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Case Report

Aggressive salivary duct carcinoma with widespread dissemination: a case report

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ABSTRACT

Salivary duct carcinoma is a rare and aggressive salivary gland malignancy with a poor prognosis. Due to the paucity of literature, very little is known about this neoplasm. We have described such a case in this case report and highlighted the clinical and histopathological features associated with this disease. A sixty-year old male patient reported in the outpatient department of a regional dental college and hospital with a circular firm growth in the mandibular anterior region and mobility of adjacent mandibular teeth. An array of investigations including radiographic, tomographic, ultrasonographic as well as histopathological were performed**.** It was diagnosed as invasive salivary duct carcinoma with distant metastases in the shoulder joint, ribs as well as pelvic bones. Currently, no National Comprehensive Cancer Network guidelines for the specific treatment of salivary duct carcinomas exist. National Comprehensive Cancer Network guidelines recommend complete surgical excision of tumors for major salivary gland tumors without nodal involvement (N0) with or without neck dissection for high-grade and T3/T4 salivary gland tumors.

Keywords: Neoplasm metastasis, Salivary gland neoplasm

INTRODUCTION

Salivary duct carcinoma (SDC) is a rare, highly aggressive malignancy. The term salivary duct carcinoma describes the malignancy that histologically resembles in situ and invasive ductal carcinoma of the breast. The parotid gland is the most commonly involved salivary gland accounting for approximately 80% of tumors, followed by the submandibular gland (8-12%) and minor salivary glands less than 10%.[1,2] SDC is an extremely rare malignancy with an estimated incidence of 1-1.2 in 1,000,000 patients, with a higher prevalence in men.[3] It is often diagnosed at an advanced stage as it metastasizes early to regional lymph nodes and distant sites. Immunohistochemically, tumor cells are diffusely positive for cytokeratin 7 and androgen receptor stains and may be GATA binding protein 3 positive. HER2/neu positivity occurs in 15-40% of patients with SDC.[2]

The mainstay of treatment for SDC is wide surgical resection along with lymph node dissection followed by adjuvant radiation therapy. The role of adjuvant chemotherapy and targeted therapies has limited benefit to date. Overall survival of the metastatic disease is poor, and 60-80% of patients with advanced-stage (T4) die within three years.[4] Adeberg et al listed 1282 cases of SDC found in the literature between 1987 and 2019.[4] This malignancy poses a challenge for physicians to provide education and information to the patients regarding the overall prognosis and treatment options given the absence of consensus guidelines.

As very few cases have been reported in the literature, this report will help in better understanding of this elusive neoplasm.

This section should include a critical review of the pertinent literature.

CASE REPORT

A sixty-year-old male patient reported in the outpatient department of a regional dental college and hospital with a chief complaint of swelling in the lower anterior mandibular region which had been present for 15 days (Figure 1).

Figure 1: Pre-operative photograph of a 60-year old male patient who reported to the outpatient department of a regional dental college. Absence of any extraoral swelling.

The patient initially underwent extraction of lower anterior teeth which were mobile. A few days after undergoing extraction the patient reported the presence of a solitary exophytic sessile swelling in the lower anterior mandibular region (Figures 2 and 3).

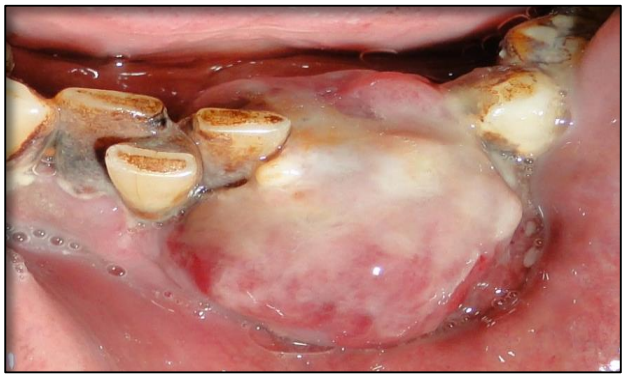

Figure 2: Intraoral examination revealed the presence of a solitary sessile mass in the lower anterior region which was firm, tender on palpation and bled on provocation.

