



Carcinosarcoma of minor salivary gland of lower lip with pelvic metastasis: A case report

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ABSTRACT

Carcinosarcoma of salivary gland is a rare and aggressive tumor composed of both carcinomatous and sarcomatous components which metastasize together. Most tumors arise in parotid gland from a preexisting pleomorphic adenoma.

Methods and Results: We report a case of carcinosarcoma of minor salivary gland of lower lip with subsequent infiltration to mandible and pelvic metastasis after recurrence. The patient was a 38 year old man who underwent surgical resection and adjuvant chemo radiotherapy and demonstrates recurrence after 9 months with metastasis to left iliac fossa.

Conclusion: Carcinosarcoma of minor salivary gland is an extremely rare malignancy with poor prognosis and is best managed by combination of surgical resection and postoperative radiation.

Key words: Carcinosarcoma, Salivary gland, Metastasis, Recurrence.

Introduction

Malignant mixed tumor of salivary glands can be divided into three different morphological types:

- 1) Carcinoma ex-pleomorphic adenoma.
- 2) Carcinosarcoma (true malignant mixed tumor).
- 3) Metastasizing mixed tumor.

More than 99% of malignant mixed tumors arise in a pre-existing pleomorphic adenoma with only 0.2 % being carcinosarcoma [1]. Carcinosarcoma is a biphasic and extremely rare tumor in which both carcinomatous and sarcomatous elements are present and metastasize together.

er. More than half of the cases of carcinosarcomas exhibited preexistence or coexistence of pleomorphic adenoma. Most carcinosarcomas have been reported in parotid gland [2]. To the best of our knowledge, there has been no reported case of intraosseous carcinosarcoma with distant bony metastasis. We present a case of carcinosarcoma of minor salivary gland of lower lip coexisting a pleomorphic adenoma with pelvic metastasis after recurrence.

Case Report

A 38 year old Iranian male was referred to the department of oral and maxillofacial surgery of Shariati hospital, Tehran, Iran with a rapidly enlarging mass in mandible labial

vestibule measuring 55* 35 mm for the past month. The lesion had firm consistency and in CT scan, it seems to be from lower lip mucosa originally with significant bone infiltration and destruction and soft tissue invasion (Figure 1,2). The patient had no neurosensory dysfunction in lower lip and chin. He had no past history of salivary gland pathology. Incisional biopsy revealed the mass as malignant mixed tumor. There was no cervical lymphadenopathy in neck and chest and brain CT scans were clear of any metastatic lesion. He underwent partial mandibulectomy from the first molar area on the ipsilateral side to the contralateral canine area. Mandible continuity was regained with the aid of a reconstruction plate (Figure3).

Histopathologic examination revealed a biphasic tumor composed of both sarcomatous and carcinomatous components. In serial section evaluation, pleomorphic adenoma was also seen in the specimen. Immunohistochemistry was positive for vimentin in the majority of tumor cells, while S100 and SMA were only positive in myoepithelial cells. Ki 67 was positive in 80 % of tumor cells. The results of IHC analysis was negative for EMA, CK and desmin.

Postoperatively, the patient underwent the combination of chemoradiation as adjunctive therapy. The prescribed regimen of his chemotherapy included ifosfamide plus mesna, cisplatin and fluorouracil. At the end of the adjunctive therapy, 9 months after the initial surgery, local recurrence became evident. The localized pain worsened and the patient had become cachectic and had lost 45 kg due to his severe malnutrition. An irregular, ulcerated exophytic and exsanguinating mass rapidly grew in the skin overlying the previous surgical site and the reconstruction plate became exposed (Figure 4). CT imaging revealed significant invasion to infratemporal space superiorly, and to submandibular space inferiorly (Figure 5). The patient also complained of reduced force in his lower left extremity. In abdominal CT scan a soft tissue mass lesion was seen in left iliac fossa which extended outside of left iliac bone. The soft tissue of the mentioned lesion extended downward around femoral head and mild bone destruction was seen. In contrast enhanced CT scans, chest and brain had no tumoral involvement. The patient underwent a more radical surgical resection including hemimandibulectomy with the resection of overlying skin. The defect was reconstructed with vascular pectoralis major myocutaneous flap. Incisional biopsy of the pelvic mass, proved the mass to be metastatic consisting of both carcinomatous and sarcomatous elements.

