

SARCOMATOID SALIVARY DUCT CARCINOMA OF PAROTID GLAND – A RARE HISTOLOGICAL VARIANT

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ABSTRACT :

Salivary duct carcinoma (SDC) is a rare and high-grade malignant tumor that arises predominantly in a major salivary gland and bears a striking histological resemblance to high grade ductal adenocarcinoma of breast. The sarcomatoid SDC is a rare subtype that comprises of both epithelial and sarcomatoid components. The authors describe a case of sarcomatoid SDC of parotid gland. Its histological features have been discussed. The patient was treated by total parotidectomy with wide local excision of the involved skin and ipsilateral neck dissection followed by adjuvant radiotherapy.

Keywords: Ameloblastomatous CCOT, Clinicopathological Conference, Mandibular Angle Region, Unilocular Radiolucency

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INTRODUCTION :

Salivary duct carcinoma (SDC) is a rare and high-grade malignant tumor that arises predominantly in a major salivary gland. It accounts for 0.9% to 6% of all parotid's tumors. Several histological variants of SDC have been previously described in literature. Sarcomatoid SDC is one such variant, which is characterised histologically by the presence of typical SDC as the epithelial component and pleomorphic spindled cells with cytological atypia as the sarcomatoid component. Histological features and the nomenclature of this entity has been a matter of debate for many years. The immunohistochemical, electron microscopic and recent molecular findings in this biphasic neoplasm suggest that sarcomatoid SDC (SSDC) is a more appropriate term than “carcinosarcomas”. SSDC is an aggressive neoplasm known for the high incidence of nodal metastasis, distant metastasis and loco-

regional recurrence. We describe a case of sarcomatoid salivary duct carcinoma in a 45 year old man who presented with involvement of skin and subcutaneous tissue.

CASE REPORT:

A 45 year old man presented to our outpatient department with complaints of a slowly progressive, painless right infra-auricular swelling for past 2 years. The swelling ulcerated 6 months back; the size of ulceration had been increasing since then and was associated with small amount of intermittent bleeding, especially on touch. He did not give any history of facial asymmetry or constitutional symptoms. He denied any addictions. Physical examination revealed 8 X 8 cm right infra-auricular swelling. Overlying skin showed a 4 X 4 cm ulceration with a reddish, granular surface which bled on touch. Rest of the skin was tense and could not be pinched/was involved by the tumor (Figure 1). On palpation, the surface was lobulated and it was variegated in consistency with soft to firm areas. Facial nerve function was intact.



Figure 1 showing the right parotid swelling with ulceration or fungation of the overlying skin

MRI revealed a 6.5 X 3.5 X 6.2 cm size solid-cystic, well-defined exophytic mass arising from superficial lobe of right parotid gland and infiltrating the overlying skin and subcutaneous tissue. Large necrotic areas were seen within the tumor. Inferior aspect of deep lobe also showed patchy focal lesions. The solid component was isointense on T1wi and iso- to slightly hyperintense on T2wi and showed mild heterogeneous post-contrast enhancement (Figure 2 -5). Necrotic lymph nodes were visualised at left level 2.

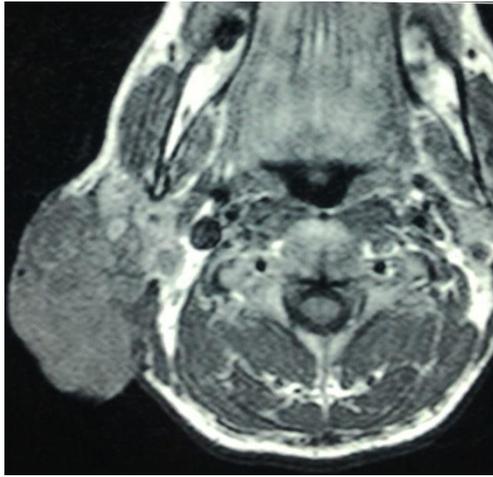


Figure 2 showing T1 isointense mass superficial lobe of right parotid

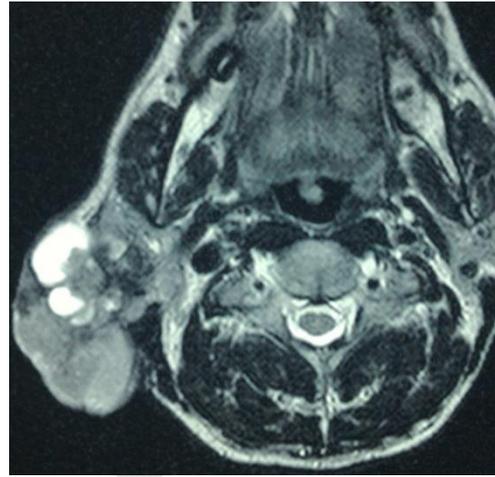


Figure 3. T2wi on MRI showing the arising from cystic or necrotic component. Solid component is iso- to hyperintense

