Large Mucoepidermoid Carcinoma of the Maxilla in a Pediatric Patient

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ABSTRACT

Mucoepidermoid carcinoma generally arises from salivary glands, representing 5 to 10% of all salivary tumors. It is rarely seen in the jaws. The purpose of this paper was to report the case of a large mucoepidermoid carcinoma arising in the maxilla of a 15-year-old. (J Dent Child 2013;80(1):50-4)

Received August 4, 2011; Last Revision March 25, 2012; Revision Accepted March 25, 2012.

KEYWORDS: CENTRAL SALIVARY GLAND TUMOR, MUCOEPIDERMOID TUMOR, MINOR SALIVARY GLAND TUMOR, SALIVARY GLAND TUMOR

Mucoepidermoid carcinoma (MEC) is a relatively common salivary gland tumor. The first detailed description and histological analysis was done by Stewart et al. in 1945.[1] It is a common parotid malignancy (89% of all salivary gland malignancies) and the most common malignant salivary gland tumor in children.[2] Approximately 70% of these tumors are found in the parotid gland, 15% to 20% in the oral cavity (other than the salivary glands), and 6% to 10% in the submandibular gland.[2]

Ectopic salivary gland tissue may be found in numerous regions of the body, including the lymph nodes, ear, thyroglossal duct, pituitary gland, skin of the neck, mastoid bone, maxilla, and mandible.[3-5] Salivary gland neoplasms arising within the jaws as primary central bony lesions are extremely rare. MEC in the mandible is the most common of the intraosseous salivary gland tumors, occurring 2 to 3 times more frequently than in the maxilla. In the jaws, it is usually misdiagnosed clinically and radiographically as either a cyst or benign odontogenic tumor. Patients exhibit a few signs and symptoms suggestive of malignant growth, and radiographs often show multilocular or unilocular lesions with well-defined margins typically seen in benign lesions.[5] Therefore, their proper diagnosis is often in doubt.

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CASE REPORT

A 15-year-old female patient reported to the Department of Oral Medicine and Radiology with a 9 month history of progressively enlarged swelling on the right side of the maxilla (Figure 1). The mass was not painful but was becoming increasingly noticeable. The patient did not notice any anesthesia or paraesthesia in the involved area. Her medical and dental histories were unremarkable, except for the extraction of a mobile maxillary right second premolar four months earlier.

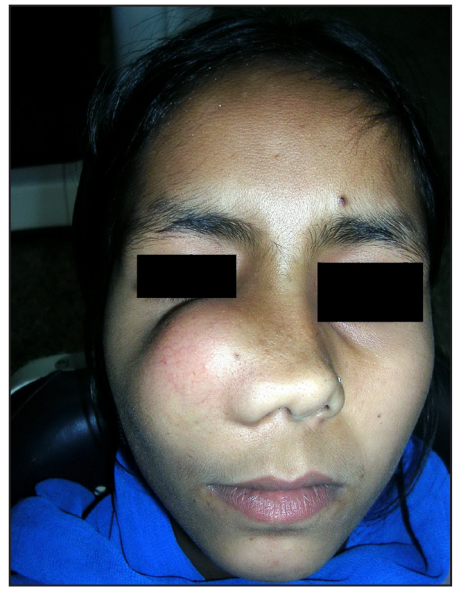

Figure 1. Frontal view showing swelling on the maxillary right side.

Upon extraoral examination, a single swelling in the right maxillary region was noticed, extending from the ala of the nose to the zygomatic region and vertically from the level of the corner of the mouth to the infraorbital margin. The skin over the swelling was normal. The swelling was nontender. Lymph nodes were not palpable. Intraorally, there was a palatal swelling on the right side extending from the permanent maxillary right canine to the right second molar crossing 1 cm across the midline (Figures 2 and 3). The consistency varied from hard to soft.

A sinus tract was present approximately 1 cm away on the palatal aspect of the permanent maxillary first molar. The buccal vestibule was also obliterated from the permanent maxillary right central incisor to the second molar. Another sinus tract was present on the buccal aspect of the permanent maxillary right second premolar. The fluid coming out of the sinuses was colorless, thick, and viscous.

A panoramic radiograph (Figure 4) showed a multilocular radiolucency extending from the posterior border of the right maxilla crossing the midline up to the permanent maxillary left lateral incisor. Superiorly it extended to the infraorbital rim and inferiorly to the alveolar ridge of teeth in the right maxilla. External root resorption of permanent maxillary right first and second molars was evident. A maxillary occlusal view (Figure 5) revealed a well-defined multilocular radiolucency on the right side of the maxilla crossing the midline. Expansion of the buccal cortical plate was observed. The nasal septum was deviated to the opposite side (Figures 6 - 8).

