

## Salivary duct carcinoma – A case report

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### Abstract

Salivary duct carcinoma is an aggressive malignancy which phenotypically resembles high-grade breast ductal carcinoma, it has a high mortality rate and parotid gland is the most common location. Here, we report a case of an extensive salivary gland malignancy involving the left middle third of face in a 65-year-old male patient. Computed tomography confirmed a large and extensive heterogenous radiopacity in the left maxillary sinus region which revealed calcification causing destruction of the posterolateral and medial wall of the left maxillary sinus and also causing severe destruction of the left lateral wall of the nose and the pterygoid plates.

**Keywords:** Ductal carcinoma, Malignancy, Recurrence.

### Introduction

Salivary gland cancers represent 3%–6% of all head and neck cancers in adults.<sup>1,2</sup> Salivary gland malignancies are a heterogeneous group of tumors that presently involves 24 histologically distinct cancer subtypes.<sup>3</sup> Salivary duct carcinoma (SDC) is a high grade adenocarcinoma which arises from the ductal epithelium of salivary glands. It is also termed as cribriform salivary carcinoma of excretory ducts and infiltrating salivary carcinoma.<sup>4</sup> These tumors justify a separate classification because of their distinctive clinical and pathologic characteristics.<sup>5</sup> Clinically, the tumors can be characterized by their aggressive behaviour with early metastasis, local recurrence, and significant mortality.<sup>6</sup> SDC is more aggressive than the other salivary malignancies, with 5-year overall survival (OS) ranging from 42% (stage I) to 23% (stage IV). It shows male predominance commonly in their fifth or sixth decade. It arises from the ductal epithelium, predominantly occurring in the major salivary glands, especially the parotid gland.<sup>5</sup>

Histologically, they have a striking resemblance to breast carcinoma of the ductal type, with intraductal and invasive components. Salivary duct carcinoma have gained acceptance because it is used in the WHO classification of tumors. It has a predictably aggressive behaviour. The incidence of SDC is relatively rare, although it is difficult to determine.<sup>7</sup> The main differential diagnosis of LG-SDC includes cystadenoma, cystadenocarcinoma, sclerosing polycystic adenosis, salivary duct carcinoma in situ/high grade intraductal carcinoma, and papillary-cystic variant of acinic cell carcinoma.<sup>8</sup> This tumour has a tendency for infiltrating into the surrounding tissues and they frequently show haematogenous and lymphatic metastasis. Salivary duct carcinoma has an aggressive clinical course and they have a poor prognosis; most series showing that more than 70% of patients die within three years of disease.<sup>7</sup>

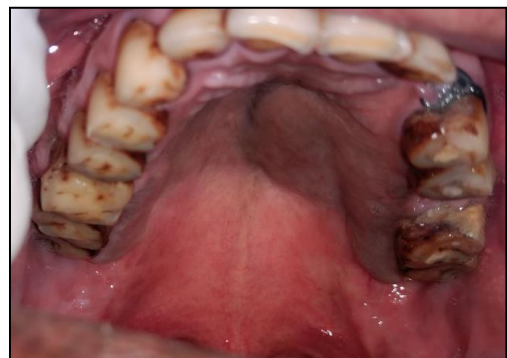
### Case Report

A 65yr old male patient reported with pain in the left middle one-third of the face since 4 months & with a swelling on the palate since 1 month. Pain was gradual in

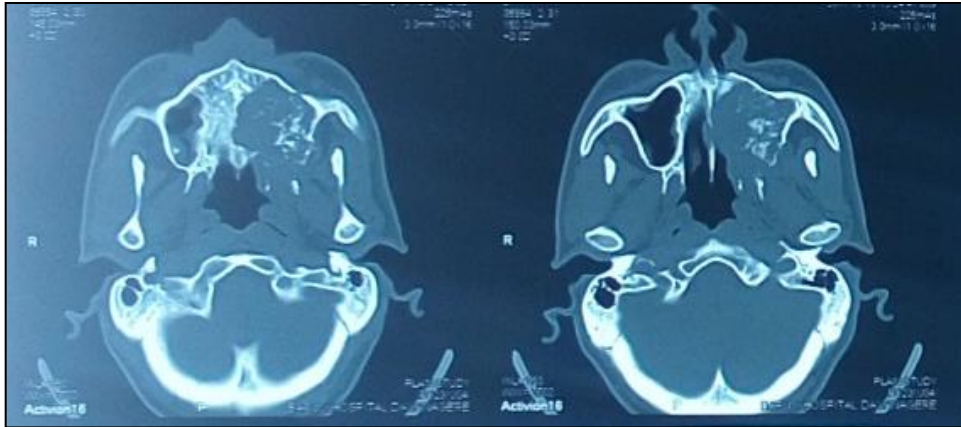
onset, severe, continuous & radiating to the left temporal & orbital region. Extra oral examination revealed a mild facial asymmetry on the left side owing to the presence of mild mid-facial swelling.

