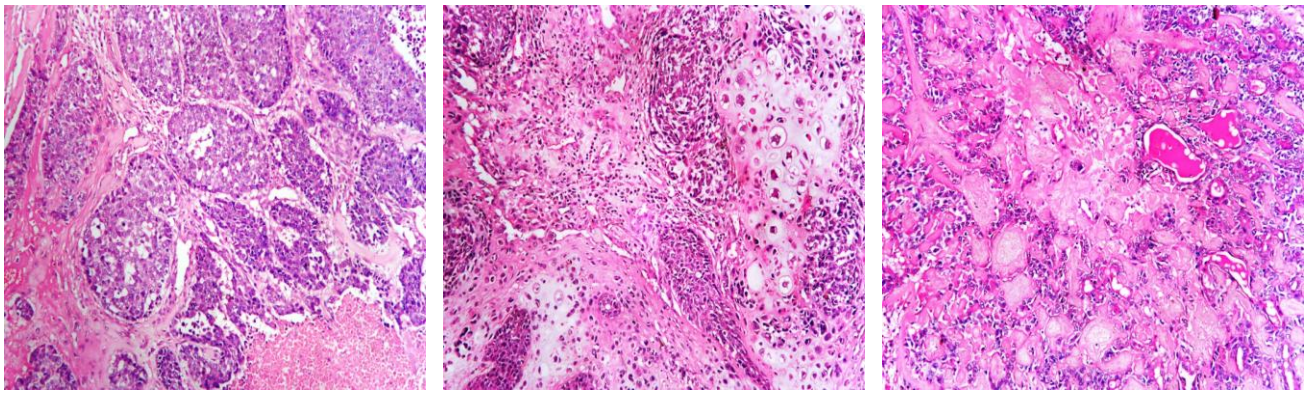
Carcinosarcoma is an extremely rare true malignant mixed tumor of salivary gland comprising 0.04-0.16% of total cases. Sixty five percent of these tumors arise in parotid gland [1]. This heterologous neoplasm is composed of both malignant mesenchymal and epithelial components [2]. It is a high-grade neoplasm with multiple episodes of recurrences and blood borne distant metastasis [3]. The malignant epithelial component is mostly formed by squamous cell and adenocarcinoma. On the other hand, chondrosarcoma forms the most common malignant mesenchymal component followed by fibrosarcoma, leiomyosarcoma, osteosarcoma, and liposarcoma in decreasing order of frequency [1].

We report a case of carcinosarcoma of the parotid gland in 47-year-old woman with adenocarcinoma as epithelial and chondrosarcoma as mesenchymal component, with ultrasonographic, cytological and histological findings.

CASE REPORT

A 47-year-old female presented to E.N.T. outpatient department of Narayana hospital, Nellore, with a mass just below the right ear lobule of 4 weeks duration with mild tenderness. She had no significant medical complaints. No history of smoking and tobacco chewing were present. On clinical examination, mass was measuring 1x1 cm, firm in consistency and located just above the angle of mandible. It was fixed to the underlying structures. Skin overlying the tumor was normal. No lymphadenopathy was present.

Basic hematological investigations, liver function test, renal function tests, chest X- ray and ultrasonography (USG) of abdomen were normal. Provisional diagnosis of pleomorphic adenoma was made and fine needle aspiration cytology (FNAC) of the lesion was planned.



Figures - Fig 1- Adenocarcinoma component with malignant cells arranged in cribriform pattern (H&E X400), Fig 2- Malignant chondrosarcoma component (H&E X400), and Fig 3- Myxoid and hyaline change representing pre-existing pleomorphic adenoma

FNAC slides showed clusters and dispersed individual malignant epithelial cells exhibiting marked nuclear pleomorphism, increased nuclear-cytoplasmic ratio, and prominent nucleoli with abundant fibromyxoid stroma and hyaline material in the background. Diagnosis of atypical pleomorphic adenoma was made.

Because of atypical cells in FNAC, the patient was considered for surgery. A preoperative USG of the right region of the neck showed a well-defined lobulated, heterogeneously hypoechoic, 19 x 11 mm tumor in the superficial lobe of right parotid gland with minimal peripheral vascularity on CDFI with insignificant lymph nodes. The USG findings were consistent with benign pleomorphic adenoma. Thereafter, the patient underwent right total parotidectomy with full preservation of the facial nerve. The specimen was received and processed by the department of Pathology, Narayana medical college and hospital. It was fixed in 10% formalin, embedded in paraffin and routine H & E sections were obtained. Grossly, the parotidectomy specimen measured 5x3x1.5cm. The cut surface revealed a relatively well-defined, grayish white tumor, firm in consistency, measuring 1.5x1.5x1 cm, with adjacent normal salivary gland parenchyma.

Microscopically, the tumor was mainly composed of two components, carcinoma and sarcoma. The former was that of moderately differentiated adenocarcinoma. Carcinomatous component is characterized by malignant cells arranged in sheets, cribriform pattern and glandular pattern (Figure 1). The sarcomatous component was mostly composed of chondrosarcoma (Figure 2). Large foci of hyalinization and myxoid change representing pleomorphic adenoma were present (Figure 3). Mitotic figures and multinucleated tumor giant cells were also noticed. Background consisted of areas of necrosis. Two regional lymph nodes were isolated, both negative for tumor. So, a final diagnosis of carcinosarcoma of parotid gland was made. Postoperative state was uneventful and four months follow up of the case showed good recovery. No metastatic deposits were detected on clinical and radiological investigations.

