TRUE MALIGNANT MIXED TUMOR (CARCINOSARCOMA) MASQUERADING AS PYOGENIC GRANULOMA OF LOWER ANTERIOR REGION: A CASE REPORT

Reddy M. Vandana*, Saxena Susmita**

The malignant mixed tumor (carcinosarcoma) of the salivary gland is an extremely rare tumor. By definition, it is composed of both malignant epithelial and malignant mesenchymal components. We report a case of carcinosarcoma of a 63 year old man that arose "de novo" and contained epidermoid carcinoma as the epithelial component and chondrosarcoma as mesenchymal component. The etiologic and histologic features are presented and in addition, the literature is reviewed.

Key words: Malignant mixed tumor, Carcinosarcoma

INTRODUCTION

True malignant mixed tumor (carcinosarcoma) is an extremely rare tumor, comprising 0.04%-0.16% of all salivary gland tumors and 0.4% of all malignant salivary gland neoplasms. Carcinosarcoma of salivary gland is a heterologous neoplasm which is composed of both malignant epithelial and malignant mesenchymal elements. They are commonly seen in association with pleomorphic adenomas. But some carcinosarcomas arise de novo. The extreme rarity of the tumor with a still rarer "de novo" origin has prompted us to report this case.

CASE REPORT

A 63-year- old male patient reported to the Department of Oral Pathology and Microbiology, Subharti Dental College, Meerut with complaint of a bulbous growth in the mouth associated with pain since 1 year.

The patient was well nourished with moderate built with no obvious extraoral abnormalities. On intra oral examination, the patient had partially edentulous arches with a localized growth on the lingual aspect of the lower anterior region. The lesion was 3cm×3cm in size, well-demarcated, sessile, and extending from the midline towards the right side till the premolar region with intact mucosal covering and firm in consistency with a corrugated surface. The base appeared to be deep-seated and could be traced till the floor of the mouth. There was no bleeding on manipulation but tenderness was evident.

History revealed that the swelling had been present for past 1 year and was painful. The growth was initially small but had slowly enlarged to the present size. There were no palpable lymph nodes. The past medical history was non-contributory and the patient gives history of loss of teeth on its own without any surgical intervention. Based on the clinical features, a provisional diagnosis of pyogenic granuloma/ peripheral Giant cell Granuloma / Peripheral ossifying fibroma was made. The lesion was excised completely under local anesthesia and submitted for histopathology. The clinical photographs and radiographs were not taken as the clinician categorized it as a frequently occurring peripheral lesion of the mucosa and no incisal biopsy had been performed prior to the excision of the lesion. Results of routine hematologic tests were within normal limits.

The surgical specimen measured 3.5cm×3 cm ×1.5 cm in size. The surface of the specimen showed creamish white irregular outer surface and brownish cream inner surface. The specimen was fixed in 10% formalin and embedded in paraffin. Histological sections were stained with routine hematoxylin and eosin (Fig. 1).

*Post graduate student;
**Professor & Head; Department of Oral Pathology and Microbiology, Subharti Dental College, Meerut.

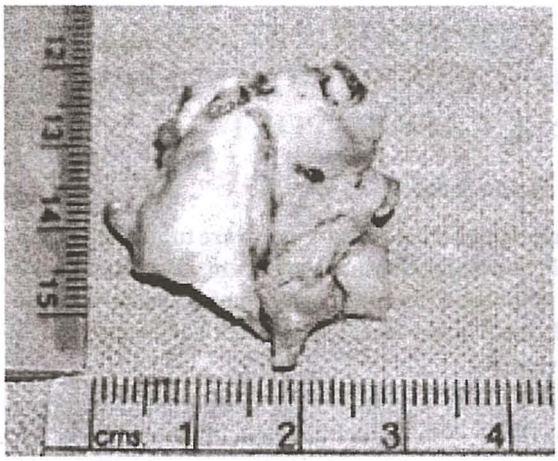


Fig. 1: Resected specimen consisting of circumscribed irregular mass, creamish white in color.

