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# Malignant fibrous histiocytoma in the right portion of the mandible with metastasis in pancreas



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## ABSTRACT

**BACKGROUND:** Malignant fibrous histiocytoma is a sarcoma of uncertain origin that can be found both in soft tissues and in bones. It is currently called undifferentiated pleomorphic sarcoma not otherwise specified and it represents a final common pathway in several tumors that are subject to the progress of dedifferentiation.

Local recurrence of the tumor in the same location where it was originated occurs in 20–30% of the total number of soft tissue sarcomas. It is less frequent in limbs and more likely to recur in retroperitoneal sarcomas and in head and neck.

Most tumors grow during the first two years after treatment.

Most sarcoma metastases, especially those in the head and neck (including malignant fibrous histiocytoma) present as a lung disease (90%). Extrapulmonary diseases are not frequent and can occur in lymph nodes (10%), bones (8%) and in the liver (1%).

**CASE REPORT:** 61 years old woman with history of malignant fibrous histiocytoma in the right portion of the mandible, resected four years ago. Subsequent checkups did not show any disease. The patient reports a constant abdominal pain not diagnosed, related to a pancreatic nodule.

**CONCLUSION:** Malignant fibrous histiocytoma is a potentially curable disease. The most important part of the treatment is complete surgical removal, usually followed by adjuvant radiation therapy. Chemotherapy is a treatment usually used only on those patients with higher recurrence risk or on those showing recurrence or who have already showed recurrence.

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## 1. Background

Malignant fibrous histiocytoma (MFH) is a sarcoma of uncertain origin found both in soft tissue and in bones [1–3]. It can appear as primary bone injury or as a result of a preexistent condition such as fibrous dysplasia or Paget disease [2,4].

MFH is the most common soft tissue sarcoma found in adults mostly aged from 20 to 70 years old [4,5]. Even though the MFH is considered the most common soft tissue sarcoma, it can also appear as a primary bone tumor [2]. Both fibrosarcoma and primary bone MFH differ from osteosarcoma in that they lack traces of malignant osteoid material [6], which can affect any bone, but in most cases, it affects the femur, followed by the tibia and the humerus [4,7].

Most patients suffer pain, swelling or a pathologic fracture [2,4,5]. In terms of radiology, tumors are not clearly defined lytic lesions and they can have a considerably soft tissue mass.

MFHs are more frequent in lower extremities and retroperitoneum and less frequent in the head and neck (3–10%) [2]. The nasal cavity and the paranasal sinuses are mostly affected (54%), and consequently, MFHs can affect the maxillary alveolar bone [2] and lead to laryngeal, maxillary, and mandibular sinus injuries. These are the most severe cases [2].

The incidence of MFH in the mandible is not frequent and incidence risk is only 3% of all MFHs in bones [2].

There are five different histologic subtypes: (1) storiform/pleomorphic, (2) myxoid, (3) giant cells, (4) inflammatory and (5) angiomatoid. The most frequent subtype is storiform/pleomorphic. Angiomatoid and myxoid present the most favorable prognosis of them all, whereas MFH with giant cells is the most harmful of all subtypes. The latter is considered a high-grade lesion in terms of local growth, recurrence and distant metastasis [2].

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Due to the variable range of histological differentiation, MFHs show no regular images in computerized tomography (CT) and magnetic resonance (MRI). In most cases, the tumor is observed as a round bulk with no clear sides and it always pervades contiguous bones and soft tissues. Density signal intensity is diverse. Necrosis can be observed in hyperintense areas as well as calcification and ossification (in 5–20% of these areas) [4,5].

Currently, there is no reported evidence of malignant fibrous histiocytoma having metastasized in pancreas and stomach.

The present work is reported in line with the SCARE criteria [8].

## 2. Case report

### 2.1. Personal history

61-year-old female patient, with no pathological history of drug use, illegal drugs, tobacco, alcohol abuse or genetic alterations, with a history of right lower jaw malignant fibrohistiocytoma, completely removed 4 years ago.

### 2.2. Pathology

#### 2.2.1. Macroscopic properties

Portion of right side mandibulectomy. A  $2 \times 2$  cm tumor formation with unclear edges can be observed.

#### 2.2.2. Microscopic properties

High-grade pleomorphic sarcoma (pleomorphic malignant fibrous histiocytoma). Clean margin.

Right submental node, jugular level 2 and facial without neoplastic cells.

Extension studies with chest CT were normal.

Ongoing adjuvant radiation therapy, no adjuvant chemotherapy according to the latest results from the European Organization for Research and Treatment of Cancer (EORTC) showing good tolerance and response to the indicated therapy.

Subsequently, craniofacial, neck, thorax and abdominal clinical and tomographic follow-up was performed, showing no pathology.

Three years after maxillary MFH resection, the patient consults for continuous epigastric pain of moderate intensity that evolved for a few days, without any other accompanying symptom.

The physical examination showed no alteration of the abdominal semiology. Laboratory and CT control is requested.

