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Case report

Nasopharyngeal mucoepidermoid carcinoma in children

M. Re [a,*], E. Pasquini [b]

[a] Department of Otorhinolaryngology, Polytechnic University of Marche, Ancona, Italy
[b] ENT Department Sant'Orsola-Malpighi Hospital, Bologna University, Bologna, Italy

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ABSTRACT

Salivary gland tumours represent 1–4% of all human neoplasms and less than 5% occur in children and adolescents. Malignant salivary gland tumours only represent 0.08% of all childhood tumours and mucoepidermoid carcinoma (MEC) is the most common histological type.

The majority of MECs in the paediatric group are histologically classified as low or intermediate grade of malignancy, favouring a good prognosis.

Complete excision of the lesion with free surgical margins is the treatment of choice and the one that offers the best local control of the disease.

Experience with minor salivary gland carcinoma arising specifically within the nasopharynx is limited because nasopharyngeal MEC is an extremely rare malignancy and there is controversy regarding its optimal treatment.

We hereby report a case of mucoepidermoid carcinoma arising from the nasopharynx in a 7-year-old girl, which was managed via an endonasal endoscopic procedure.

To our knowledge the case we describe is the second case of nasopharyngeal MEC in paediatric age reported in literature and is the only one occurred in the first decade of life.

1. Introduction

Salivary gland tumours represent 1–4% of all human neoplasms and less than 5% occur in children and adolescents [1–4]. In the paediatric population the incidence of benign and malignant salivary gland tumours is similar [2,5,6] and malignant neoplasms are more common in the intraoral minor salivary glands [3]. Malignant salivary gland tumours only represent 0.08% of all childhood tumours [7].

Mucoepidermoid carcinoma (MEC) is the most common malignant salivary gland tumour in children and adolescents [8–10], accounting for about 25% of the epithelial tumours and slightly less than 50% of all malignant tumours [7].

Mucoepidermoid carcinoma (MEC) is a distinctly malignant epithelial salivary gland neoplasm that is divided into low, intermediate, and high grade types, based on its morphologic and cytological features [11]. Approximately 60% of MECs occur in the major salivary glands and 35% in the minor glands [11–13].

Children undergoing multi-agent chemotherapy and especially head and neck radiotherapy are at an increased risk of developing malignant salivary gland tumours and warrant long-term follow-up [7].

Most studies have shown that epithelial salivary gland tumours of children and adolescents are more common in females than in males and are more common in the second decade of life, their occurrence being rare in the first decade [1–3,8,14–17].

The clinical presentation of MEC may vary according to the histologic grade. Low- and intermediate-grade MECs may appear as slow-growing masses; high-grade MEC generally appears as a rapidly enlarging mass [3,11–13,18–22]. Regardless of the location, an asymptomatic swelling is the presenting symptom in 75% of cases. In patients with MEC of the minor salivary glands the tumours are often disguised as benign neoplastic and inflammatory conditions [12,13,18,19].

Enlarged cervical nodes, related to metastasis, occurred in approximately 30% of patients but nodal enlargement was seldom found in patients with minor salivary gland neoplasms at presentation [13].

Prognosis of MECs is a function of the histological grade, adequacy of excision, tumour grade and location and clinical staging [3,11–13,18–23]. Low-grade tumours have a 5-year survival rate of 90–100% although 5% of major gland and 2.5% of

* Corresponding author at: Department of Otorhinolaryngology, Polytechnic University of Marche, Via Tronto 10/A, 60126 Ancona, Italy. Tel.: +39 071 5964624; fax: +39 071 5964415.
E-mail address: remassimo@hotmail.com (M. Re).

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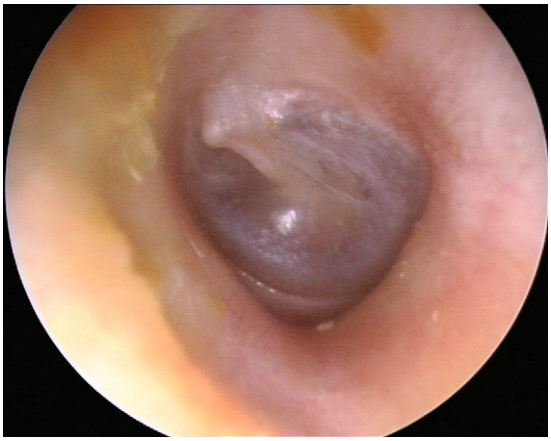

Fig. 1. Endoscopic evaluation of left ear with signs of chronic effusive *otitis media*.

