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Metastatic Primary Gastric Histiocytic Sarcoma: A Case Report and Clinical Insights

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Abstract Case Report

Histiocytic sarcoma (HS) is an exceedingly uncommon lymphohematopoietic tumor characterized by an aggressive clinical trajectory and poor treatment response. The diagnosis depends on verifying its histiocytic origin and ruling out other undifferentiated cancers. Hodgkin's Sarcoma is often located in lymph nodes, with the gastrointestinal tract being the most prevalent extranodal location. As of yet, only a limited number of stomach histiocytic sarcomas have been documented. We discuss the case of a 64-year-old patient who exhibited persistent abdominal discomfort for two months and significant decline in overall health. The findings from oesophagogastroduodenal fibroscopy and biopsy, corroborated by immunohistochemistry analysis, indicated stomach histiocytic sarcoma. The thoraco-abdomino-pelvic CT scan indicated a stomach tumor with metastasis to the liver. The patient was sent to oncology for chemotherapy treatment. Timely identification and precise diagnosis of this uncommon neoplasm are crucial, since they might influence patient prognosis. The collection and assessment of stomach histiocytic sarcomas is crucial for advancing prognosis and therapeutic therapy.

Keywords: Histiocytic Sarcoma, Stomach, Case Report, Immunohistochemistry.

INTRODUCTION

Histiocytic sarcoma (HS), also known as true histiocytic lymphoma, is a very rare lymphohematopoietic malignancy occurring mainly in adults, and tumor cells display the morphological and immunophenotypic characteristics of mature histiocytes [1, 2]. Histiocytic sarcoma tumor cells are positive for a histiocytic marker (CD68, CD163 or lysozyme) and negative for a Langerhans cell marker (CD1a or langerin). And they are generally negative for markers specific to B cells (PAX5 or CD19), T cells (cytoplasmic CD3) and myeloid cells (myeloperoxidase). HS usually occurs in lymph nodes, skin and a number of other extra-ganglionic sites, among which sites in the intestinal tract are common [3]. To date, only a small number of gastric histiocytic sarcomas have been reported. In the present study, we reported a case of primary histiocytic sarcoma in a man who consulted the gastroentero-hepatology department at Arrazi Hospital, CHU Mohammed VI, Marrakech. We also performed an updated systematic review to summarize the clinical features of gastric histiocytic sarcoma.

CLINICAL CASE

This is a 64-year-old patient with a history of intermittent melenas, several episodes over the past 6 months, never investigated, and no known family digestive or extra-digestive neoplasia. Presented with chronic abdominal pain and profoundly altered general condition. Whose symptomatology began 2 months after admission with the onset of abdominal pain in the epigastric area and in the CDH, in the form of cramps of moderate intensity with no irradiation or analgesic position, and no other digestive or extra-digestive signs, all assessed in a context of asthenia and significant weight loss (15 kg in 2 months). Clinical examination on admission revealed a conscious patient with a WHO performance score of 1 and a BMI of 16.2 kg/m2. Blood pressure was 110/60 mmHg. The heart rate was 80 beats per minute and the respiratory rate 16 cycles per minute, with discoloured conjunctivae. Abdominal examination revealed diffuse abdominal tenderness accentuated in the region of the CDH and epigastrium, no signs of portal hypertension, no signs of hepatocellular insufficiency, no palpable mass, no organomegaly. The rest of the somatic examination was unremarkable, notably no palpable adenopathy.

The initial laboratory work-up revealed normocytic normochromic anemia at 9.4 g/l, a

prothrombin level of 80, and an inflammatory syndrome with hyperleukocytosis at 12700 elements/mm3 and CRP at 70. Carcinoembryonic antigen and 19-9 carbohydrate antigen were negative. In view of this clinical picture, an oesogastroduodenal fibroscopy was performed, which revealed an infiltrating ulcerating tumour process extending from the lesser curvature to the angulus (figure 1).

Biopsy of the gastric process showed malignant tumour proliferation.

Tumor cells were monomorphic to pleomorphic with a fusiform shape. Tumor cell cytoplasm was abundant and eosinophilic (Figure 2).

An immunohistochemical study was performed. Tumor cells were positive for anti-CD68, anti-CD15 and the Ki67 proliferation index was very high, estimated at 80%. In addition, they were negative for the epithelial cell marker cytokeratin AE1/AE3, the Langerhans cell marker CD1α, the dendritic cell marker CD35 and other markers including CD20, follicular, smooth muscle actin (SMA), ALK and S100.

On the basis of histological and immunohistochemical examinations, the malignant tumor of the stomach was diagnosed as a histiocytic sarcoma.

Abdominal ultrasound showed a normal-sized liver with regular contours and several heterogeneous hypodense nodular lesions (Figure 3).

A thoraco-abdomino-pelvic CT scan was performed as part of the extension work-up, which was in favor of a gastric tumor metastatic to the liver. The patient was referred to oncology for chemotherapy according to the CHOEP protocol. The patient subsequently died of his progressive disease.

