SALIVARY DUCT CARCINOMA: A CASE REPORT

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ABSTRACT

A Salivary Duct Carcinoma (SDC) is an uncommon high grade, aggressive malignant with high rate of recurrence having frequent perineural and lymphovascular invasion. Salivary duct carcinoma accounts for 1% to 3% of all malignant salivary gland tumours. About 200 cases have been reported in the English literature. Histologically SDC is similar to a breast ductal carcinoma. Due to rarity of this malignancy, no consistent therapeutic concept and protocol is available in literature. However, total parotidectomy with neck dissection and adjuvant radiotherapy appear to be appropriate treatment for a salivary duct carcinoma.

KEYWORDS

Salivary Duct Carcinoma.


INTRODUCTION

Salivary duct carcinoma is a rare tumour and accounts for 1% of all head and neck malignancies and 1% to 3% of all malignant salivary gland tumours. Although first described by Kleinssas in 1968, it was not included in WHO classification till 1991. Later, it was included in 2nd version of WHO classification of salivary gland tumours. SDC is most commonly found in older age male. It is highly malignant and aggressive tumour predominantly of the parotid gland and occasionally in submandibular glands. SDC has invasive growth with early regional and distance metastasis and should be differentiated from metastatic ductal carcinoma of the breast as well as high grade mucoepidermoid carcinoma, because of difference in clinical behaviour, treatment and prognosis.

CASE REPORT

A 53-year-old male patient presented to OPD with complaint of swelling in left parotid region since 6 months. There was history of pain in the swelling. On examination, a mass of size 5 x 5 cm present in the left parotid region. Swelling was nontender, firm and mobile. There was no palpable cervical lymph node and no abnormal findings in oral cavity. There were signs of facial nerve palsy. Chest X-ray and blood investigations were normal. USG revealed mass arising from superficial lobe of parotid gland with a diagnosis of benign salivary gland tumour superficial parotidectomy done. On gross examination, a 5 cm gland with 5 cm ill-defined, non-encapsulated, nodular lesion present. Microscopic examination showed malignant salivary gland neoplasm with closest resemblance to salivary duct carcinoma. Immunochemistry was positive for Her-2/neu and EMA.

DISCUSSION

Most reports indicate that SDC is an aggressive tumour with the most dismal short-term prognosis of all salivary gland tumours. Patients commonly present with a painless, rapidly growing parotid mass often with facial nerve involvement and cervical adenopathy. It frequently involves the extracranial portion of the facial nerve and has a propensity to metastasize through the temporal bone via perineural spread. Gingival metastasis has also been reported. Patients are usually elderly men with a mean age ranging between 55 to 61 years. A review by Jamal et al reported a male predominance of salivary duct carcinomas (Average 71%), and they most commonly occurred in the parotid gland (Average 87%) with only a few cases having been reported in the submandibular and minor salivary glands. This tumour has invasive growth with early lymphovascular and perineural invasion. Local recurrence and distant metastasis to the lung, liver and bone are frequent leading to a poor prognosis and are the most common cause of tumour-associated morbidity and mortality. Seventy percent of the patients with SDC die of their disease within 3 years of diagnosis due to widespread metastasis.
Salivary duct carcinoma can be established from pleomorphic adenoma (Gland carcinoma ex-pleomorphic adenoma) or de novo. It has also been reported to arise from long-standing chronic obstructive sialadenitis. Imaging findings, especially CT scan and MRI features are non-specific, but they are helpful in the diagnosis of malignancy and in the management. Positive diagnosis is based on histologic examination. The means of diagnosis consist of fine needle aspiration cytology, which is useful but not always reliable, and surgical specimen.

Histopathologically, SDC bears a striking similarity to ductal carcinoma of the breast and is composed of intraductal and invasive components. Similar to the breast carcinoma, the intraductal component may appear as a cribriform, papillary, or solid growth pattern, often with comedo-like central necrosis. The invasive carcinoma consists of irregular glands and cords of cells that frequently elicit a prominent desmoplastic reaction.

Delgado et al, based on the degree of intraductal or infiltrative component have classified SDC into 3 subtypes: 1) Predominantly intra ductal, where 90% of the tumour is intraductal; 2) Predominantly infiltrative, when less than 20% of the tumour is intraductal; or 3) Infiltrative when the tumour is entirely infiltrative. The significance of this classification is not known but it has been seen that the predominantly infiltrative tumour has dismal prognosis. Histologic variants of a salivary duct carcinoma include a low-grade salivary duct carcinoma, sarcomatoid variant, mucin-rich variant and invasive micropapillary SDC. Low-grade salivary duct carcinomas have a favourable behaviour in contrast to the aggressive pattern of high-grade salivary duct carcinomas. The sarcomatoid variant shows a biphasic pattern of salivary duct carcinoma and sarcomatoid elements and has highly aggressive behaviour just like high-grade salivary duct carcinomas.

The prognosis for a micropapillary salivary duct carcinoma is worse than that for a high-grade salivary duct carcinoma. Micropapillary invasive salivary duct carcinomas have a worse prognosis, while low-grade salivary duct carcinomas have a better prognosis. Perineural invasion and lymphatic invasion show no obvious prognostic significance. Some evidence revealed that salivary duct carcinomas arising from a pleomorphic adenoma may have a better prognosis than those originating de novo.

Immunohistochemical findings are not useful, but a constant overexpression of keratin, HER-2/neu, CEA and c-erb-B2 have been described. Frequently, androgen receptors and prostate-specific antigen expression have been reported. Some authors reported that Her-2/neu expression together with an elevated proliferation rate is associated with an unfavourable clinical course. Overexpression of c-erb-B2 was associated with a poor prognosis. In a study, all of the patients who overexpressed c-erb-B2 died of their disease in less than one and a half years, but the c-erb-B2 negative patient was still alive after 10 years.

Due to limited data and rarity of this disease, no consistent therapeutic concept and protocol exists for this tumour entity. Majority treat patients with SDC with a radical surgical approach and adjuvant external radiotherapy. If facial paralysis is present, a radical parotidectomy is mandatory. Recurrence was seen in all the patients reported in literature with parotid SDC who have undergone parotidectomy and did not undergo lymph nodal neck dissection.

CONCLUSION

SDC is a rare, highly aggressive tumour with the short-term prognosis and high incidence of early lymph node metastasis and frequent local recurrence after surgical excision. A total parotidectomy with neck dissection and adjuvant radiotherapy appear to be appropriate treatments for a salivary duct carcinoma.

REFERENCES