DISCUSSION

Malignant mixed tumor of the salivary gland is composed of three distinct clinical entities - carcinoma ex pleomorphic adenoma (the most common type), metastasizing mixed tumor and carcinosarcoma [3]. The term true malignant mixed tumor was first described by King et al in 1967 [4].

The tumor genesis of carcinosarcoma is controversial and two hypotheses are suggested. According to some authors, carcinosarcoma and pleomorphic adenoma of the salivary gland may share a common precursor, possibly a myoepithelial cell (divergence hypothesis). On the other hand, it has been shown that in some carcinosarcomas, both components arise de novo (convergence hypothesis) [5]. In the present case, evidence of pleomorphic adenoma in multiple histologic sections studied, supports divergence hypothesis. Till now seven cases of carcinosarcoma have shown histologic evidence of pre-existing or co-existing pleomorphic adenoma [6, 7-9, 10, 11, 12].

Table 1 - Carcinosarcoma (True Malignant Mixed Tumors) of Salivary Glands: A review of literature

Source, year	Age/ Sex	Location	Sarcomatous component	Carcinomatous component
Garner, 1989 [15]	57/F	Parotid gland	Chondrosarcoma, osteosarcoma	Adenocarcinoma
Bleiweiss, 1992 [17]	64/M	Submandibular gland	Osteosarcoma	Ductal carcinoma
Carson, 1995 [7]	51/F	Parotid gland	Osteosarcoma	Adenocarcinoma
Alvarez-Canas, 1996 [6]	64/M	Parotid gland	Fibrosarcoma	Ductal carcinoma
Gogas, 1999 [18]	77/M	Submandibular gland	Chondrosarcoma, osteosarcoma, rhabdomyosarcoma	Ductal carcinoma
Kwon, 2001 [19]	47/M	Parotid gland	Rhabdomyosarcoma, Fibrosarcoma	Squamous cell carcinoma
Owaki, 2003 [20]	71/M	Submandibular gland	Chondrosarcoma	Myoepithelial carcinoma
Gnepp, 1993 [1]	43 case review		See discussion	
Present case	47/F	Parotid gland	Chondrosarcoma	Adenocarcinoma

Gnepp et al studied 43 cases of carcinosarcoma of the salivary glands [1]. The maximum number of these tumors (33%) was observed in parotid glands followed by submandibular glands (19%) and palate (14%). There was no sex predominance and the mean age at presentation was 58 years. The histology of the epithelial component comprised of salivary duct carcinoma as the most common (19 cases), followed by adenocarcinoma (6 cases), poorly differentiated carcinoma (5 cases), undifferentiated carcinoma (4 cases), squamous cell carcinoma (3 cases), and epidermoid carcinoma (1 case). Focal areas of mixed differentiation were also present in 5 cases. In this case report, patient is 47 year old female. The epithelial component is composed of adenocarcinoma whereas mesenchymal component is mainly formed by chondrosarcoma.

We also discuss some cases of carcinosarcoma of salivary gland in table 1. In the current case report, by cytology only carcinomatous component characterized by sheets of malignant epithelial cells was well noticed. Among mesenchymal components, only fibromyxoid and hyaline material were present and no sarcomatous component was noticed. This may be due to presence of focal chondrosarcoma component. Only in three reports [9, 10, 13], cytology features of carcinosarcoma parotid have been studied making this case unusual and supplementary to the limited literature.

The differential diagnosis of unilateral parotid lesion included benign mixed tumor, primary versus metastatic sarcomas, carcinoma ex-pleomorphic adenoma and muco-epidermoid carcinoma. In our case, tumor cells display atypical features including cellular and nuclear pleomorphism, hyperchromatism, mitotic figures and invasive growth. These features help in distinguishing it from benign mixed tumor. Carcinoma ex-pleomorphic adenoma and mucoepidermoid carcinoma were excluded on the basis of presence of sarcomatous component.

USG findings and clinical examination were suggestive of pleomorphic adenoma. FNAC has contributed substantially in its preoperative diagnosis as atypical pleomorphic adenoma. The definitive diagnosis of carcinosarcoma parotid was made based on histopathological examination. The prognosis is determined by clinical and histological stage of the disease. The prognosis is poor with 58-85% of patients dying of the disease. So, there should be close follow up of these patients for symptoms and signs of local recurrence and metastasis [14-16].

CONCLUSION

The reported case is a rare case of carcinosarcoma (true mixed malignant tumor) of the parotid gland arising from pre-existing pleomorphic adenoma with adenocarcinoma and chondrosarcoma as predominant epithelial and mesenchymal component respectively. The diagnosis of carcinosarcoma can be missed on USG examination where it was reported as benign lesion and fine needle aspiration where it was reported as pleomorphic adenoma with atypical features. Only histopathology is diagnostic for carcinosarcoma of salivary gland.

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