Case Report

Advance Oropharyngeal Mucoepidermoid Carcinoma in a 9-year-old Boy: A Case Report and Review of Literature

Abdulrazak Ajiya, Iliyasu Yunusa Shuaibu[1], Mansur Adamu Yahuza[2]

Department of Otorhinolaryngology, Faculty of Clinical Sciences, College of Health Sciences, Bayero University, Kano, Nigeria, [2]Department of Radiology, Bayero University/Aminu Kano Teaching Hospital, Kano, [1]Department of Surgery, Division of Otorhinolaryngology, Faculty of Clinical Sciences, College of Medical Sciences, Ahmadu Bello University, Zaria, Nigeria

Abstract

Primary malignant salivary gland tumours are uncommon among the paediatric population, accounting for <10% of all head-and-neck tumours in childhood. Less than 5% of all salivary gland cancers occur during childhood, most of them are diagnosed over the age of 10 years and are histologically low or intermediate grade. Mucoepidermoid carcinoma (MEC) occurring in the oropharynx of children and arising from the posterior pharyngeal wall is rare and probably never reported. We hereby report a case of advanced MEC arising from the posterior wall of the oropharynx in a 9-year-old boy, managed via a transcervical excision in a resource-poor setting.

Keywords: Childhood, mucoepidermoid carcinoma, oropharyngeal

Introduction

Primary malignant salivary gland tumours are uncommon among the paediatric population, accounting for <10% of all head-and-neck tumours in childhood.[1] Paediatric major salivary tumours are well known to the otolaryngologist. However, minor salivary gland malignancies are quite rare and are usually found on the palate.[2] Less than 5% of all salivary gland cancers occur during childhood, most of them are diagnosed over the age of 10 years and are histologically low or intermediate-grade.[3]

Mucoepidermoid carcinoma (MEC) is a common malignant salivary gland neoplasm presumed to arise from the reserve cells of the salivary gland ducts.[3] Girls are mostly affected.[4] It is the most common malignant lesion and represents around 35%–50% of all malignant salivary gland tumours in childhood.[5]

MEC primarily involves the salivary gland but can also be found in the upper aerodigestive tract, tracheobronchial tree, lacrimal sac, thyroid and liver.[6] Only a few paediatric and adolescent cases have been well-documented in the minor salivary glands. A review of English literature in 2012 revealed only 15 well-documented cases of MEC of minor salivary gland origin in this age group.[7]

MEC occurring in the oropharynx of children and arising from the posterior pharyngeal wall is rare and probably never reported. We hereby report a case of advanced MEC arising from the posterior wall of the oropharynx in a 9-year-old boy, managed via a transcervical excision in a resource-poor setting.

Case Report

A 9-year-old boy with progressively enlarging left-sided neck mass referred from a secondary facility to our clinic with progressive dysphagia to solids, choking and difficulty breathing over a 3-month duration. The patient volunteered a history of left earache early in the onset of disease. There was no significant past medical history. Physical examination revealed an acutely ill-looking child, stridulous, in respiratory distress, dehydrated and drooling saliva. A huge fleshy mass

Address for correspondence: Dr. Iliyasu Yunusa Shuaibu, Department of Surgery, Division of Otorhinolaryngology, Faculty of Clinical Sciences, College of Medical Sciences, Ahmadu Bello University, Zaria, Nigeria.
E-mail: iyshuaibu@abu.edu.ng

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was seen occupying the oropharynx making it difficult to assess the anatomical subsites of the region. A diagnosis of upper airway obstruction secondary to the advanced oropharyngeal tumour was made and the patient had emergency tracheostomy with examination under anaesthesia and biopsy [Figure 1]. Histology revealed MEC. Computed tomography scan showed a huge ill-defined irregularly shaped exophytic moderately enhancing mass centred to the left oropharyngeal wall [Figure 2a]. It extends from the posterior pharyngeal wall to the nasopharynx superiorly obliterating the fossa of Rosenmuller and inferiorly into the hypopharynx. Anteriorly, the mass fills the vallecula and distorts the epiglottis and base of the tongue [Figure 2b]. No vascular encasement or bony destruction is seen. Extensive lymph node enlargement is seen in the deep and posterior cervical groups. Definitively, the patient had a transcervical excision of the tumour with selective neck dissection and excision of the left submandibular gland [Figure 3]. The intraoperative finding was that of a huge fleshy mass from the posterior pharyngeal wall of the oropharynx and palpable matted lymph nodes at levels Ib, II and III of the neck.