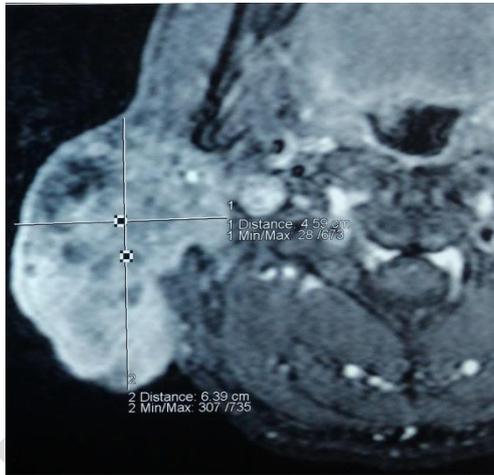


Figure 4 and 5 showing heterogeneous post contrast enhancement

FNAC revealed clusters of myoepithelial cells with vacuolated cytoplasm and few epithelial cell clusters. Mild atypia was seen in both groups of cells. There was no evidence of mitotic figures. The findings were suggestive of a benign or low grade malignancy.

The presence of skin involvement and lymph nodes suggested a high grade tumor. Consequently, the patient underwent total parotidectomy including wide local excision of the overlying involved skin (Figure 6). Facial nerve was identified and preserved. Selective neck dissection encompassing left level II, III and IV was done. The skin defect was reconstructed by posterior cervical advancement flap. Post-operatively, the patient developed grade 3 paresis of the facial nerve which recovered gradually over a period of four weeks (Figure 7).



Figure 6 showing the resected primary tumor at 3 weeks postop.



Figure 7 showing the facial functions

Histopathological examination of the resected specimen revealed tumor cells to be arranged in nests separated by thick fibrovascular stroma showing tubules, solid islands and cribriform pattern at places. Dilated ducts lined by adenocarcinomatous cells were seen. Focally, the tumor showed features of squamous cell carcinoma. Foci of spindling of tumor cells identified. Base of excision was formed by unremarkable salivary gland. This was suggestive of ductal adenocarcinoma with squamous cell differentiation with sarcomatous change. Two lymph nodes at level 2 showed evidence of metastasis. The patient underwent postoperative adjuvant radiotherapy. He has been disease free at 6 months of follow-up.

DISCUSSION:

Salivary duct carcinoma is a high grade malignant tumor, first described by Kleinsasser in 1968.¹ However, it was not until 1991, that SDC was formally recognised as a distinct entity by inclusion into the second version of WHO classification of salivary gland tumors.² Also known as cribriform salivary carcinoma of excretory ducts, it bears a striking histological resemblance to high grade ductal adenocarcinoma of breast. It is a rare tumor constituting around 1-3% of all salivary gland malignancies.^{2,3} The parotid gland is by far the most commonly involved (75%-88% of cases) with the submandibular gland and a combination of various minor salivary gland sites affected roughly equally.^{4,5} Most patients present after the age of 50 years. Even though it resembles ductal carcinoma of breast architecturally and cytologically, SDC is paradoxically more common in males. The male: female ratio is at least 4:1.⁶

SDC can occur de novo or as the malignant component of carcinoma ex pleomorphic adenoma. Microscopically, SDC is characterised by the presence of both intraductal and invasive components. The intraductal component is described as a cribriform, papillary, or solid growth pattern; often with comedo-like central necrosis whereas the invasive component consists of irregular glands and cords of cells that frequently elicit a prominent desmoplastic reaction.⁷ Delgado *et al.* classified SDC into three subtypes based on the degree of intraductal and invasive components. It has been seen that the tumors with predominantly

infiltrative or invasive components have dismal prognosis.⁸ Salivary duct carcinomas tend to be very high grade cytologically, with enlarged pleomorphic nuclei, coarse chromatin, prominent nucleoli, and numerous mitoses including many atypical forms. The cells have moderate to abundant amounts of eosinophilic to amphophilic cytoplasm, frequently imparting an apocrine-like phenotype.⁶

