Malignant fibrohistiocytoma of the parotid region. Report of a case

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Summary
Most salivary gland tumors are benign, malignant lesions accounting for 15-30% of the total. The most frequent site of salivary gland neoplasms is the parotid gland (80% of all cases). We present a case of malignant fibrohistiocytoma with atypical features due to its location (in the parotid region), size and rapid growth. Generally, this type of tumor arises in the lower limbs and in the abdomen. When located in the parotid gland, these lesions appear as a firm, slow growing and painless mass. Due to the low frequency of such lesions and their clinical behavior, the imaging study and fine-needle aspiration biopsy findings tend to diagnose them as pleomorphic adenoma. The definitive diagnosis requires microscopic study of the resection piece using immunohistochemical techniques. The treatment of choice is surgery, occasionally associated to radiotherapy. The success of treatment is dependent upon complete resection of the tumor - long term follow-up being necessary due to the risk of recurrence or distant metastasis.

Key words: *Malignant fibrohistiocytoma, sarcomas, parotid tumors.*

Introduction
Malignant fibrohistiocytoma (MFH) is an infrequent neoplasm that may arise in any body region, though it is generally found in the limbs (particularly the legs) and abdomen (retroperitoneum) (1). The term "malignant fibrohistiocytoma" was introduced in 1963, in reference to tumors composed of histiocytes and fibroblasts. At present, these lesions are considered to be a variant of high grade sarcoma. (2). MFH tends to develop in the skin in the form of small, slow-growing nodules, though its behaviour is more aggressive when the lesions are located in deep-lying tissues (3). The region of the head and neck is affected in 3% of cases (2,4), the nasosinusal tract being the most common location (1,2). Involvement of the parotid region has been reported in about 17 cases to date (5).

While the histological origin is unclear, it has been postulated that the tumor derives from histiocytes that morphologically and functionally differentiate to form fibroblasts (4).
The present study describes a case of MFH located in the region of the parotid gland.

Case Report
An 84-year-old woman presented with a steadily growing, firm and painless tumor in the right parotid region. The patient had been treated in the Department of Dermatology for actinic keratosis and dermatofibroma of the face two years before. The magnetic resonance imaging (MRI) and maxillofacial computed tomography (CT) findings revealed a nodular lesion in the right parotid gland, with

bilateral adenopathies of nonsignificant size. Puncture biopsy showed a hematic background with scant cellularity. Individual cells without atypia were noted, together with fusocellular elements. The CT, MRI and cytological findings suggested a mixed parotid tumor.

At surgery, a considerable increase in lesion size was observed, with evidence of skin involvement (Figure 1). A superficial parotidectomy was performed, with removal of the adjacent skin tissue (Figure 2a and 2b).

The histological study showed a fusocellular tumor with areas of hyalinization and abundant mitoses, occupying the gland and infiltrating the dermis – though without reaching the epidermis. Immunohistochemistry proved positive for vimentin, HHF-35, and focally for smooth muscle actin and S-100 protein. CD117, H-caldesmon, CD68, CD34, CD31, CKAE1/A3 and desmin proved negative. On the basis of these findings, malignant fibrohistiocytoma of the parotid gland was diagnosed (Figure 3).

Following hospital discharge, the patient was referred to the Oncology Department for neck radiotherapy (60 Gy over the surgical bed). One year after surgery the patient remains disease-free.

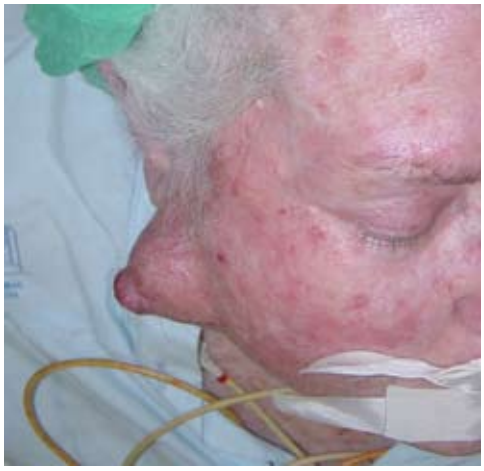

Fig. 1. Right parotid tumor with apparent skin involvement.

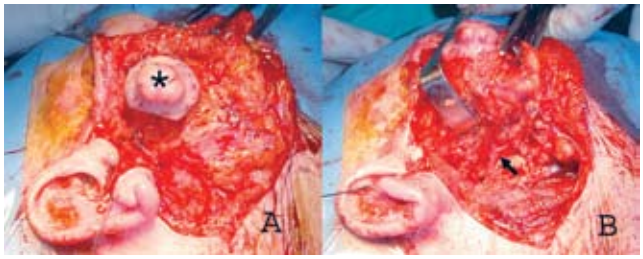

Fig. 2. Partial parotidectomy. 2a) Surgical field with resection of an affected cutaneous focus (asterisk). 2b) Resection completed with preservation of the facial nerve (arrow).

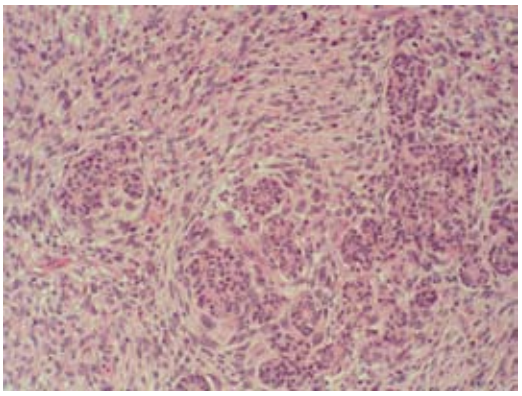

Fig. 3. Spindle cells arranged in short fascicles and showing a storiform pattern, with numerous mitoses. Remnant salivary ducts are present (hematoxylin-eosin, x100).

Discussion

Between 5-20% of all sarcomas are ear, nose and throat (ENT) lesions. In this context, rhabdomyosarcoma is the most common malignancy, followed by MFH. The latter is more frequent in adults and is very rare in early childhood (2). Males are more often affected (66%), and the maximum incidence is in the 50-70 years age range (1,6).

Location in the parotid region is not common; as a result, MFH is not usually contemplated in the differential diagnosis of parotid disorders. When located in this region, MFH tends to manifest as a steadily growing, painless mass – though facial paralysis may be observed in some cases (7). In clinical practice, it is frequently mistaken for pleomorphic adenoma – the definitive diagnosis being established by studying the surgical piece. This can occur with other types of infrequent tumors when they are located in this gland, such as solitary fibrous tumors (8), basal cell adenoma (9) or sebaceous adenoma (10), among others.

Atypical rapid growth was observed in this case, though this feature is not unusual (11).

The relapse rate is 16-52%, and is fundamentally determined by the presence of tumor-positive surgical resection margins (2). Initial aggressive surgical management is therefore indicated (12). Surgery is the first choice treatment. In this case, the absence of facial nerve involvement and the characteristics of tumor resection margins allowed preservation of the nerve. Radiotherapy is considered postoperatively, though some authors include malignant fibrohistiocytoma as a radioinduced tumor (6). Chemotherapy seems to be of scant utility (7)

Distant metastases appear in one-third of cases, involving mainly lung, bone and liver. Lymph node involvement is less common (6).

Despite its infrequency, MFH should be considered in the differential diagnosis of parotid gland tumors, comprising rhabdomyosarcoma, leiomyosarcoma, liposarcomas and pleomorphic carcinoma, among other lesions. Surgery is the management of choice, with or without radiotherapy.

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