The lesion measured 3 by 2 cm in maximum dimension and extended from the lower right incisor to the lower left premolar region obliteration of the labial vestibule. The mucosa overlying the swelling was covered with necrotic slough. It was associated with sharp and shooting pain and was not relieved with analgesics. The lesion was accompanied by fever but there was no history of bleeding or pus discharge, trismus, or paresthesia of the lower lip and chin region. On palpation, the lesion was tender, nonfluctuant, non-compressible, and bled easily. The buccal cortical plate was expanded and lower anterior teeth adjacent to the lesion also demonstrated grade III mobility. The submental lymph nodes were palpable, non-tender, mobile, and not fixed to overlying skin or underlying structures. The panoramic radiograph revealed irregular punched-out radiolucency in the lower anterior region and bone loss concerning lower molars (Figure 4).

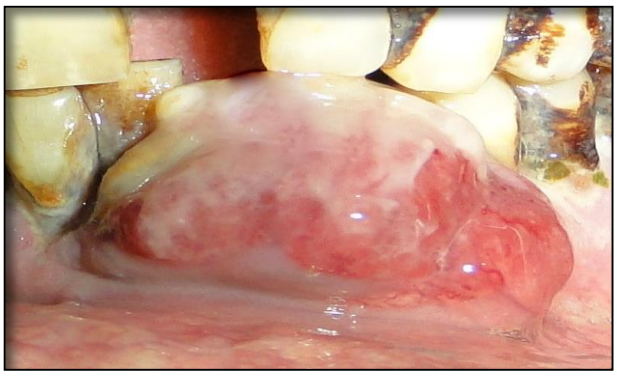

Figure 3: The lesion was also ulcerated and covered with necrotic slough.

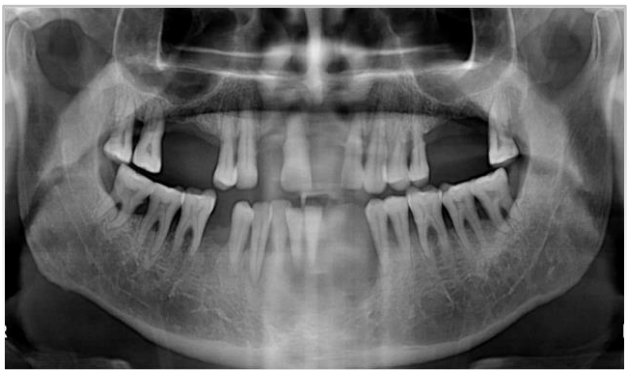

Figure 4: Panoramic radiograph demonstrating presence of an irregular, poorly defined radiolucency in the mandibular left incisor-premolar region.

He underwent an incisional biopsy under local anesthesia and the specimen was sent for histopathological evaluation. The histopathological evaluation revealed ductal necrosis with fenestrated epithelium and invasion of the underlying muscles with tumor islands (Figure 5). A diagnosis of salivary duct carcinoma of minor salivary was established. The patient reported a few days later with severe radiating pain in the left shoulder causing difficulty in standing and walking. USG of the left shoulder revealed tender nodules in the soft tissues around the shoulder girdle. Serum phosphorus and alkaline phosphatase levels were also elevated. The magnetic resonance imaging (MRI) of the left shoulder joint revealed altered signal areas in the scapula and humerus with peri-capsular soft-tissue components suggestive of metastasis. Positron emission tomography–computed tomography (PET-CT) scan revealed increased uptake over the left shoulder joint area, right 1st, 2nd, and 4th rib areas, T12 vertebrae, and the pelvic bones suggestive of metastases.

Immunohistochemistry of the oral mucosal lesion was HER2/neu and cytokeratin 7 positive and negative for mucicarmine. The patient received palliative radiotherapy in the left shoulder and T12 vertebral region for 2 weeks and was advised to report for palliative chemotherapy. However, the patient succumbed to the disease a few weeks later.