In histopathologic evaluation, the mandibular lesion showed a biphasic pattern of both polydifferentiated sarcomatous and carcinomatous parts. The sarcomatous component predominates and was mainly composed of chondrosarcoma (Figure 6-A) whereas the carcinomatous part was mainly an adenocarcinoma, not otherwise specified (NOS) (Figure 6-B). In serial sections no remnants of pleomorphic adenoma were seen in the specimen. Significant perineural and vascular invasion was evident, along with numerous mitotic figures and polychromatic nuclei (Figure 6-C). In IHC study, the specimen was positive for S100 and vimentin and Ki67 was expressed in nearly 80 % of tumor cells. Cytokeratin and EMA were positive in few isolated cells and ductal structures, whereas Desmin was positive in vessel walls and surrounding muscle tissue. The patient was referred to the oncologist for the appropriate adjunctive therapy.

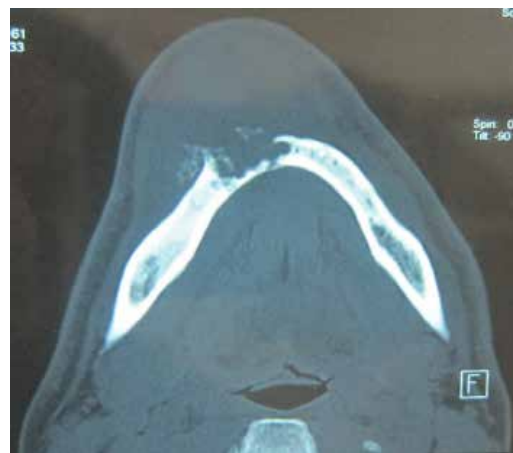


Figure 1. Axial cut of the CT scan of the lesion with significant bone destruction and soft tissue invasion.



Figure 2. Orthopantomogram of the destructive lesion on mandible.

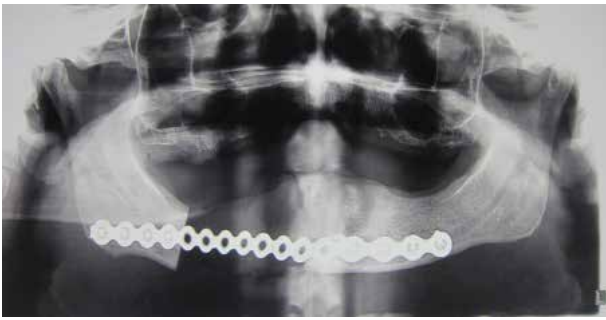


Figure 3. Partial mandibulectomy with the placement of a reconstruction plate.



Figure 4. Clinical view of the recurrence after 9 months.

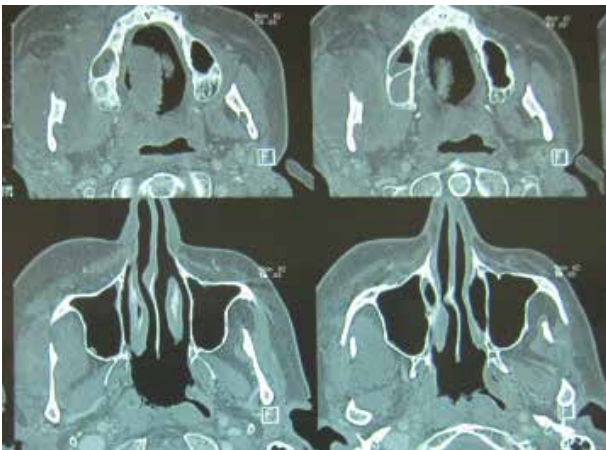


Figure 5. Significant invasion of the recurrent lesion to infratemporal space.

