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Malignant fibrous histiocytoma of the head and neck. Case report

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SUMMARY

Malignant fibrous histiocytoma (MFH) is a rare tumor included in the group of soft tissue sarcomas. Approximately 1-3% of these tumors affect the head and neck, with about 100 cases reported to date. A case affecting the retromolar trigone and involving the postero-lateral portion of the left hemi-tongue is reported. Etiological and pathological aspects are discussed. Diagnosis, treatment and prognosis of this neoplasm are reviewed.

KEY WORDS:

Malignant fibrous histiocytoma, head and neck, soft tissue sarcomas.

RÉSUMÉ

Le fibro-histiocytome malin est une tumeur rare faisant partie du groupe des sarcomes des tissus mous. Environ 1-3% de ce type de tumeur est localisé dans la région de la tête et cou, dont 100 cas ont été rapportés à ce jour. Le cas rapporté affecte le trigone rétromolaire envahissant la portion postéro-latérale de l'hémi-langue. Les aspects étiologiques et pathologiques sont discutés. Le diagnostic, le traitement et le pronostic sont passés en revue.

MOTS CLÉS:

Fibro-histiocytome malin, tête et cou, sarcome des tissus mous.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a rare tumor included in the group of high-grade soft tissue sarcomas. In spite of its rarity it is the most frequent of these tumours (40%) (Alvergard and Berg, 1989). MFH was first described by O'Brien and Stout in 1964. Approximately 100 cases have been reported to date (Bhuvanesh Singh *et al.*, 1993).

The most common locations are extremities and retroperitoneum, involvement of the head and neck being relatively rare, at 1-3% (Weiss and Enzinger, 1978). The first symptom depends on the tumor site, although it usually appears after a long period of time since the beginning of the pathological alterations (Bhuvanesh Singh *et al.*, 1993).

The tumor consists of fibroblast-like and histiocyte-like cells, often arranged in a storiform pattern (Bhuvanesh Singh *et al.*, 1993).

We report a case of MFH affecting the left retromolar trigone.

CASE REPORT

A 71-year-old caucasian male outpatient presented a three-month history of gum mass, that had lately become painful.

Personal background: perforated ulcer requiring gastrectomy 40 years before; smoking habit (approximately 20 cigarettes/day); upper and lower dentures for 7 years. The patient refered the biopsy of a lesion affecting the lateral region of the left hemi-tongue 6 years before, which revealed granuloma but no signs of malignancy.

Examination showed a 4 cm×2 cm exophytic tumor in the retromolar trigone that infiltrated the ipsilateral portion of the left hemi-tongue (Fig. 1). Cervical nodes were not discovered by palpation.

A deep biopsy of the lesion was performed. High-grade malignancy MFH was diagnosed. The tumor showed a fusocelular proliferation with atypical and pleomorphic multinucleated giant cells. There was substantial vascular neogenesis, as well as mixoid areas (Fig. 2). Immunohistochemical markers showed the tumor cells to be negative for cytokeratin, vimentin, factor VIII and S-100 protein.

In the ipsilateral prophylactic neck dissection all the cervical nodes showed sinusoidal histiocytosis, without signs of metastasic invasion. The patient received telecobaltotherapy (55.8 Gy) of the tongue and oral cavity. Curietherapy was subsequently applied on the primary lesion with threads of Iridium^{192} (14.4 additional Gy on the tumor; 35 cGy/hour), with a large security margin. The tumor disappeared macroscopically in a brief period of time.

At follow-up after 8 months, a whitish lesion was seen on the left retromolar trigone (Fig. 3). Tumor recurrence was confirmed by histopathology.

The patient died after 1 month.

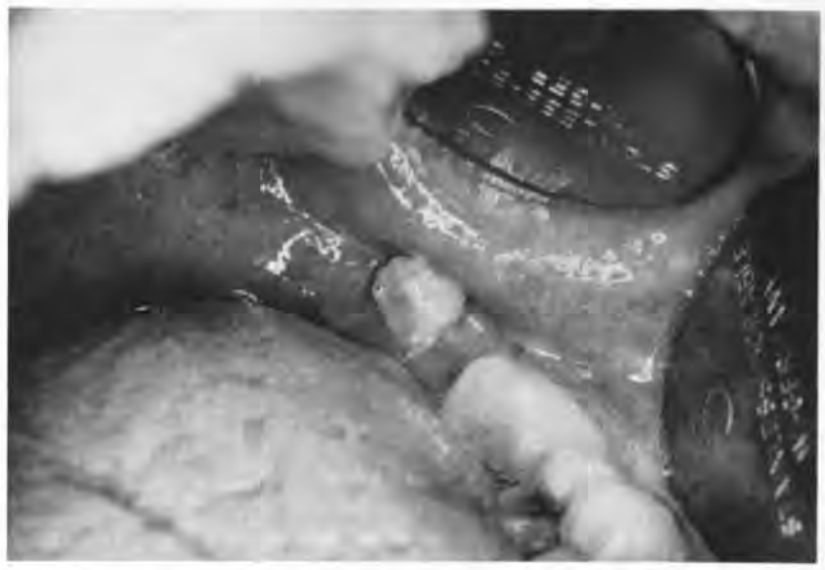

Fig. 1: Primary lesion.
Fig. 1: Lésion primitive.