### 2.3. Laboratory tests

Erythrocytes  $4.1 \times 10^{12}/L$ ; hemoglobin 134 g/l; hematocrit 0.41; leukocytes  $5.3 \times 10^9/L$  (0.76 segmented neutrophils, 0.18 lymphocytes, 0.6 monocytes); urea 3.2 mmol/L; creatinine 92 mol/L; blood sugar 9.7 mmol/L; ionogram within regular values.

### 2.4. Additional procedure

Abdominal CT: partially necrotic  $3 \times 2 \times 7$  cm nodule with hypodense center in the tail section of the pancreas.

Normal CA 19.9 and CEA.

CT IMAGES

- Abdominal CT: pancreas, nodule image with peripheral contrast. Hypodense center

In view of the impossibility of an accurate etiological diagnosis, and in accordance with pathological antecedents, the general surgery department, together with the multidisciplinary committee, and the patient's consent, decides on surgical procedure.

### 2.5. Treatment

Diagnostic laparoscopy was performed showing negative results for peritoneal implants and hepatic metastasis. A 4 cm pancreatic tumor was found, possibly harmful for splenic vessels.

A laparotomy and a cavity evaluation were performed. Besides the tumor in the tail section of the pancreas, a 3 cm tumor in the body section of the stomach was also found. Splenectomy, distal pancreatectomy and partial gastrectomy with Roux-en-Y anastomosis were performed.

Samples are sent for anatomical pathology study.

Sample images

- Nodule in the body section of the pancreas.
- Lesion in the body section of the stomach.

#### 2.5.1. Clinical progress

Patient undergoes post-surgical treatment in the critical care unit showing positive clinical evolution. On the tenth day of admission, due to positive evolution, hospital discharge, with indications, and scheduled medical check-ups are indicated.

Two years after surgery, the patient continues under scheduled check-ups, showing no oncological disease, and with high risk of relapse, without a standardized adjuvant treatment.

#### 2.5.2. Histological report

Stomach and pancreas: high-grade pleomorphic sarcoma (pleomorphic malignant fibrous histiocytoma). COMPATIBLE WITH LOWER MAXILAR PRIMARY INJURY

### 2.6. Clean margin

Immunohistochemistry techniques with monoclonal antibody through the streptavidin-biotin-peroxidase method showing DAB chromogen on the slide showed the following: Negative pankeratine and CD45; positive lysozyme, vimentin, S-100 (only in giant cells) and CD68.

SLIDE IMAGES FOR PATHOLOGICAL ANALYSIS

- Immunohistochemistry of malignant fibrous histiocytoma.
- Histology of malignant fibrous histiocytoma.

## 3. Discussion

According to the World Health Organization, bone MFH is a malignant neoplasia composed of fibroblasts and polymorphic cells with a mostly storiform pattern [6]. Mandibular involvement resulting from MFH is not frequent (less than 30 cases reported) and is mainly located in the back of the mandible.

Soft tissues in extremities and in the retroperitoneum are usually the most affected [7]. A 70% of the cases start as primary tumors and 30% derive from preexistent conditions (such as radiation therapy on the affected area) [2,5].

The best choice is surgical resection with wide margins related to adjuvant chemotherapy [4–6]. Intralesional or narrow margin resection is related to high rates of recurrence and metastasis. Chemotherapy without surgical resection is not effective [2,5,7].

Local recurrence after exeresis is 16–52% and it depends on the size, spreading and positive microscopic margins [2]. Most recurrence cases occur during the first 2 years after treatment [7], and most cases with metastatic dissemination in sarcomas, particularly in the head and neck, are present as lung diseases (90%). Metastasis in extrapulmonary locations are not frequent and can occur in lymph nodes (10%), bones (8%) and in the liver (1%) [1].

Regarding histologic diagnose, with highly undifferentiated cells, it required a wide immunohistochemistry panel to deter-



**Fig. 1.** Abdominal CT: pancreas, nodule image with periferal contrast. Hypodense center.



**Fig. 2.** Abdominal CT: pancreas, nodule image with periferal contrast. Hypodense center.

mine tumor cell lines. Considering that pankeratineand CD45 were negative, squamous cell carcinoma and lymphoma were discarded, respectively. Positive lysozyme and vimentin contribute to the diagnose, since they indicate the presence of histiocytes and mesenchymal cells. S-100 is positive, but only in multinucleated giant cells, which are seen among several polimorphic cells. Positive CD68 in cytoplasm of several cells is specific for histiocytes in general and, int his case, defines the diagnose (Figs. 1–7)

Currently, there is no reported evidence of MFH having metastasized in pancreas and stomach. This work offers the possibility of considering new sites of metastasis in the bibliography.

