



A case report on inflammatory myofibroblastic tumor of the maxillary sinus

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ABSTRACT

Inflammatory myofibroblastic tumor is a rare lesion of uncertain etiology. It most commonly occurs in the lungs, but there are reports of extrapulmonary location. There is a documented different clinical behavior between the pulmonary and extrapulmonary lesions. Extrapulmonary location is characterized by unaggressive behavior and a tendency for recurrence. The treatment is based on surgery and adjuvant radiotherapy. We report the case of a 36-year-old patient suffering from IMT of the maxillary sinus.

Keywords: Inflammatory myofibroblastic tumor, Maxillary sinus, Etiology, Myofibroblastic proliferation, Internal calcifications, Metachronous primary lesion

INTRODUCTION

An Inflammatory myofibroblastic Tumor (IMT) is a rare lesion composed of myofibroblasts, fibroblasts, and inflammatory cells of uncertain etiology and disputed nosology. Because of these unknown factors, IMT was referred to by several different terms until the World Health Organization (WHO) classified IMT as a distinct entity in 1994. The tumor occurs most commonly in the lungs where it presents a genuine benign behavior. Its extrapulmonary location, although rare, has also been reported and is characterized by a different, more aggressive behavior, namely, the nasal cavity, paranasal sinuses, and pterygopalatine fossa. In this article, we present a case of a 36-year-old patient suffering from IMT of the maxillary sinus.

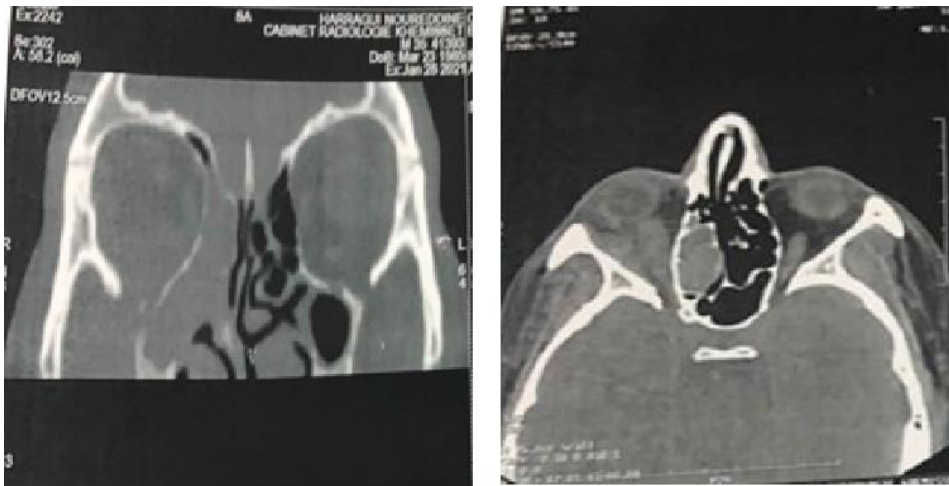
CASE STUDY

This is a 36 year old patient, who has presented for 2 months a right cheek mass, which

gradually increases in volume, without any

notion of nasal obstruction, without rhinorrhea or epistaxis. On nasal endoscopy, we note the presence of a translucent mass at the level of the right middle meatus. The CT scan reveals a right ethmoidomaxillary sinusitis with orbital extension (Figure 1). MRI found a locally advanced infiltrating right maxillary lesion process infiltrating the temporal, infra-temporal, masticatory space, and the orbital floor, associating a contralateral maxillary location (Figure 2). The patient underwent endoscopic endonasal resection of the mass (Figure 3). The histologic examination was in favor of a lesional process dominated by a fibro-inflammatory reaction, requiring phenotyping immunostaining to support the diagnosis. The immunohistochemical diagnosis was of an inflammatory myofibroblastic tumor.

The patient underwent adjuvant radiotherapy. The patient has been regularly followed up every 3 months with magnetic resonance imaging and chest radiographs. The evolution was favorable.



A

B

Figure 1. CT-scan (A) coronal sections (B) axial sections, Ethmoidomaxillary sinusitis with orbital extension.