Intraoral examination revealed a well defined solitary swelling in the left side of the posterior hard palate region extending from the premolar to the molar region and extending from the mid palatine raphe to the free gingival margin. The swelling was firm and non fluctuant and severe tenderness was elicited on palpation.

CT images taken revealed ill defined soft tissue density lesion in the hard palate and maxillary alveolus extending into the left maxillary sinus involving the floor, medial, anterior and postero-lateral walls showing post contrast enhancement. An incisional biopsy was done & histopathology report revealed tumor cells in cribriform ductal component with Comedo necrosis and few solid ducts. Individual tumor cells showed pleomorphism with prominent nucleoli and the adjacent area showed desmoplastic stroma and areas of calcification. At some places bony trabeculae with osteocytes were seen along with congested blood vessels features which were suggestive of salivary duct carcinoma. Patient is currently undergoing radiotherapy.



**Fig. 1: Intraoral examination**



**Fig. 2: Computed tomography scan**

### Discussion

Salivary duct carcinoma (SDC) is rare malignancy which has a highly aggressive nature and estimated to represent 1% to 3% of all the salivary malignancies. Kleinsasser in the year 1968 first described this lesion as Speichelductcarcinome owing to its histologic similarity to ductal carcinoma of the breast. The lesion appears to be predominately in the parotid gland, and the literature states that most patients die within 3 years, with an overall survival reported as low as 42% for stage I disease and 23% for stage IV disease.<sup>5</sup>

The tumour has a predilection for male gender and the age ranges from 22 to 91 years, but most patients are older than 50 years. The tumour is said to be originated from the excretory and interlobular ducts of the major salivary glands. This tumour has a propensity for infiltrating into the surrounding tissues and frequently shows haematogenous and lymphatic metastasis.<sup>9</sup>

Huh KH et al reviewed the radiologic descriptions of SDC in the literature. He stated that in a case panoramic radiograph in the mandible revealed a poorly circumscribed radiolucency. The computed tomography of the case further demonstrated the invasion into the adjacent pterygoid muscle, a subcutaneous mass adjacent to the masseter muscle, an irregular mass with heterogeneous high density, and the nearly total replacement of the parotid gland by the tumor. MR images revealed low signal intensity on both T1-weighted images and T2-weighted images with an ill-defined infiltrative margin and an ill-defined high- and low-signal area on the enhanced T1-weighted image. He states that these radiologic findings may reflect the malignant features of SDC. An ill-defined, infiltrative margin can be detected on computed tomography and MR images and also on plain radiographs in cases involving hard tissues. The literature shows that the MR imaging is the imaging modality of choice because of its greater signal intensity, degree of enhancement, and the detail it provides on the internal architecture and characteristics of the margin, all of which can be useful in the evaluation of the aggressiveness of the lesion. However SDC cannot be differentiated from other high-cellularity malignant tumors by means of the mentioned imaging features. But high-grade squamous cell

carcinoma, mucoepidermoid carcinoma, and adenoid cystic carcinoma originating in the palate can be considered in the differential diagnosis because of the radiologic findings.<sup>10</sup>

On immunohistochemistry studies of ductal carcinoma, the cells are usually positive for cytokeratin 7 (CK7), CK20, and HER2 but negative for S100 and from 67% to 83% of SDCs express the androgen receptor (AR).<sup>11</sup> In patients with progressive SDC, trastuzumab is found to improve disease-free and overall survival. Androgen deprivation therapies (ADTs) have also been explored in small studies and have produced promising results.<sup>12</sup>

Overall, SDC is found to be one of the most aggressive salivary malignancies. According to the literature, death occurs in 60–80% of patients, usually within the first 5 years; about 33% develop local recurrence and 50% develop distant metastases, at different sites including bone, lungs, brain, liver and skin. It has been found that the morphologic variants behave in a similar way to the usual type of SDC, although there is evidence that the most aggressive is the micropapillary variant.<sup>4</sup> For invasive SDC the standard treatment at present is complete surgical excision with radical neck dissection which is followed by radiotherapy to the tumor bed and possibly chemotherapy.<sup>9</sup>

### Conclusion

Salivary duct carcinoma is a malignancy that will be rarely seen by most physicians henceforth providing education and information to patients about prognosis remains challenging. Good local control can be achieved by surgery and postoperative radiotherapy in SDC patients, but high distant metastasis remains a major challenge.

**Conflict of Interest:** Nil.

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