A CT scan study revealed a large, expansive, multilocular cystic, minimally enhanced lesion involving the right maxilla. The lesion was extending superiorly to the inferior orbital margin and the right ethmoidal sinuses with superior displacement of the floor of the right orbit. Inferiorly, it involved the maxillary alveolar ridge and was in close relation to the roots of the underlying teeth. Medially, it was obliterating the nasal cavity, with displacement of the nasal septum toward the left side. The lesion extended posteriorly to the pterygoid plates on the right side.

Considering these findings, a working diagnosis of ameloblastoma was made. Differential diagnoses of odontogenic myxoma, giant cell granuloma, odontogenic keratocyst, and mesenchymal/salivary gland tumor were considered.

A fine needle aspiration cytology showed tiny myxoid stromal fragments, occasional multinucleated giant cells, and anucleated squamous-like cells. An incisional biopsy, done under local anesthesia, was performed from the lesion's buccal aspect after reflecting the full thickness of the mucoperiosteal flap. The thin cortical bone was easily penetrated, and a piece of purplish brown gelatinous material was removed from a honeycomb-like cavity.

Histopathological reports showed that the tissue was composed of 3 types of cells: (1) mucous, (2) intermediate, and (3) epidermoid. Mucous or goblet cells formed solid masses and lined cystic spaces. Epidermoid cells

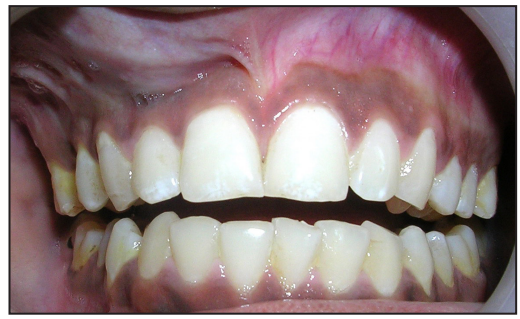

Figure 2. Intraoral view showing obliteration of the vestibule on the right side.

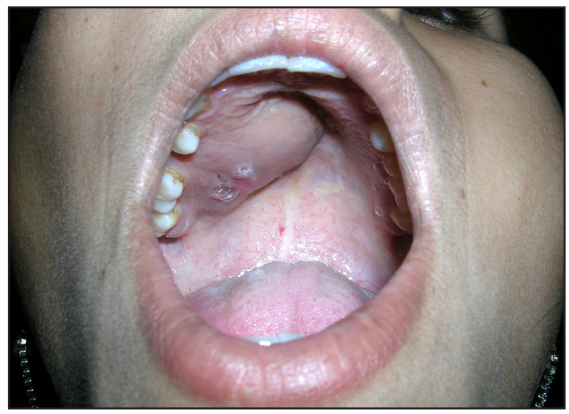

Figure 3. Intraoral view showing palatal swelling.

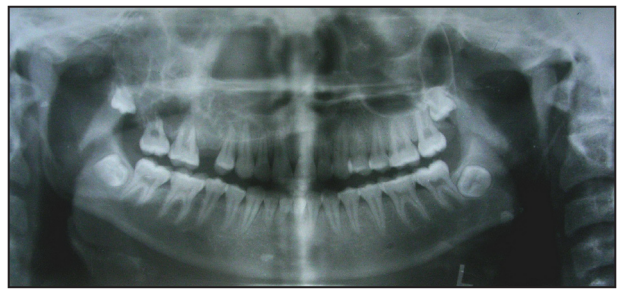

Figure 4. Panoramic view showing multilocular radiolucency involving the right maxilla and root resorption of the right maxillary 1st and 2nd molars.

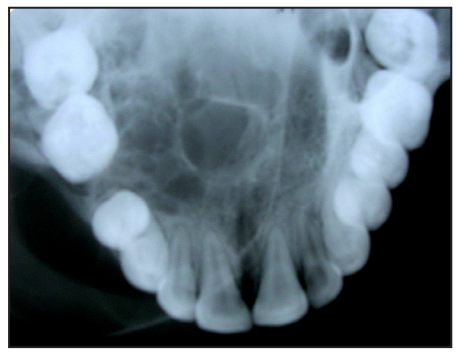

Figure 5. Maxillary occlusal view revealing a well-defined multilocular radiolucency on the right side along with expansion of the buccal cortical plate.

had identifiable intercellular bridges and were arranged as solid masses. Intermediate cells were small, with dark staining nuclei.