Histologic examination of the tumor displayed an intimate mixture of carcinomatous and sarcomatous elements, with the latter predominating. The malignant epithelial component was mainly epidermoid carcinoma. It showed ulcerated stratified squamous epithelium with chronic inflammation and multiple vascular channels. Tumorous proliferation of cells in cords and clumps was seen which were squamous in nature with dysplastic features. The malignant mesenchymal component was composed of clumps of tumor cells with aberrant mitotic figures and clear



Fig. 2: Section showing superficial epithelium with subepithelial tumor tissue (H and E, X40).

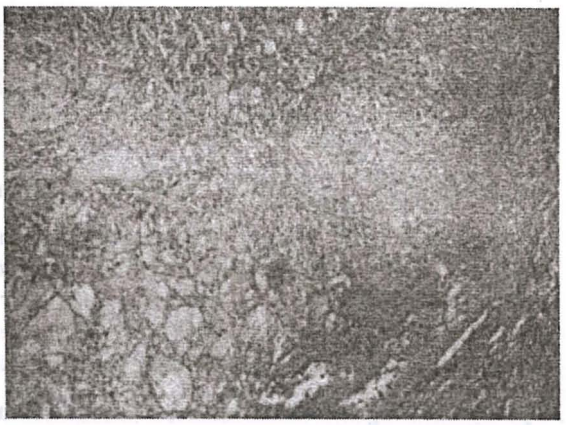


Fig. 3: Section showing cartilage like tissue and collections of mucous secreting cells (H and E, X100).



Fig. 4: Cartilagenous area showing nuclear pleomorphism and mitotic figures (H and E, X 400).

cell like appearance with areas of hyalinization resembling cartilage, which was suggestive of chondrosarcoma (Fig. 2, 3, 4). PAS positive nature of epithelial component of the tumor was noted with PAS staining (Fig. 5).

The biphasic histopathologic appearance prompted us to diagnose it as carcinosarcoma, which was seconded by multiple observers.



Fig. 5: PAS positive staining reaction of epidermoid component of the tumor (PAS, X100).

DISCUSSION

Based on the classification proposed by the Armed Forces Institute of Pathology, carcinosarcoma of the salivary gland does exist, although it is rare. In fact, it is so rare that according to the World Health Organization's histologic classification of salivary gland tumors, malignant mixed tumor only refers to the carcinoma in pleomorphic adenoma. Mixed carcinoma and sarcoma of the parotid gland was first described in 1951 by Kirklin et al of the Mayo Clinic, and sarcomatous component was thought to be the stroma of the tumor (1). The term true malignant mixed tumor (carcinosarcoma), which means a tumor consisting of both carcinomatous and sarcomatous elements, was first used by King in 1967 (2). Since that time, there have been approximately 62 cases of carcinosarcoma of the salivary glands reported in the English-language literature.

Gnepp summarized 43 cases of carcinosarcoma of the salivary glands published in the literature (1993). The mean age at presentation for 34 patients with available data was 58 years (range, 14–87 years). There was no sex predominance. Among 37 patients with available data, 18 were female and 19 male. Twenty-seven tumors were from parotid glands, 8 from submandibular glands, 5 from the palate, 2 from unspecified major salivary glands, and 1 from the tongue. Six tumors had microscopic evidence of arising in a benign mixed tumor. Our patient was a 63 year old male who presented with a painless mass of 1 year duration that suddenly increased in size causing pain. These clinical data are consistent with those reported by others. There have been another 19 cases either not included in Gnepp's article or reported afterward till date. Clinical data were available in 16 of these cases (mean age, 70 years; range, 51–86 years; 8 women and 8 men). Among these additional 19 cases, 14 were from parotid glands, 2 from submandibular glands, and 3 from the palate. There were 8 cases that showed clear histologic evidence of a preexisting or coexisting pleomorphic adenoma and 5 cases that failed to demonstrate such a relationship. In the other 6 cases, it was not stated in the reports whether the carcinosarcoma was associated with a pleomorphic adenoma or not (3).