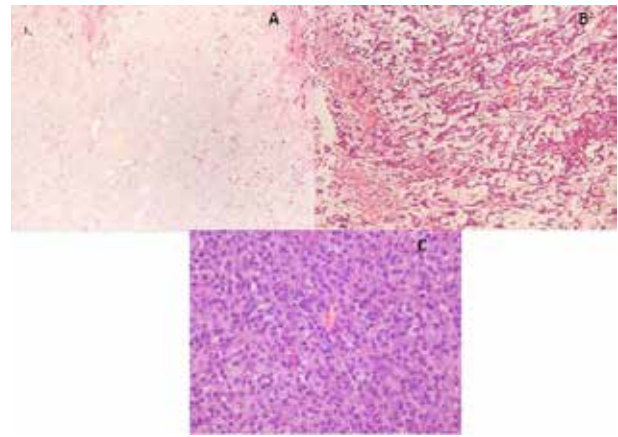


Figure 6. Histopathologic evaluation of the specimen (H&E). A: sarcomatous component. B: carcinomatous component. C: numerous mitotic figures and polychromatic nuclei.

Discussion

Minor salivary gland tumors are comparatively uncommon and comprise approximately 10% of all salivary tumors [3]. Less than half of these tumors were malignant, palate is the most common anatomic site [4]. Pleomorphic adenoma or benign mixed tumor is the most common salivary neoplasm [4]. Malignant degeneration is a potential complication. The risk of malignant transformation is probably small, but it may occur in as many as 5% of all cases [5]. It is also possible that malignant mixed tumors arise de novo, without a preexisting benign mixed tumor. Malignant mixed tumor is an uncommon malignancy which comprises 2-6% of all salivary gland tumors [6]. Among the three mentioned subtypes, carcinosarcoma is extremely rare accounting for less than 0.2% of all salivary gland tumors [7]. Its prognosis is poor; around 75% of patients either die from their disease or develop recurrent local tumor or metastases [6]. Most carcinosarcomas develop in parotid gland. Palate is the most common site of carcinosarcoma of minor salivary glands [8]. Few cases have been reported in floor of the mouth [9] and tongue [10]. The tumor most often occurs in advanced ages and is more frequent in men than in women [7]. Histologically, carcinosarcoma is characterized by the presence of malignant epithelial and stromal components which metastasize together. The most common carcinomatous and sarcomatous elements yet reported are adenocarcinoma and chondrosarcoma, respectively [7].

The histogenesis of carcinosarcoma is a matter of controversy. Some authors believe that benign and true malignant mixed tumors share a common precursor, probably myoepithelial cell (monoclonal hypothesis)

[11,12]. This theory seems more accurate for cases in which pleomorphic adenoma coexists or preexists carcinosarcoma. Others support the theory which implies independent malignant transformation of the carcinomatous and sarcomatous elements, which then unite [13].

On rare occasions, salivary gland tumors arise centrally within the jaws. The most common intrabony salivary tumor is intraosseous mucoepidermoid carcinoma. For those intrabony tumors associated with impacted teeth, it has been postulated that they originate from pluripotential odontogenic epithelium. Other possible sources include actual minor salivary glands entrapped developmentally within the bone or from mucous glands of maxillary sinus for maxillary tumors. So far no intraosseous malignant mixed tumor has been reported in english language literature.

Due to its rarity, there is no established therapeutic approach for this malignancy. Staffieri et al [7] found that the combination of surgery and radiation had a significantly lower rate of recurrence than surgical excision alone. The effectiveness of chemotherapy is still unclear.

Our patient was a middle aged man who presented with a non painful rapidly growing mass in mandible. In the present case, despite being multi differentiated, the carcinomatous part was mainly an adenocarcinoma and the sarcomatous part was a chondrosarcoma.

In immunohistochemical investigation after the first and second resection, Ki 67 was positive in 80% of tumor cell. Ki-67 is a nuclear non-histone protein that is present at low levels in quiescent cells but is increased in proliferating cells. Its overexpression is frequently seen in a variety of malignant tissues and is considered as a negative prognostic factor in many malignancies [14]. The aggressive course of disease in our patient could partly be attributed to high level of Ki 67 expression in tumor cells.

Carcinosarcoma de novo is an extremely rare entity in salivary gland malignancy and exhibits a broad range of morphological presentations. Considering its aggressive biological course, the combination of radical surgical excision and radiotherapy accompanied by close follow up visits seems to be the most appropriate treatment protocol.

Conflict of Interest

There is no conflict of interest to declare.

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