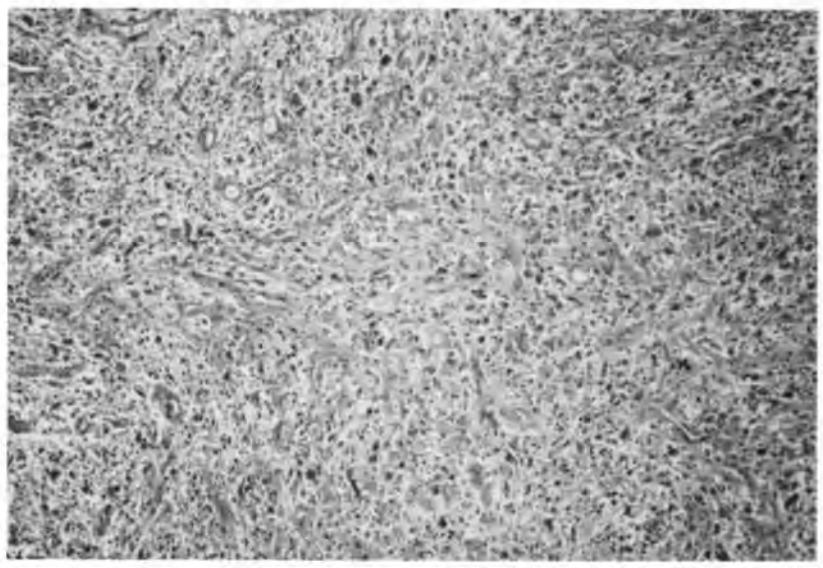

Fig. 2: Photomicrograph showing fibroblast-like cells and histiocyte-like cells.
Fig. 2: Histiologie de la tumeur montrant de cellules d'allure fibroblastique et d'allure histiocytique.

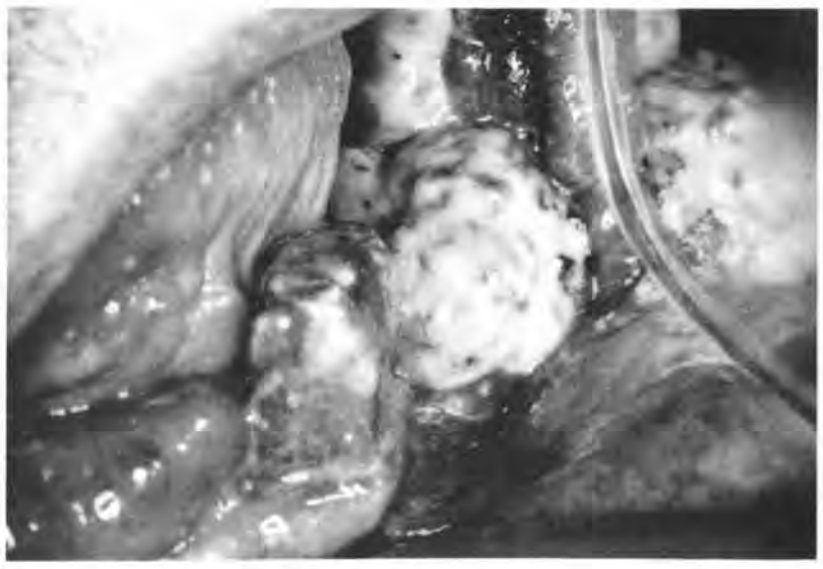

Fig. 3: Recurrent lesion (after radiotherapy).
Fig. 3: Récidive de la lésion après radiothérapie.

DISCUSSION

Malignant fibrous histiocytoma may appear at any age, but is more frequent between the fifth and seventh decades of life. It is more common among Caucasians, and exhibits a male predominance (about 3:1) (Sawyer *et al.*, 1993).

Although the etiopathogenia of the disease is open to controversy, its association to radiodermatitis has been postulated; in 75% of the cases reported to date MFH was seen to affect exposed areas. Gonzales-Vitale *et al.* have described a case of intracranial

MFH induced by radiotherapy (Gonzales-Vitale *et al.*, 1976). Other factors may be involved in the development of this neoplasm, such as repeated traumatisms, whose relationship to epidermoid carcinoma of the tongue has been well documented (Fretzin and Helwig, 1973). In this sense, poor dental conditions or inadequately fitting dental prostheses favor injury of the tongue.