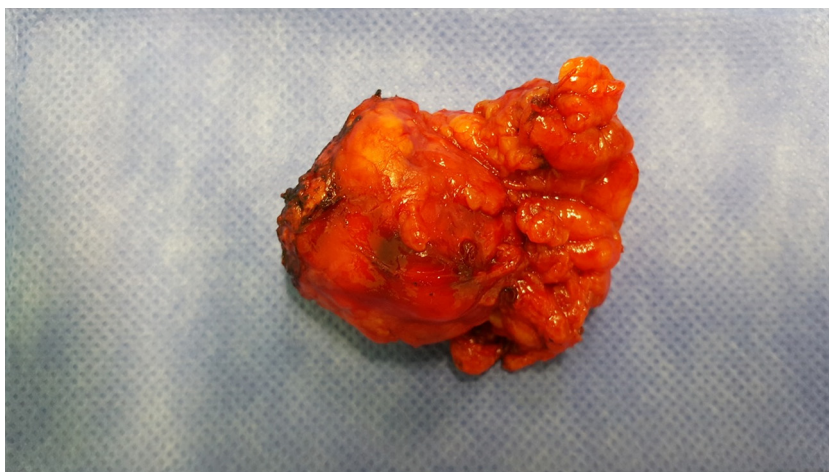
**4. Conclusion**

The case presented is interesting since the primary tumor located in the right portion of the mandible, as well as location for metastasis are not frequent. In addition, it is relevant to mention that after 3 years with no disease there is no metastasis in lungs, which occurs in 90% of the cases, nor in the liver.

There is no bibliography on metastasis in pancreas and in stomach as it is in this case.

Treatment for this pathology requires a multidisciplinary and well elaborated approach, as the time it takes to choose a treatment since the diagnose is made is extremely important.

This encourages and shows us that medicine is a wide field of study and every day we can discover new cases in which tumors behave in unusual ways and affect unusual organs drawing our attention to scenarios that may seem unlikely or inexistent up to that moment.



**Fig. 3.** Nodule in the body section of the pancreas.



Fig. 4. Lesion in the body section of the stomach.

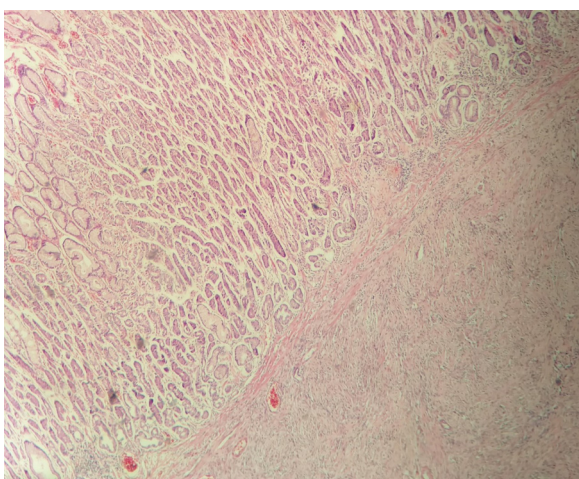


Fig. 5. Histology of malignant fibrous histiocytoma.

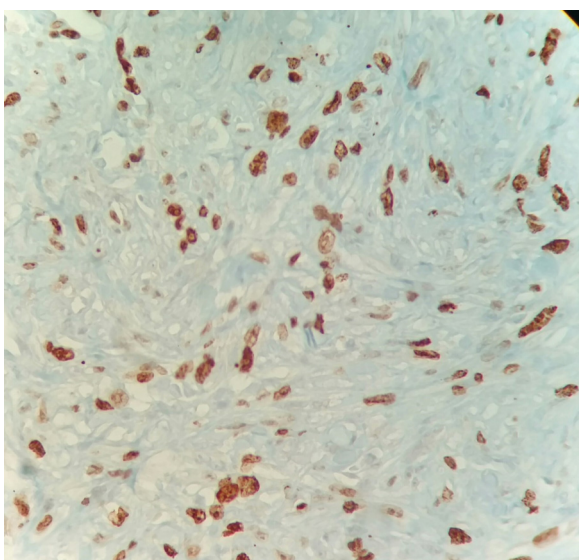


Fig. 6. Immunohistochemistry of malignant fibrous histiocytoma.

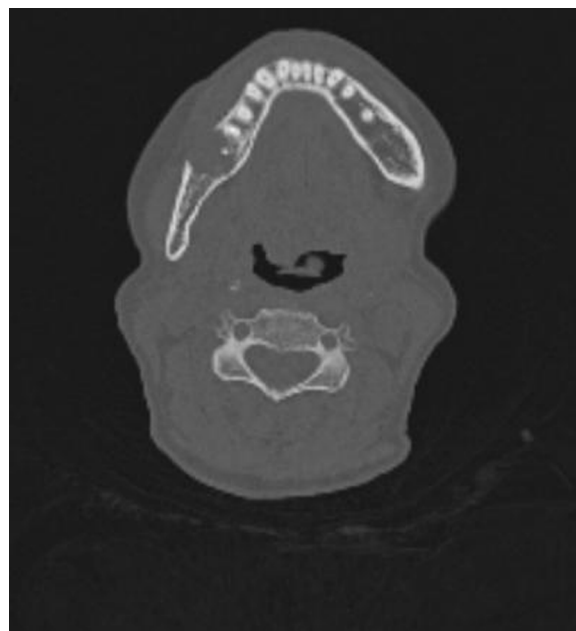


Fig. 7. Malignant fibrous histiocytoma in the right portion of the mandible.

**Conflicts of interest**

None.

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**Ethical approval**

The patient’s family have given their consent for science. I sent the informed consent.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Javier Biurrun Chamale.  
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