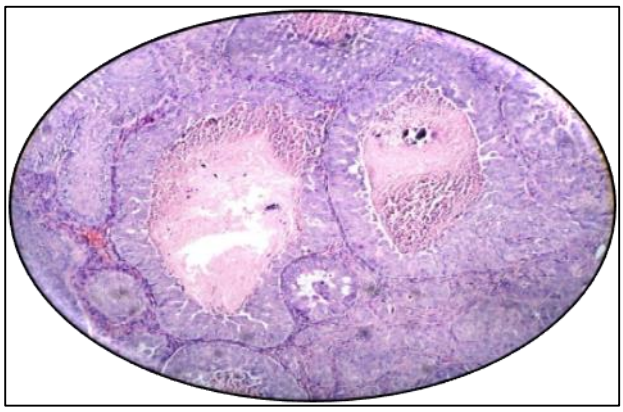

Figure 5: Histopathological evaluation revealed comedo-like intra-ductal necrosis with fenestrated epithelium/ roman bridge appearance as well invasion of underlying muscle with tumour cells.

DISCUSSION

Kleinsasser et al described salivary duct carcinoma (SDC) in 1968 as a salivary malignancy that histologically resembles in situ and invasive ductal carcinoma of the breast.[2,5] Most tumors arise de novo, although 20% of cases arise secondarily from preexisting benign pleomorphic adenoma (i.e., carcinoma ex pleomorphic adenoma).[2] The most common presenting symptom is a painless neck mass. Locally advanced disease can involve the facial nerve resulting in facial weakness.[6] According to the case series published by Jaehne et al, which included 50 cases, around 66% were present at the T3/T4 stage.[7,8] SDC predominantly affects male patients (66-75%), presenting with a wide age range from 34 to 83 years and a mean age of diagnosis at 62.5.[8] SDC is a highly aggressive high-grade salivary malignancy with a tendency for early regional, distant metastasis, and a high-grade recurrence. The most common sites of distant metastasis are the lung, bone, brain, and liver. Perineural invasion, lympho-vascular invasion, extracapsular spread, higher N stage, and facial nerve involvement are often considered negative prognostic indicators, although findings are inconsistent. Cheng et al described that perineural invasion and LVI were frequently present in approximately 57-69% and 61-70% of patients, respectively. Extra-nodal invasion occurred in 58% of patients.[2] The series published by Jaehne et al. found local disease recurrence in 48% of patients at 17.4 months after initial treatment and distant metastases in 48% after an average of 28 months.[8] The mean survival of patients in this report was 36 to 56 months. The 5-year survival rate for stage I disease was 42%, stage II was 40%, stage III was 30.8%, and stage IV was 23.2%.[8] Patients with parotid gland involvement had a better prognosis than those with submandibular or minor salivary gland involvement. Currently, no National Comprehensive Cancer Network (NCCN) guidelines for the specific treatment of SDC exist.

NCCN guidelines recommend complete surgical excision of tumors for major salivary gland tumors without nodal involvement (N0) with or without neck dissection for high-grade and T3/T4 salivary gland tumors. The recommendation for those with node involvement (N+) is complete surgical resection combined with neck dissection. In addition, they recommend adjuvant radiotherapy for high-risk features like intermediate or high grade, close or positive margins, neural/perineural invasion, lymph node metastasis, lymphatic/vascular invasion, and T3/T4 tumors. Postoperative radiotherapy for SDC is an appropriate therapeutic option regardless of stage and margin status.[9] Excellent local control rates can be achieved with extensive (local) surgical treatment and postoperative RT.[10] Randomized clinical trials demonstrating the benefit of cytotoxic chemotherapy are lacking. SDC treatment is most often with targeted therapy. Androgen receptor expression is seen in over 90% of the cases of SDC, making the tumor a potential target for androgen deprivation therapy.[9] Additionally, HER2 expression occurs in between 15 and 40% of cases.[2] Therapy with trastuzumab for HER2 overexpressing tumors improves overall survival.[9] A case series of 50 tumor samples published by Jaehne et al showed that the intensity of HER2/-neu expression correlates with the early development of distant metastasis and a poor 5-year survival rate. The 2-year, 3-year, and 5-year survival in patients with HER2 1+ were 88.9%, 55.6%, and 11.1% compared to 51.4%, 17.1%, and 0%, respectively, in those with HER2 3+ positivity.[8] Further research regarding androgen deprivation therapy and other targeted therapy is warranted. There are no consensus guidelines in the management of metastatic disease.

CONCLUSION

The rarity of salivary duct carcinoma and the lack of consensus guidelines necessitates the reporting of these cases. Finally, we hope clinical trials employing robust historical data will soon address the treatment of metastatic SDC, which remains elusive.

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