Based on the literature of these 62 reported cases of true malignant mixed tumor of the salivary glands, the most common carcinoma was squamous cell carcinoma or adenocarcinoma, whereas the most common sarcoma was chondrosarcoma (4). In the current case, the carcinomatous component was squamous cell carcinoma and the PAS positive nature of the tissue leads us to believe that the origin might be from salivary glandular element. The sarcomatous component showed chondrosarcoma. Although it is recognized now that true malignant mixed tumors of the salivary glands do exist, their histogenesis and pathogenesis are still under discussion. Clinical and histologic studies showing their relationship to pleomorphic adenomas imply that true malignant mixed tumor (carcinosarcoma) and pleomorphic adenoma of the salivary gland may share a common precursor cell in that the myoepithelial cell is a major component in their development. On the other hand, it has been shown that some carcinosarcomas arise de novo (5). Also it has been demonstrated that certain carcinosarcomas do not show evidence of myoepithelial cell origin or differentiation and that they may arise independently of pleomorphic adenoma. Our case under discussion disclosed no existence of pleomorphic adenoma, and the carcinomatous and sarcomatous components did not show

any evidence of myoepithelial cell differentiation. Our observation is in agreement with that of Latkovich (1998) in that there is a subset of carcinosarcomas of salivary glands that arise de novo, independent of pleomorphic adenoma. It is suggested that neoplastic cells may originate either from an undifferentiated multipotential cell or from two distinct precursor cells (6).

The present case might have originated from minor salivary glands of the floor of the mouth as there was no evidence of submandibular and sublingual gland involvement. Thus, it can be categorized under true malignant mixed tumor of oral minor salivary glands, which is a rare entity with approximately 10 cases documented in the literature (7). The reason for concluding it as salivary gland origin is all carcinosarcoma's arise from salivary glands. Postoperative follow up has been done till 4 month after complete excision of the lesion and apart from slight numbness in lower lip and chin area, the patient remains asymptomatic. Local recurrence of the lesion has not been detected till date, which leads us to believe that complete excision of the lesion was perhaps adequate in treating the tumor. However, periodic follow up is recommended as previously reported cases of this entity have shown frequent recurrence and distant metastasis.

REFERENCES

1. Kirkin JW, McDonald JR (1951): New parotid tumors; histopathology, clinical behavior and end results, Surg Gynecol Obstet, 92: 721-33.
2. King OJr (1967): Carcinosarcoma of accessory salivary gland: first report of a case, Oral Surg Oral Med Oral Pathol, 23: 651-659.
3. Kwon and Gu (2001): True malignant mixed tumor (Carcinosarcoma) of parotid gland with unusal mesenchymal component: a case report and review of the literature, Archives of Pathol and Lan Med, 125: 812-815.
4. Quang Liu, Jain-shan ZHU (2005): Parotid gland carcinosarcoma with follicular dendritic cell sarcoma as mesenchymal component: a case report, CMJ,118: 516-518.
5. Kavita Mardi, Jaishree Sharma (2004): True malignant mixed tumor (carcinosarcoma) of parotid gland: a case report, Indian J Pathol Microbiol, 47: 64-66.
6. Alvarez-Canas (1996): True malignant mixed tumor (carcinosarcoma) of the parotid gland: report of a case with immunohistochemical study, Oral Surg Oral Med Oral Pathol Oral Radiol Endod, 81: 454-8.
7. John KH, John CC, Christopher MP (1997): AJNR, 18:1944-1948.
8. Carolina Thome Capuano A, Carvalhosa AA (2007): Immunoprofile of a carcinosarcoma of the submandibular gland, Oral Surg Oral Med Oral Pathol Oral Radiol Endod, 103: 398-402.
9. Yura S, Yamashita T (2007): A case of carcinosarcoma arising in the submandibular gland, Oral Surg Oral Med Oral Pathol Oral Radiol Endod, 103: 820-824.
10. Gotte K (2000): Salivary gland carcinosarcoma: immunohistochemical, molecular genetic and electron microscopic findings, Oral Oncology, 36: 360-364.

ATTENTION CONTRIBUTORS

All contributors are requested to send two hard copies of the article along with a complete copy on a CD, preferably containing scanned figures and photographs with legends to avoid spelling and other mistakes.

- Chief Editor