Several histological variants of SDC such as sarcomatoid variant, low grade or mucin rich SDC have been described, including documentation of their biologic behavior. Sarcomatoid SDC was first described in the year 2000 by Henley et al in three cases of parotid malignancies. The histology and nomenclature of this biphasic neoplastic variant has been a matter of debate for many years. Henley et al reported how each case was a composite of usual-type SDC and sarcomatoid carcinoma. While SDC showed typical cribriform architecture, anaplastic, spindled cells constituted the sarcomatoid areas. By immunohistochemistry they showed that the sarcomatoid component was immunoreactive to epithelial markers, thus, distinguishing this entity from “true” carcinosarcomas.⁹

Nagao et al documented the clinicopathological and immunohistochemical features of 8 cases of sarcomatoid SDCs. They recommended that for biphasic carcinomatous and mesenchymal neoplasms in the field of salivary gland pathology, the term “sarcomatoid variant of SDC” (sarcomatoid SDC) be used instead of carcinosarcoma when the carcinomatous component of those lesions is SDC and the term “sarcomatoid carcinoma, NOS” be used for lesions in which the carcinoma component is not a distinct type of tumor.¹⁰ Thus, sarcomatoid SDC is characterised by the presence of cohesive clusters and flat sheets of cells with a cribriform pattern, in combination with an atypical spindle cell component.¹¹

Histopathological examination of the tumor in our patient showed it to be a composite of epithelial and sarcomatoid elements. The epithelial or the SDC component was characterised by the presence of adenocarcinomatous cells lining dilated ducts, cribriform, growth pattern and tumor cells arranged in nests separated by thick fibrovascular stroma. The sarcomatoid component on the other hand was characterised by multiple foci of spindled cells with cytological atypia. Additionally, there was a focus of squamous cell carcinoma.

We could find only one report documenting the presence of squamous cell carcinoma component in sarcomatoid SDC. Mori et al reported a case of sarcomatoid SDC of submandibular gland in which they described the presence of atypical epidermal cell clumps with keratinisation, similar to our case.¹² SDC commonly arises from the parotid gland and clinically presents as a rapidly growing mass with a propensity to local recurrence and early distant metastasis. It is known to commonly involve the facial nerve with an increased tendency to perineural spread. Facial paralysis is observed in 40% to 60% of cases.¹³ Most treated patients with SDC present with early loco-regional recurrence with high propensity for distant metastases to the lung, bone and liver.² Seventy percent of the patients with SDC die of their disease within 3 years of diagnosis due to widespread metastases.²

Nagao *et al.* in their review of patients with sarcomatous SDC, observed cervical lymph node and distant metastasis in 33.3 % patients. They also reported their observation that the 5-year

survival rate and median survival time for patients with sarcomatoid SDC was less than those with conventional SDCs at their institution. The overall survival, however, was not significantly different. They concluded that sarcomatoid SDC is a highly aggressive tumor, similar to conventional SDC.¹⁰

Preoperative workup consists of MRI as the imaging modality and fine needle aspiration cytology. FNAC, although useful, is not always reliable in characterising SDC or its variants. FNAC was indeterminate in our patient and only raised the suspicion of a low grade malignancy. Although, there are no specific findings of this particular histology on MRI, it is helpful in the diagnosis of a malignancy, evaluation of its extent, lymph node status and involvement of surrounding structures and facial nerve. Involvement of skin and subcutaneous tissue, irregular margins, multifocal involvement, heterogeneous post-contrast enhancement and presence of necrotic lymph nodes at ipsilateral level II were sufficient to clinch the diagnosis of a malignancy pre-operatively.

The limited data in literature due to rarity of this malignancy dictates that no consistent therapeutic concept and protocol exists for this tumor entity. Most of the surgeons, however, prefer to treat patients of salivary duct carcinoma with radical surgical approach and adjuvant external radiotherapy.¹⁵⁻²¹ We recommend total conservative parotidectomy with selective neck dissection as the minimum surgery for this high grade entity. Regional lymph node metastasis occurs in 60% of cases and is often present at initial diagnosis⁵, thus, justifying neck dissection as an integral part of surgery even in patients without nodal disease at presentation. Due to scarcity of data about this tumor, it is difficult to evaluate the efficacy of adjuvant radiotherapy. While many authors favour postoperative radiotherapy for all patients with SDC, others recommend it based on the pathological stage, grade, margin status and perineural invasion.¹⁵ Ours was a patient with stage IV SDC, thus, necessitating adjuvant radiotherapy. Although, he has been disease-free so far, long term follow-up is recommended.

CONCLUSION:

Sarcomatoid variant of salivary duct carcinoma is a rare and aggressive form of salivary malignancy. The characteristic histopathological feature is the presence of typical SDC in association with atypical spindle cells. Total conservative parotidectomy with selective neck dissection followed by adjuvant radiotherapy is the minimum treatment of this malignant entity.

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