Mitosis was infrequent in mucous and intermediate cells. Islands of epidermoid and clear mucous cells were seen in a dense fibrous connective tissue background with scattered lymphocytes, neutrophils, and eosinophils. The mucous component was predominant, and there were large cystic areas in other sections. Based on the microscopic features, a diagnosis of mucoepidermoid carcinoma was made (Figures 9 and 10).

The patient was then scheduled for resection of the lesion, with adequate clearance followed by adjuvant external beam radiotherapy at a dose of 45 Gy over 20 fractions. To prevent radiation caries, the patient was instructed to increase her fluoridated water intake, maintain optimal oral hygiene, and wear a custom tray with 1% sodium fluoride gel for 5 minutes daily. After 1 year, the patient underwent secondary maxillary reconstruction using an ileac crest graft. The patient has been on a regular follow-up for the past 3 years, and there has been no evidence of recurrence.

DISCUSSION

The pathogenesis of MEC is still unknown. It generally has a female predilection, affects the salivary glands, and only rarely is located in the jaws. The peak incidence of occurrence is in the fourth and fifth decades of life. Primary central MEC of the jaws accounts for only approximately 2% to 4% of all MECs reported.[5,6] It is probable that odontogenic epithelium is capable of giving rise to mucous secretory cells and most likely serves as the progenitor for central salivary tumors of the jaws.[7]

Several theories have been proposed to explain the occurrence of salivary gland tumors in the jaws.[7] Four possible origins have been considered:[8-10]

1. entrapment of retromolar mucous glands within in the mandible, which later undergo neoplastic transformation;
2. embryonic remnants of submandibular and sublingual glands trapped within the mandible during development;
3. neoplastic transformation and invasion from the lining of the maxillary sinus;
4. neoplastic transformation of mucous-secreting cells from the epithelial lining of dentigerous cysts associated with impacted (unerupted) third molars.

More recently, intraosseous salivary tissue was demonstrated in approximately 1% of bone specimens of all jaw bones,[5] providing new evidences for the origin of intraosseous salivary carcinomas.[11] Histopathologically, the tumors are graded[12] for malignancy as:[12-14]

1. low grade—highly differentiated neoplasia with a predominance of macrocysts and microcysts, with presence of intermediate and mucin-producing cells;

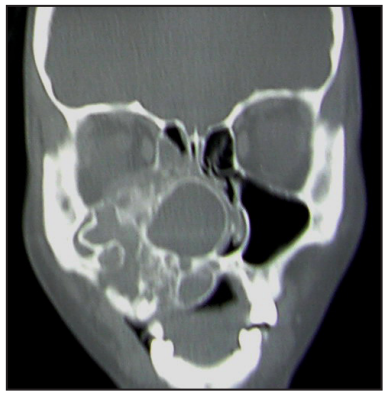

Figure 6. Coronal CT section showing multilocular radiolucency obliterating the right nasal cavity and displacing the nasal septum.

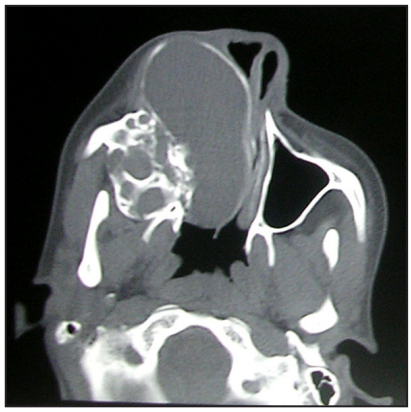

Figure 7. Axial CT section (bone window) showing the lesion crossing the midline.

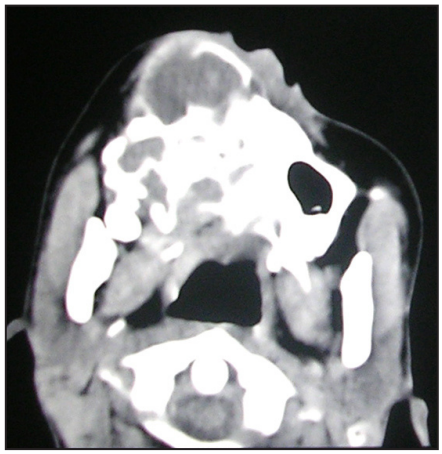

Figure 8. Axial CT section (soft tissue window) showing buccal expansion of the lesion.