A 3 cm by 10 cm mass, weighing 240 gram was removed [Figure 4] and histology revealed sections showing infiltrating clusters of malignant epidermoid cells admixed with neoplastic mucous cells and intermediate type cells and disposed within a fibro collagenous stroma containing moderate inflammation.[Figure 5] The submandibular gland was unremarkable. The patient was transfused whole blood intraoperatively and was fed postoperatively via a nasogastric tube. Sutures were removed on the 8th post-operative day, the tracheostomy tube on the 13th day and the nasogastric tube on the 17th. The patient had a barium swallow on the 17th day post-operative, which showed the normal study. He was discharged on the 20th day post-operative. The patient had been on regular follow-up since then without a sign of recurrence.

Discussion

MEC was first described by Volkmann in 1895.[6] Although rare, MEC is the most common malignant salivary gland tumour in childhood accounting for 50% of cases.[7] The minor salivary glands are roughly around 500–1000 glands distributed throughout the oral cavity and oropharynx, mostly found in the palate, other areas where they are less commonly found include buccal mucosa, retromolar trigone, tongue and floor of the mouth.[2] In the present case, the tumour arises from the posterior wall of the oropharynx. This is the first reported of such to the best of our knowledge.

Most patients present with an isolated, painless and slow-growing mass. Our patient had a history of otalgia probably from the affectation of nerves in the pharynx. The clinical presentation of MEC may vary according to the histologic grade. Low and intermediate grade may appear as slow-growing masses; high grade generally appears as a rapidly enlarging mass.[8] The disease in our patient evolved

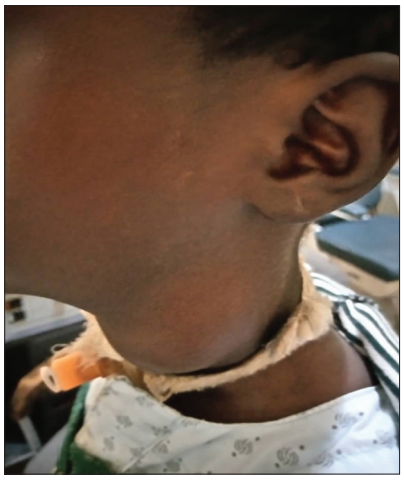

Figure 1: Huge neck mass and tracheostomy tube *in situ*

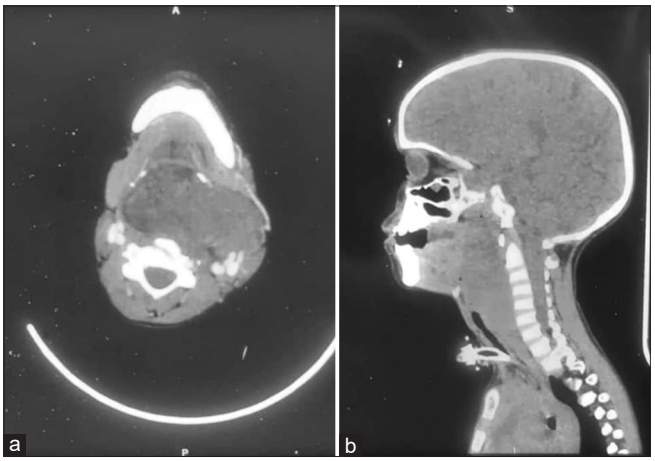

Figure 2: (a) A huge exophytic oropharyngeal mass. (b) Mass in the oropharynx extending to the nasopharynx, hypopharynx and vallecula

Figure 3: Transcervical tumour excision

just over 2 months. Enlarged cervical lymph nodes related to metastasis occurred in 30% of patients, but nodal enlargement was seldom found in patients with minor salivary gland neoplasms at presentation.[9] Our patient presented with neck