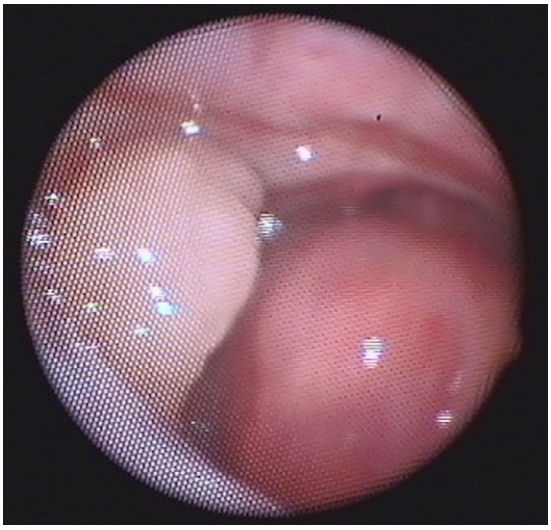

Fig. 2. The fibroendoscopic endonasal evaluation shows a firm exophytic submucosa neoformation of the nasopharynx. The contrast between the mucosa lining the tumour and the lymph adenoid tissue is evident; the left Eustachian tube is not visible.

minor gland low-grade tumours metastasized to regional lymph nodes or resulted in death [11,24].

The majority of MECs in the paediatric group are histologically classified as low or intermediate grade of malignancy, favouring a good prognosis [3,7,25].

Complete excision of the lesion with free surgical margins is the treatment of choice and the one that offers the best local control of the disease [3,11–13,18–22,24,26–31].

Radiation therapy is recommended for high-grade carcinomas, stage III and IV tumours and incompletely excised tumours [32]. The role of chemotherapy in the treatment of salivary MEC remains speculative [11].

MEC occurring in the nasopharynx is extremely rare and not well characterized.

We hereby report a case of mucoepidermoid carcinoma arising from the nasopharynx in a 7-year-old girl, which was managed via an endonasal endoscopic procedure.

2. Case report

A 7-year-old girl, with left chronic effusive otitis media resistant to medical therapy (Fig. 1), was referred to our clinic.

The fibroendoscopic endonasal evaluation showed a firm and exophytic submucosa neoformation of the nasopharynx with attachment area on the anterior margin of left Eustachian tube nasopharyngeal orifice (Fig. 2). The other head and neck findings were within acceptable limits. The child underwent a computed tomography (CT) examination which confirmed the presence of a left nasopharyngeal neoplasm without obvious aspects of erosion or infiltrations of the surrounding structures and of the left medial pterygoid process particularly (Fig. 3). No lymph nodes were observed in the latero-cervical regions.

The patient underwent the complete removal of the nasopharyngeal neoformation by endonasal endoscopic approach (Hopkins rod lens telescope, 0° view, 4 mm diameter).

For a better exposure of the nasopharynx, we removed the adenoid tissue and part of the tail of the left inferior turbinate with a microdebrider and then, through a subperiosteal dissection, we removed "en bloc" the lesion which appeared tough, well defined and of chondroid aspect. The left Eustachian tube nasopharyngeal orifice did not appear infiltrated but only compressed.

Extemporaneous histological evaluations of multiple biopsies from the tissue surrounding the lesion were negative. A nasal Merocel pack was left in situ for 1 day.

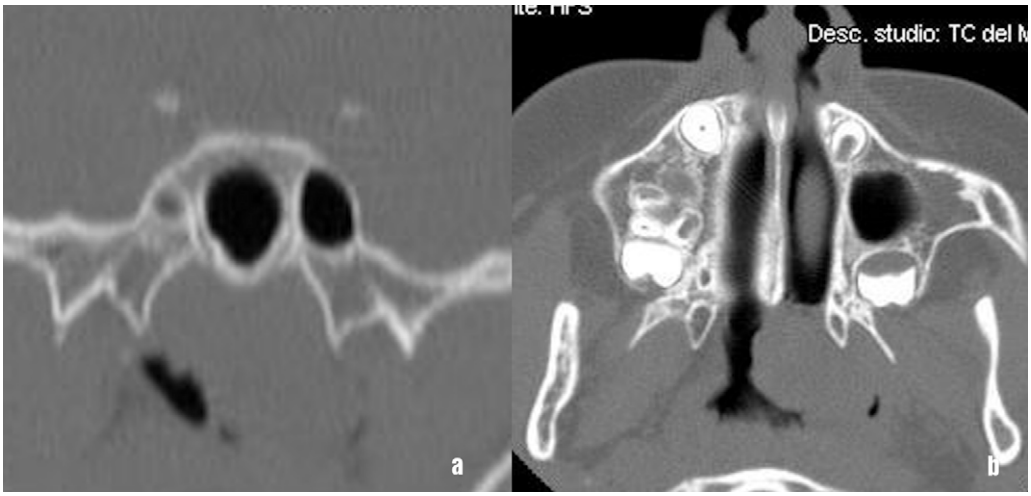

Fig. 3. Computed tomography (CT) examination shows the presence of a left nasopharyngeal neoplasm without obvious aspects of erosion or infiltrations of the surrounding structures, especially the left medial pterygoid process.