Histology shows MFH to consist of fibroblast-like and histiocyte-like cells arranged in characteristic whorls. Both components exhibit variable pleomorphism and mitotic activity.

In this sense it was initially thought that the tumor derives from histiocytes which could differentiate into functioning fibroblasts. However, immunohistochemical evidence suggests that fibroblasts are the original cells involved (Wood and Enzinger, 1986).

Recent cell culture and tumor ultrastructural investigations (Fu *et al.*, 1975) have demonstrated the presence of a small number of undifferentiated mesenchymal cells that might also be regarded as the original cells in MFH.

MFH has been divided into 5 categories, as a function of the predominant histological characteristic involved. Thus, storiform-pleomorphic, myxoid, giant cell, inflammatory and angiomatoid variants have been reported. The storiform-pleomorphic pattern is the most frequent presentation, and all pleomorphic MFH belong to this category, including the present case. The myxoid variant in turn corresponds to approximately 25% of all cases of MFH (Das Gupta, 1983), while the angiomatoid form usually affects patients under 20 years of age (Enzinger, 1979).

The diversity of the histological presentation and the pleomorphic nature of MFH complicates pathological diagnosis. In this sense, differential diagnosis must be established with fibroxantoma, neurilenoma, granuloma, fibrosarcom, hemangiopericytoma, myoblastoma, histiocytosis X and pleomorphic rhabdomyosarcoma.

MFH affects the head and neck in only 1-3% of cases. The distribution rate of each location is shown in Table I. The clinical presentation depends on the tumor site, though patients are often initially asymptomatic. Nasal obstruction, epistaxis, pain, swelling of the check and headache may occur when the nasal cavity or paranasal sinuses are affected. The most frequent symptom in laryngeal involvement is hoarseness and occasionally airway obstruction. A painless enlarging mass is the most common presentation when MFH affects the oral cavity, parotid gland and neck (Bhuvanesh Singh *et al.*, 1993).

Only histopathological study is able to establish diagnosis. Immunohistochemical markers for histiocytes (e.g., alpha-1-antitrypsin) and mesenchymal cells are used to differentiate MFH from squamous tumors. Electron microscopy in turn reveals the absence of both tonofilaments and desmosomes, which are typical findings in epithelial neoplasms.

Important controversy exists as to the treatment of choice: wide resective surgery appears to bring the best results in small tumors. Although MFH appears to be encapsulated, simple enucleation results in recurrence in 44-73% of cases (Alvergard and Berg, 1989), for the tumor exhibits microscopic spread along the fascial planes and muscle layers beyond the limits of the apparent capsule. Extensive resection is thus recommended.

When MFH affects the head and/or neck, metastasis tend to involve the cervical lymph nodes. Consequently, neck dissection is adviseable in advanced tumors, and should always be performed when palpable nodes suggestive of metastasis are present.

A number of authors have reported better results when applying postoperative radiotherapy: Fagundes *et al.* in a series of 49 patients, observed no recurrences in patients with negative resection margins who received adjuvat radiotherapy at doses above 5000 cGY (Fagundes *et al.*, 1992). Although the results of treatment limited to radiotherapy have not been fully evaluated, total regression and the absence of recurrences has been reported in a number of cases. In this sense, Hayter *et al.* reported

TABLE I: Specific locations of MFH in the head and neck[17].
TABLEAU I: Localisations spécifiques de fibrohistiocytome malin au niveau de la tête et du cou.

Location	Percentage
– Sinonasal tract	30%
– Craniofacial bones	15-20%
– Larynx	10-15%
– Soft tissue of the neck	10-15%
– Major salivary glands	5-15%
– Oral cavity	5-15%

one case of maxillary MFH that was regarded as inoperable. Ten months after radiotherapy, histopathological evaluation demonstrated the absence of tumor cells (Hayter *et al.*, 1985). In the present case we adopted the same treatment, and recurrence occurred after 8 months.

Chemotherapy has also been used as the primary treatment modality. Leite *et al.* reported a good response to chemotherapy in 33% of patients with recurrence of MFH metastasis (Leite *et al.*, 1977). Adriamycin appears to be the most effective drug.

The evolution of MFH depends on several aspects. Thus, negative prognostic factors include tumor size (40% mortality in tumors over 6 cm in size), bone involvement (21% recurrences, 42% metastases and 37% mortality), and tumor depth or invasion. The pathological variant also influences survival — the pleomorphic presentation being more aggressive, with earlier metastases and poorer response to surgery (Block *et al.*, 1986). Location is decisive: tumors of the larynx, maxillary sinus and mandible have mortality rates of 44%, 40% and 30%, respectively, and thus involve the poorest prognosis (Bhuvanesh Singh *et al.*, 1993).

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