CASE REPORT

Salivary Duct Carcinoma of the Parotid Gland: Report of Two Cases in Females

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ABSTRACT

Salivary duct carcinoma is a rare tumor, commonly involving parotid gland. It typically affects middle aged to elderly males. The tumor has an aggressive behaviour and is notorious for early metastasis, high rate of local recurrence, and high mortality. Treatment is mainly surgical but other modalities are also used. We are reporting 2 cases in females who had different presentation, course of disease, treatment, and prognosis.

Key Words: Salivary duct carcinoma. Malignant parotid tumors. Salivary gland tumors.

INTRODUCTION

Salivary duct carcinoma (SDC) is a distinctive primary neoplasm of the major salivary glands. It was first described by Kleinsasser et al. in 1968 and is characterised by remarkable resemblance to in situ and invasive ductal carcinoma of breast, hence described as SDC. About 200 cases have been reported in the English literature. The tumor has predilection for older men in the sixth to seventh decades of life. A number of patients experience facial nerve palsy and / or pain, and have cervical lymphadenopathy at presentation.1-3

SDC is characterised by an aggressive behaviour, early metastasis, local recurrence, and significant mortality. It has several histological variations and a rare variant, low grade SDC shows minimal invasion and better prognosis.2-4

CASE REPORT

Case 1: A 50-year female presented with a gradually increasing swelling in the preauricular region extending just below the right ear lobule for 3 years, which on examination was 3x3 cm non-tender, firm to hard in consistency, smooth surfaced with well-defined margins and mobile; but on teeth clenching, seemed fixed to underlying muscles. There were palpable lymph nodes in right anterior cervical chain. Fine needle aspiration cytology (FNAC) revealed high grade carcinoma such as SDC; and carcinoma ex-pleomorphic adenoma was not excluded. MRI reported a 4x2x2 cm, ill-defined heterogeneously enhancing, abnormal MR signal, right parotid mass with its major bulk lying in the superficial part of the gland, and level II and III cervical lymphadenopathy. Superficial parotidectomy with preservation of all branches of facial nerve was performed in 2014. Mild postoperative paresis of buccal branch of facial nerve recovered in 2 weeks. Histopathology confirmed SDC with clear resection margins and positive lympho-vascular invasion; on IHC it was positive for GCDFP 15, HER-2/neu and CD-117 while ER and PSA were negative. (Figure 1A and 1B). She underwent radiotherapy and is well after 2 years of surgery.

Case 2: A 68-year female presented with a gradually increasing swelling just below the right ear lobule for 4 years and was diagnosed on FNAC as pleomorphic adenoma, but she did not pursue any treatment. Her repeat FNAC was consistent with carcinoma (differentials were high grade muco-epidermoid and squamous cell carcinoma). CT scan showed 2.8 x 2.2 x 3.4 cm mass in superficial lobe of parotid gland with 2 adjacent lymph nodes (Figure 2). Superficial parotidectomy was done and histopathology reported SDC with involved excision margins, hence radiotherapy was advised. But a month after surgery, she developed a swelling at the operated site. CT scan showed a mass measuring 3.4 x 2.8 x 2.2

Figure 1(A): Histological features of salivary duct carcinoma on H&E. Figure 1 (B): The same tumor showing positive immunostaining for GCDFP 15.
cm involving right parotid gland with involvement of level II and V lymph nodes. CT neck in February 2012 showed a soft tissue mass in parotid area, extending to involve overlying skin and also deep lobe. MRI showed multilobulated 4.5 x 3.4 cm ill-defined heterogenous enhancing mass involving posterolateral aspect of right superficial parotid gland and an enlarged level II B right cervical lymph node. Her radical parotidectomy was performed in April 2012. Facial nerve was sacrificed and mass excised as well as defect closed with a rotation flap. On histopathology, she had a tumor 6.5 x 4.5 x 2 cm in size, consistent with SDC with a single intraparotid lymph node and involvement of medial margin of the excised tissue. She received 33 courses of radiotherapy followed by chemotherapy. Repeat CT on October 2012, showed a mass in superficial parotid lobe, 1.9 x 2.7 x 2.3 cm as recurrent growth and 0.46 x 0.61 cm pulmonary nodule at the apex of right lung. She died at home after 2 weeks and cause of death remained unknown.

DISCUSSION

SDC is a rare malignant tumor. It predominantly affects males (83%) between 55 - 65 years, commonly in the parotid gland (85 - 92%), followed by submandibular gland and other rare sites like intraoral minor salivary glands. It may arise from a pre-existing pleomorphic adenoma (27%).

SDC presents as a painless, firm mass, commonly associated with facial nerve palsy (40 - 60%) and cervical lymphadenopathy (35 - 73%). It frequently involves the extracranial portion of the facial nerve and has a propensity to metastasise through the temporal bone via perineural spread. Local recurrence and distant metastasis to lungs, bones, liver, brain and skin have been reported in 45 and 54% patients, respectively. Grossly, the cut surface is usually firm, grey white and may show areas of hemorrhage, cyst formation and necrosis. Microscopically, the tumor may have cribriform, papillary, solid with comedo-necrosis, invasive micropapillary or even sarcomatoid appearance. Low-grade and mucin-rich variants have also been reported. Cytologically, cells usually have an apocrine appearance, large nuclei and prominent nucleoli, moderate amount of eosinophilic cytoplasm and numerous mitotic figures. The role of immunohistochemistry is limited. The tumor cells express cytokeratin, EMA and CEA and frequently stain positive for GCDFP-15, CD117, PSA, PAP, androgen receptor and HER-2/neu. S100 is usually negative.

FNAC is useful but not always reliable, however, histopathology is the gold standard for diagnosis. Imaging techniques like CT scan and MRI are very useful for assessment of tumor dimensions, margins and status of lymph nodes involved. Differential diagnosis are high-grade mucoepidermoid carcinoma, adenocarcinoma not otherwise specified (ADC-NOS), oncocytic carcinoma, Warthin's tumor with nuclear atypia and acinic cell carcinoma. It is important to exclude metastatic carcinoma, particularly from breast, prostate and lung.

Therapeutic approach is non-consensual. Many authors recommend total parotidectomy even in T1 tumors because of life threatening local recurrence. If facial paralysis is present, a radical parotidectomy is mandatory. Surgery may include neck dissection. Adjuvant measures are recommended in aggressive tumor. Postoperative radiation therapy is indicated in cases of extraparotid extension, cervical lymph node metastases, positive surgical resection margins and lymphovascular and perineural invasion.

Chemotherapy with or without anti-androgen therapy for metastatic disease and treatment with anti-HER-2/neu monoclonal antibodies, such as trastuzumab may be an effective modality in advanced cases. Kourda et al. reported the utility of both anti-androgen therapy and chemotherapy.

The outcome is mostly unfavourable, as 45 - 65% of patients die at a mean duration of 3 - 5 years after diagnosis. Prognostic criteria are non-consensual consisting mainly of young age, tumor size greater than 3 cm, infiltrative tumor margins, local recurrence, lymphatic and distant metastases, necrosis, percentage of infiltrating and intraductal component, extraparotid extension, perineural invasion. histologic type, micropapillary variant and HER-2/neu overexpression. Both of our patients had different presentations, tumor behaviour and outcome.

REFERENCES

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