Mucoepidermoid carcinoma of the salivary glands are rarely found in childhood. We report the case of a 6-year old boy presenting with 2-month history of a submandibular swelling. In oral examination, a non-tender, firm mass was detected in the submandibular region involving the left cheek and left angle of the mandible. It was firm in consistency with obvious areas of multiple nodularity along the inferior border of the mass. Clinically there were no palpable lymph nodes. Radiographs revealed a radiolucent lesion in the left body of the mandible. The tumor showed sheets and nests of predominantly epidermoid cells along with occasional large pale mucous-like cells and frosted-glass appearance infiltrating the fibro-connective tissue. The lesion was diagnosed as mucoepidermoid carcinoma, high grade type.

Key words: Mucoepidermoid carcinoma, Submandibular gland, childhood

INTRODUCTION

Mucoepidermoid carcinoma (MEC) is a malignant neoplasms of the salivary glands which is rarely found in childhood. Its incidence rate has been reported to be 0.44 per 100,000. Most of them arise from the parotid gland (64 – 80%) while other locations that are less frequent include submandibular gland, sublingual gland, minor salivary glands and lacrimal gland. Submandibular gland cancer is extremely rare in childhood. MEC however is the most common malignant neoplasm of the salivary glands arising from the excretory ducts. At diagnosis MEC shows distant metastasis in less than 5% of cases, lungs being the commonest site.

Skin metastasis at diagnosis are exceptional event in high grade MEC of the salivary gland. The main therapeutic method in treatment of MEC like in most salivary gland malignancy is surgical resection. According to the literature, clinical stage, histologic grading and surgery margin status significantly correlated with the prognosis of MEC. In addition, some investigators reported that gender, age at presentation and primary site were also closely related to the survival of patients with MEC. In the present paper we describe a case of a submandibular gland mucoepidermoid carcinoma in a 6-year old boy.
CASE STUDY

A 6 year old male child (Fig1) presented with 2 months history of submandibular swelling which had rapidly increased in size. The informant who was the father claimed the swelling initially was about the size of an almond fruit and confined to left submandibular region before attaining the size at presentation. There was associated history of numbness of the left half of the lower lip of 2 weeks duration and painful mastication due to teeth mobility around the left lower jaw resulting into moderate weight loss.

Clinical examination revealed a child, not in any respiratory distress, moderately pale. There were no other positive findings in the general physical examination.

There was a lower jaw mass involving the submandibular region bilaterally with more prominence on the left side. There was involvement of the left cheek and left angle of the mandible. It was firm in consistency. There were areas of multiple nodularity along the inferior border of the mass. Clinically there were no palpable lymph nodes.

Intraorally there was a mass in the floor of the mouth which was not raising the tongue up. It was also firm in consistency. There was mobility of teeth in the left lower quadrant.

Fig 1: Picture of a 6yr old boy with mucoepidermoid carcinoma of the submandibular gland

Fig 2: Showing local invasion of the left body of the mandible from the mucoepidermoid carcinoma of the left submandibular gland

Fig 3: A photomicrograph of high grade mucoepidermoid carcinoma of the submandibular gland H&E X 125
Radiographs showed involvement of the left mandible with areas of radiolucency with irregular margins (Fig 2). Total body scan looking for distant metastasis revealed no significant findings. Laboratory evaluations including full blood count (FBC), liver function tests (LFT), and serum chemistry profiles were all normal. Chest X-Ray was negative for any signs of metastasis.

Incisional biopsy result revealed sheets and nests of predominantly epidermoid cells along with occasional large pale mucous-like cells and frosted-glass appearance infiltrating the fibro-connective tissue. These features are consistent with mucoepidermoid carcinoma, high grade (Fig 3).

**DISCUSSION**

In the literatures, few cases of malignant salivary gland tumors in childhood have been reported. They occur frequently in the adult age, with a peak incidence in the sixth decade of life. Only a few number of cases under the age of 14 years have been reported. Despite its rarity in children mucoepidermoid carcinoma (MEC) is still the most common malignant salivary gland tumor, comprising approximately 30% of all salivary gland malignancies. Parotid gland tumors comprised 0.08% of all primary cancers and 6% of all second cancers in childhood.

Submandibular gland involvement is extremely rare in childhood. In the submandibular glands, MEC is the second most common cancer in the general population, following adenoid cystic carcinoma. Previous exposure of children with childhood malignancies to irradiation or chemotherapy have been associated to risk of developing secondary malignancies of the submandibular gland. In the case reported there was no previous history of any exposure to either chemotherapy or radiotherapy, it may however be associated with a genetic predisposition.

The progressive, rapid growth and aggressive local invasion of the skin and the mandible with resultant teeth mobility are evidence of malignancy. A preoperative exploratory biopsy is not actually recommended for the danger of cell spillage and implantation. However an exception is represented by unresectable tumors in which the biopsy has the diagnostic value. The diagnosis of mucoepidermoid carcinoma and its grading can however be achieved only after histological examination. Skin metastases are an exceptional event in high-grade MEC (HG-MEC) of salivary glands. There was no evidence of distant metastasis. Metastasis in MEC is observed in less than 5% of the cases and the lung being the most commonly involved site.

MEC is primarily made up of three cell types in widely varying proportions: intermediate, mucous, and epidermoid. However based on prognostic indications MEC can be classified histologically as low, intermediate, and high-grade types.

High-grade tumors are poorly differentiated, and they are made up primarily of squamous epithelial and rarely intermediate cells. Low-grade tumors are well differentiated and are made up primarily of mucous-secreting and few intermediate cells or cystic space filled with abundant mucin. The histologic features of intermediate-grade tumors fall in between. It has been suggested that intermediate grade tumors has a clinical behavior closer to that of low grade tumors, they are composed of predominantly of intermediate cells and occasional mucous cells.

Higher-grade tumors show evidence of cytologic atypia, a high mitotic frequency, and areas of necrosis, and they are more likely to show neural invasion. Stromal hyalinization is common and sometimes extensive. Higher-grade MECs may be infiltrative or fixed as being observed in the case presented.

The prognosis of MEC is dependent on the clinical stage, site, grading, and adequacy of surgery. Pires et al. reviewed the literature and reported that overall 5-year survival rates ranged from 0 to 43% for patients with high-grade mucoepidermoid cancers of the salivary glands, 62 to 92% for patients with intermediate-grade tumors, and 92 to 100% for patients with low-grade tumors.

The reported case is a high grade tumor consisting primarily of squamous epithelia cells. High-grade MEC are invasive, difficult to excise with a local relapse rate of 25%, disseminating to distant sites in 30% of cases. Five-years survival rate is lower than 50%.

Treatment of MEC is primarily surgical, especially in patients with an early stage of disease. Mucoepidermoid carcinoma has been considered a radioresistant tumor. Radiation therapy alone however has been used in patients with advanced unresectable disease.
The use of adjunctive radiation has also been reported to improve loco-regional control but its influence on overall survival is not clear. In the case presented the tumor was regarded unresectable going by the clinical staging and histologic grading however radiotherapy was planned.

Chemotherapy can also be used in cases of locally advanced, metastatic or recurrent disease. The relationship between the primary site of the MEC and its prognosis is controversial. It has been reported that the differences in the primary site does not affect the survival of MEC, although submandibular MEC has been reported to have poor prognosis.

CONCLUSION

Mucoepidermoid carcinoma from a submandibular gland is extremely rare in children. A rapidly progressing swelling in a child gives a high suspicion of a malignancy. However incisional biopsy is of high diagnostic value. Staging, grading and site are great prognostic indicators in the treatment of this neoplasm.

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