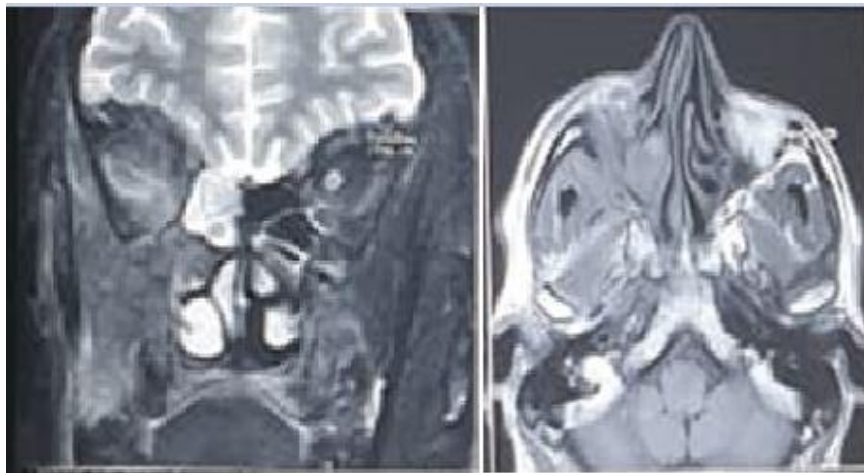


Figure 2. Nasopharyngeal MRI: voluminous expanding infiltrating process of the right maxilla, with heterogeneous signal, measuring 47 × 45 × 80 mm.

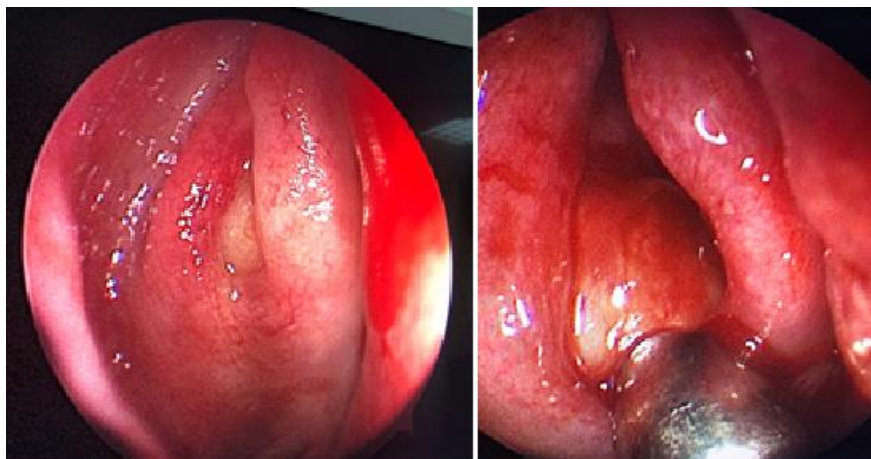


Figure 3. Intraoperative images.

RESULTS AND DISCUSSION

IMT is a myofibroblastic process associated with prominent infiltration of inflammatory cells, especially lymphocytes and plasma cells, in various patterns of myofibroblastic proliferation

(Kempson RL et al., 2001; Miettinen M 2003). Although the lesion was established as a single entity in 1994 by the WHO classification, because of its histologic, immunohistochemical and clinical diversity, IMT has been defined

with various terms related either to its histologic features or to its neoplastic origin and malignant potential. Thus, the terms used have been as follows: inflammatory pseudotumor, xanthogranuloma, histiocytoma, plasma cell granuloma, inflammatory fibromyxoid tumor, and myofibroblastic proliferation (Sciot R et al., 1997).

IMT occurs most commonly in the 2 first decades of life and it is rare over the age of 30 (Kempson RL et al., 2001). When the tumor arises in the lungs, which are the most common sites of tumor occurrence, it appears as a nodular lesion with completely benign behavior and the term inflammatory pseudotumor may be used as a synonym (Kempson RL et al., 2001). However, the occurrence of the tumor at extrapulmonary sites, though histologically the same, is accompanied by some special characteristics such as multifocal appearance, infiltrative local growth, vascular invasion, the tendency for local recurrences, and metastatic potential (Montgomery E et al., 2001; Watanabe K et al., 2000). All the above, in conjunction with some reports of clonal chromosomal changes (Sciot R et al., 1997) found in the tumor myofibroblasts, confirm the theories that IMT of the extrapulmonary type could be a neoplastic process (Karakok M et al., 2002) and the term inflammatory fibrosarcoma (IFS) was used to describe the most aggressive varieties with documented metastasis (Miettinen M, 2003; Watanabe K et al., 2000). It is worth mentioning that the WHO does not classify IFS as a single entity; however, based on the study by Meis and Enzinger (Meis and Enzinger, 1991) on 38 cases of IFS, several authors share the opinion that there is a difference between these 2 tumors (focused on the metastatic potential of each). Coffin et al. (Coffin et al., 1995) concluded that IMT should be considered an intermediate tumor with a weaker potential to recur and metastasize than other sarcomas and that the official term should be considered the most appropriate to describe this lesion.

