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Muco-Epidermoid Carcinoma of the Parotide in a Nine-Year Girl

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ABSTRACT

Mucoperidone carcinoma of the parotid is a very rare tumor and represents 1 to 4% of all parotid tumors. It can be observed at any age, especially in the adult to the 5th decade, but very rarely in the child, nineteen cases have been reported in the world literature, with an equality between the two sexes. We report the case of a 9-year-old girl admitted to consultations for a left lateral-cervical swelling that gradually increases in volume, and has evolved for 3 years. Cervical ultrasounds with an MRI were performed objectifying a tumor of the parotid gland. Total parotidectomy was performed. The extemporaneous biopsy was in favor of a low grade mucoepidemic carcinoma. The surgical procedure was completed by functional lymph node dissection. This is a slow-growing tumor with a non-specific symptomatology. The treatment is essentially surgical. Its prognosis depends on the histological features of the tumor.

Introduction

The parotid mucoperideroid carcinoma (CME) is a very rare tumor and accounts for 1 to 4% of all parotid tumors [1]. Its preferred location is the parotid gland, and can be observed at any age, especially in adults towards the 5th decade [2], but very rarely in children under 10 years [3]. Nineteen cases have been reported in the world literature, with an equality between the two sexes [3].

Observation

This is a 9-year-old girl with no significant pathological antecedent which has been present for three years in a swelling of the left parotid area which gradually increases in volume, painless, without a facial paralysis. The clinical examination finds a mass of the left parotid region, 3 cm in diameter, from the pretragian region forward and angulo-mandibular below, to the retro-auricular region behind and raising the lobule of The left ear, firm, with no inflammatory signs in view neither palpable adenopathy nor facial asymmetry. The endobuccal examination does not find any pus or blood from the orifice of the canal of the stenon. Examination of the scalp does not detect suspicious lesions. The cervical ultrasound showed a tissue formation developed within the left parotid, heterogeneous hypoechogene measuring 32 * 22 mm, poorly limited, with some images of calcification. MRI showed a lobulated left intra-parotid mixed mass with a capsule (Figure 1), with intra-parotid adenopathies of the jugulo-carotid lesion. The patient received a total parotidectomy with sacrifice of the facial nerve that was infiltrated by the tumor. The extemporaneous examination was in favor of a grade I mucoepidemid carcinoma, hence the surgical complement by homolateral jugulo-carotid II and III lymph node dissection (Figure 2). The histological study showed three types of tumor cells often entangled, mucosecreting cells, squamous cells and intermediate cells producing a muco-epidermoid carcinoma of low grade of malignancy. Disease was free from lymph node metastasis (Figure 3). The patient received a dose of radiotherapy of 55Gy in postoperative treatment, due to the invasion of the facial nerve. The suites were satisfactory.

Iconography

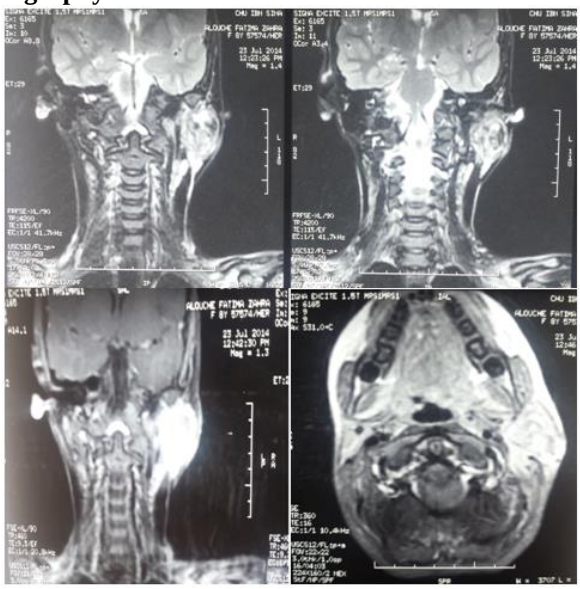

Fig. 1. MRI in coronal and axial section shows the tumor of the left parotid.

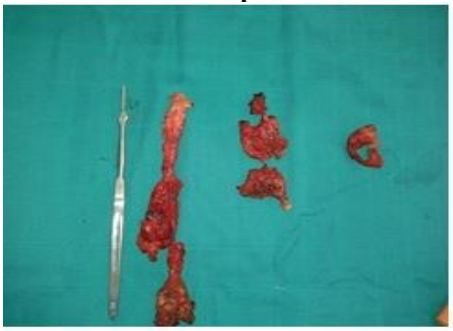

Fig.2 . Operative part of the parotid and lymph node dissection.