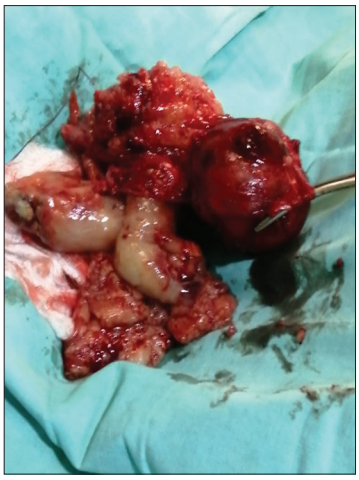

Figure 4: A huge excised mass

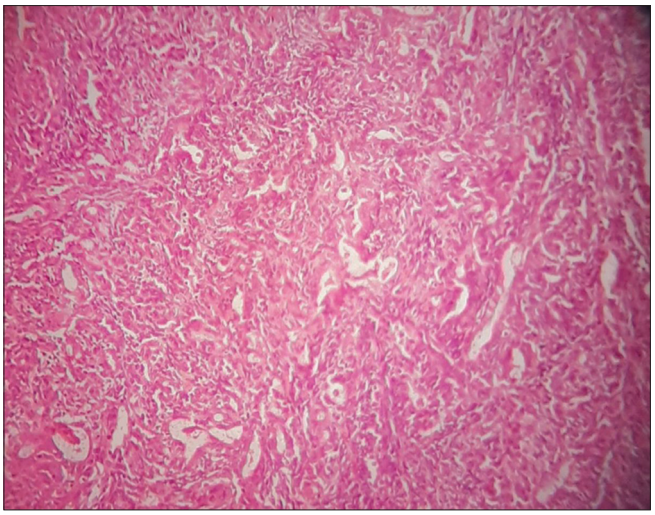

Figure 5: Intermediate grade Mucoepidermoid carcinoma showing islands of malignant epithelial cells as well as clear cells containing mucin. (H and E ×10)

nodes at multiple levels in the neck. Up to 5% of major glands and 2.5% of minor gland low-grade tumours were reported to metastasise to regional lymph nodes. MEC among children is commonly reported among girls.[4,10] On the contrary, our patient was a boy.

Diagnosis of MEC may be difficult based on imaging alone, and therefore, tissue diagnosis is usually needed. On computed tomography, low-grade tumours frequently appear as well-defined masses with moderate enhancement, but high-grade tumours exhibit ill-defined margins.[11,12] The scan in our patients showed an ill-defined but moderately enhancing mass.

Low-grade tumours are well-circumscribed and mucoid material is abundant. On the contrary, high-grade tumours have a greater epithelial component. Also, epithelial pearls and intercellular bridging may be found. In some higher-grade specimens, special stains may be required to demonstrate mucin production, thereby differentiating these tumours from squamous cell carcinoma.[2] The microscopic feature in our specimen showed more of the malignant epidermoid cells commensurate to a intermediate-grade lesion.

Complete surgical extirpation of the mass with free surgical margins is the treatment modality of choice in both children and adults. It is also the one that offers the best chance of local control of the disease.[9] Radiation therapy with 5500–6500 cGy is recommended for high-grade carcinomas, stage III and IV tumours, incompletely excised tumours and/or nodal involvement. However, strong indications should exist to justify post-operative radiotherapy in children, as adverse sequelae are common as high as 60%.[13] The role of chemotherapy in the treatment of salivary MEC remains speculative. Prophylactic neck dissection is not warranted for clinically negative neck, as recurrence is dependent on tumour stage and extent of surgical resection.[8] Our patient had complete surgical excision with selective neck dissection and has been referred for post-operative radiation therapy.

Low-grade tumours have recurrence rates of 15%–20% in contradistinction with 30%–40% recurrence rates seen among patients with high-grade tumours. The 5-year survival of 85%–95% for patients with low-grade tumours has been reported variously. On the contrary, patients with high-grade tumours have survival rates of 30%–50% from several reported series.[14] However, the prognosis appears to be better for children than adults. It is still early to be able to reliably comment on the prognosis in the index patient.

Conclusion

In the evaluation of children with oropharyngeal tumours arising from the posterior and lateral walls of the oropharynx with associated palpably enlarged cervical lymph nodes, MEC should be entertained. A transcervical approach to tumour excision with neck dissection with or without post-operative radiotherapy is a useful treatment modality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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