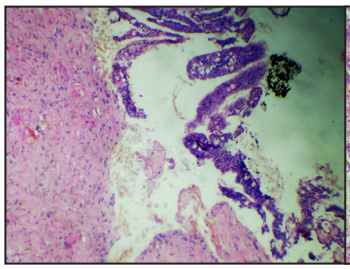

Figure 9. Low power view (10x) showing epidermoid and mucus secreting cells.

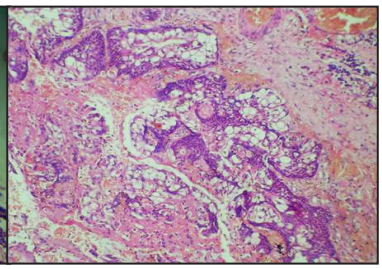

Figure 10. Epidermoid and clear mucous cells in dense fibrous tissue background (100x magnification).

2. intermediate grade—with a predominance of intermediate cells and a few cysts and presence of mucin-producing cells and islands of epidermoid cells;
3. high grade—poorly differentiated neoplasia with a predominance of intermediate and epidermoid cells in solid blocks, with mucin-producing cells present.

The commonly accepted criteria for diagnosis of central MEC include the following: (1) presence of intact cortical plates (cortical perforation, however, does not exclude primary intraosseous carcinoma type 4); (2) radiographic evidence of bone destruction; (3) histological confirmation; (4) positive mucin staining; (5) absence of a primary lesion in the salivary glands or elsewhere that can mimic an MEC's histological features; and (6) exclusion of an odontogenic tumour.[15-17]

Radiographically, all lesions appear as radiolucencies with reasonably well-defined, usually scalloped, margins. Lesions in the anterior mandible tend to have a soap bubble appearance, while posterior lesions more frequently manifest as a large vacuolated or cystic configuration.[18] A few lesions have been reported as diffusely destructive with vague or poorly defined borders.[18]

Brookstone and Huvos[7] have suggested 3 grades of classification for intraosseous MEC: stage I, without expansion and rupture of the cortical plate; stage 2, with expansion but without rupture of the cortical plate; and stage 3, with rupture of the cortical plates or presence of regional metastasis. In our case, the radiographic presentation was of a large multilocular lesion.

CT scans and MRIs also serve as additional diagnostic aids in assessing the extent of a lesion and involvement of vital structures such as the orbit, nasal fossa, neighboring sinuses, and pterygoid plates—particularly in the maxilla and lymph node metastasis.[19] The MRI has the same importance as in other salivary gland MECs to check for perineural or lymphovascular invasion. This affects the treatment and prognosis of the disease.

Surgery has been the predominant treatment for these lesions and has been categorized broadly as either conservative (enucleation, curettage, marsupialization, and marginal resection, with or without adjacent therapy) or radical (segmental resection, with or without treatment of associated neck and/or adjacent therapy).[7] It seems that radical resection offers the best chance of tumor eradication. Neck dissection is part of the treatment in cases where metastasis to the cervical nodes is suspected.[20] Freje et al.[21] recommend radiotherapy for high-grade tumors. Without the help of clinical staging, however, any conclusions about treatment are of limited value.[7] Long-term follow-up is required due to the possibilities of late recurrence or regional metastasis.[20,22]

Our case involved the entire right maxilla and right ethmoidal sinuses. There was also obliteration of the right nasal cavity, thus the case does not fall strictly into the category of primary central mucoepidermoid carcinoma. Other possibilities, however, such as metastases and odontogenic tumor were excluded. It is difficult to discern these lesions in the maxilla because, they become indistinguishable from those arising from the antral mucosa or entrapped salivary glands within the maxilla.[22] Without adequate examination of the resection specimen, it may be difficult to determine if a maxillary tumor actually arose within the substance of the maxilla or only represents intraosseous extension of a tumor originating within the sinus mucosa.[22]

CONCLUSIONS

This report describes a rare case of a large pediatric MEC involving the maxilla, an entity that must be included in the differential diagnosis of a multilocular maxillary lytic lesion. The rareness of this type of tumor in pediatric patients warrants careful and individual consideration for management. Considering the 3-year postoperative follow-up period, completeness of the resection, and absence of metastatic disease, a favorable prognosis is suggested for this patient.

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