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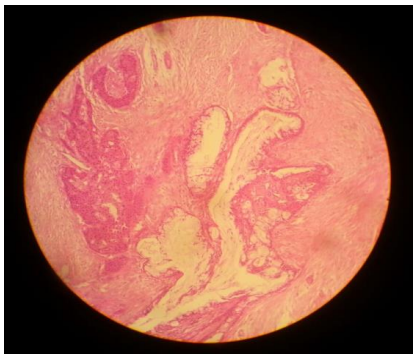

Fig. 3. Histological aspect of the operative part objectifying the double glandular and squamous component.

Discussion

Malignant tumors of the parotid gland are very rare in children. They mainly affect adults and the grandchild, exceptionally before ten years [3]. The first description of the mucoperideroid carcinoma of the parotid gland was made by Stewart et al. In 1945 [4]. The usual age of onset is between the 3rd and 5th decades [1]. However, some authors have reported cases in children. Three teams reported three cases of parenchymal CME in children four years, four and a half years and eight years respectively [3, 5, 6]. Given the rarity of this histological type in children under ten years of age, data from the literature are still insufficient, its management is modeled on that of the adult and its prognosis remains difficult to evaluate. The latter is good with a five-year survival rate greater than 90% [3]. Clinical presentation is not child-specific, and muco-epidermoid carcinoma often produces a solitary, mobile, parotid tumor reminiscent of pleomorphic adenoma, a slow, painless growth of small size (2-3cm) [1], With occasional relapses, rapid increase in volume and fixation to surrounding tissues without pain or facial paralysis. Morphological examinations, dominated by ultrasound and MRI, show a cystic lesion with no specific signs of malignancy. Diagnosis of malignancy may be suspected by cytopuncture and PET scan, however surgical exploration with histological study of the part remains the key to diagnosis [5, 6]. Histologically, mucoepidermoid carcinoma occurs in the form of double differentiated lobules, composed of glandular cells containing mucus, epidermoid cells and intermediate cells capable of differentiating into epidermoid cells or glandular cells [1]. There are 3 histological types: mucoepidermoid carcinoma well differentiated from low grade malignancy, mucoepidermoid carcinoma slightly differentiated from high grade malignancy and mucoepidermoid carcinoma of moderate differentiation: intermediate malignancy. Mucoperidone carcinoma of the parotid of the child has the characteristic of being often of low grade of malignancy, as in our child. Recent studies in immunohistochemistry have shown the expression of a gene encoding intercellular adhesion molecules called MEL-CAM (CD146) present on chromosome 11 in 92.7% of cases [7]. Its expression is more important depending on the degree of malignancy. The higher the grade, the stronger the expression. The treatment of choice of the CME of the parotid is the parotidectomy respecting the facial nerve. It should be personalized according to the signs of malignancy or deep localization that may lead us to sacrifice the facial nerve [3]. Lymph node dissection is indicated in high-grade tumors or the risk of lymph node metastases is greater than 50%. Postoperative radiotherapy, of all ages, has only limited indications to reduce the risk of local and regional recurrences.

Due to the complications it causes, and in a population with a long life expectancy, which is the case for children, radiotherapy is rarely indicated. It is therefore indicated in cases of malignant tumor of malignancy, perineural invasion, invasion of adjacent soft tissues or lymph nodes, and / or in the presence of an unresectable tumor residue. It will be practiced with great caution, protecting noble structures, and preference will be given to conformal radiotherapy with intensity modulation (IMRT) if available. Complications include xerostomia, trismus, osteo-radionecrosis and, above all, radiation-induced cancers [8, 9]. The role of chemotherapy in the management of muco-epidermoid carcinomas is not clearly defined; It is currently reserved for nonresectable or metastatic tumors. The molecules used are the same as for squamous cell carcinomas, namely cisplatin, bleomycin, fluorouracil and methotrexate [10].

Conclusion

The CME of the parotid in children is a very rare tumor whose diagnosis is histological. His treatment is basically based on surgery, associated with radiotherapy. It is a tumor that is characterized by different degrees of malignancy that condition its prognosis.

Conflicts of Interest

The authors do not declare any conflicts of interest.

Contributions of the Authors

All the authors contributed to the realization of this work.

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