Histologically, IMT is composed of inflammatory cells, myofibroblasts, and fibroblasts components that can range in each tumor from those dominated by spindle cells to those primarily comprised of inflammatory cells. Generally, there are 3 patterns of myofibroblastic proliferation (Kempson RL et al., 2001; Miettinen M, 2003). The first pattern, resembling granulation tissue or nodular fasciitis, is characterized by elongated myofibroblasts with variably abundant eosinophilic cytoplasm and large vesicular nuclei set within a loose or myxoid stroma. Typically, there are few plasma cells and the inflammatory component consists mainly of neutrophils, lymphocytes, and eosinophils. The

second pattern is more cellular, similar to fibrohistiocytoma. The fibroblasts and myofibroblasts are closely approximated, set within a compact stroma. These spindle cells may be also arranged in island formations surrounded by myxoid or hyalinized stroma with numerous normal mitotic figures. Lymphocytes are many and are sometimes arranged in follicles, but the predominant inflammatory cell type is the plasma cell. In the third pattern, reminiscent of a desmoid tumor, the stroma is densely hyalinized and cellular with a few plasma cells and lymphocytes within the stroma. In some rare cases, the predominant cellular population consists of large histiocyte-like or ganglion cell-like cells, which have large, round, vesicular nuclei with prominent nucleoli presenting a remarkable polymorphism.

IMT appears as a solid mass that may be accompanied, in 15% to 30% of cases, by general signs such as fever, hypochromic microcytic anemia, thrombocytosis, elevated erythrocyte sedimentation value, and hypergammaglobulinemia, all of which surprisingly resolve after treatment (Miettinen M, 2003). Apart from the lungs, it can arise anywhere in the body. As far as the head and neck region are concerned, IMT has been reported to occur in the larynx, orbit, mouth, parapharyngeal space, tonsils, thyroid, parotid, and lacrimal glands (Miettinen M, 2003). We found only 2 reports involving the maxillary sinus in the international literature (Karakok M et al., 2002; Gale N et al., 2003).

On computed tomography, IMT appears as a mild enhancing soft tissue mass without any internal calcifications. Bone destruction is unusual. Being a fibrohistiocytic lesion, it should be histologically differentiated from similar lesions such as malignant fibrous histiocytoma, nodular fasciitis, fibromatosis, fibrosarcoma, inflammatory histiocytoma, and leiomyosarcoma (Karakok M et al., 2002; Montgomery E et al., 2001; Watanabe K et al., 2000).

A simple excision is proposed and mainly applied treatment. However, there are documented reports of local recurrence up to 15%, (Miettinen M, 2003) an incident that can be reflecting inadequate resection of the lesion or tumors with behavior nearest to the previously described "IFS." The difficulty of completely excising the tumor in most cases is related to its location and its vicinity to valuable structures, which by definition are numerous in the head and neck area. The multiple recurrences of this reported case may be the result of the low resistance of the maxillary plates and the special features of the tumor such as rapid growth, the tendency to

bone infiltration, and resistance to radiotherapy. After studying 84 cases of IMT, Coffin et al. (Coffin et al., 1995) share this opinion and conclude that IMT can be a cause of death because of uncontrolled local growth.

Metastases are also reported, but the incidence is less than 5% (Karakok M et al., 2002; Kempson RL et al., 2001). However, bearing in mind the fact that extrapulmonary IMT is multifocal, it is difficult to distinguish between a real metastasis and asynchronous or metachronous primary lesion (Kempson RL et al., 2001).

Adjuvant radiotherapy has also been reported for the treatment of IMT, (Karakok M et al., 2002; Gale N et al., 2003) but because of the small number of reports, it is difficult to adopt safe theories. We believe that it should be used in cases where the tumor recurs rapidly and presents aggressive local behavior or when an inadequate resection is performed. However, even with such a treatment protocol, the tumor did recur, exhibiting a tremendous potential for local infiltration, resistance to combined therapy, and multiple recurrences.

CONCLUSION

IMT is a rare lesion of uncertain etiology. It most commonly occurs in the lungs, but there are reports of extrapulmonary location. There is a documented different clinical behavior between the pulmonary and extrapulmonary lesions, indicating a more aggressive behavior of the latter, which is characterized by a tendency for recurrence and a slight but existing metastatic potential. We concluded that IMT is a tumor with local malignant behavior, suggesting aggressive and combined therapy. Even with such a treatment plan, IMT may repeatedly recur.

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