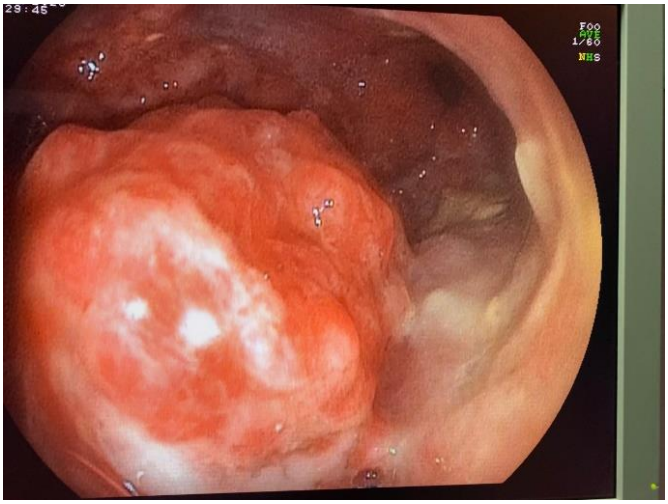

Figure 1: Ulcerating and infiltrating tumor process

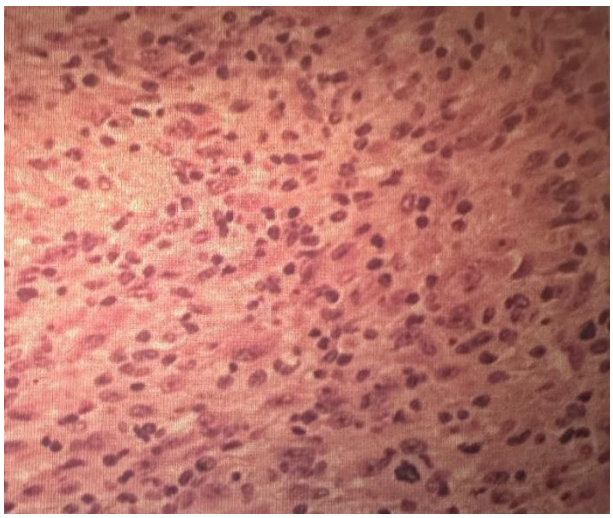

Figure 2: Histological image (× 600): Monomorphic to pleomorphic tumor cells with a spindle-like shape

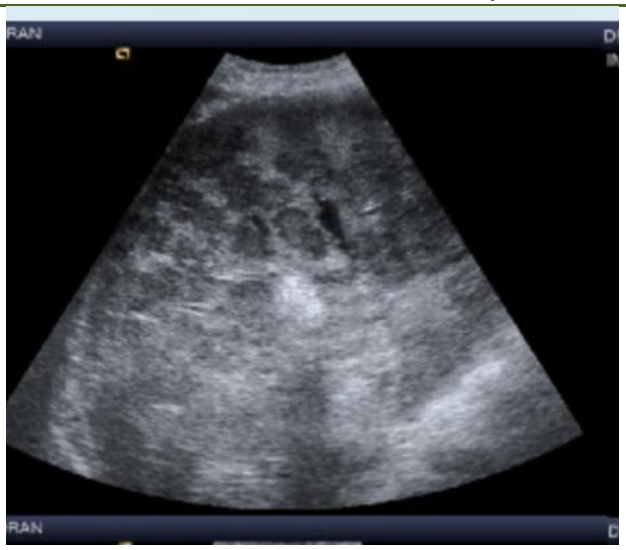

Figure 3: Multinodular liver with metastatic appearance

DISCUSSION

In 1970, Mathé *et al*., described a collection of malignant neoplastic cells with the histological characteristics of large macrophages and abundant eosinophilic cytoplasm. He was the first to introduce the term histiocytic sarcoma for tumors with histological similarities to macrophages, previously called reticulosarcoma and later histiocytic lymphoma. [8].

Histiocyte-derived malignant histiocyte disease was ambiguous in its definition. However, it was described by the World Health Organization (WHO) in 2001 as a malignant proliferative disorder with morphological and immunological properties similar to mature histiocytes, with the expression of one or more histiocytic markers other than dendritic cells, excluding acute monocytic leukemia", and the name was unified as HS [4]. In 2008, the WHO integrated all macrophage cell-induced malignancies into the HS category, and the frequency of HS was<1% of all hematopoietic lymphoid malignancies [5].

Histiocytic sarcoma of the stomach is extremely rare and only a small number of cases have been reported [6].

It represents less than 1% of hematologic malignancies, and a limited number of cases have been reported in the medical literature. Most patients are adults, and a male predilection is found in some studies [7].

Their diagnosis requires the use of immunohistochemistry and molecular biology techniques to distinguish them from lymphocytic proliferations.

Gastric HS occurred mainly in middle-aged or elderly people, with a mean age of 57.9 years, which is in line with our case report [8].

Patients usually presented with prolonged abdominal pain prior to diagnosis; other major symptoms were melena, dyspeptic symptoms [9, 10], as in the case of our patient.

Lesions may be ulcerative with erosive hemorrhage [11, 12].

HS can involve several sites including the colon, jejunum, oesophagus, pancreas and liver [13].

Hornick *et al.*, [3] have previously reported 14 cases of extra nodal HS and 13 cases of true histiocytic lymphoma. Of these, a total of 12 cases located in the gastrointestinal tract were reported in 11 patients, and in only one case was the lesion located in the stomach. Gastric HS can be located anywhere in the stomach: The cardia, fundus, antrum, lesser curvature, greater gastric curvature or bulb.

Regional lymph node involvement has been found in half the cases examined [9].

The imaging features of gastric HS are non-specific, and it is difficult to differentiate it from other gastric malignancies by radiological assessment alone [12].

Cases arising predominantly at extraganglionic sites may cause a diagnostic pathological dilemma, particularly in the absence of a high degree of atypia or pleomorphism of tumor cells masquerading as inflammatory pseudotumor [14].

Diagnosis of HS relies primarily on confirmation of its histiocytic lineage and exclusion of other poorly differentiated tumors such as lymphoma, carcinoma, sarcoma and melanoma [3].

HS is defined immunophenotypically by the expression of one or more histiocytic markers (CD68, CD163, lysosome), and the absence of Langerhans cells (CD1a, langerin), follicular dendritic cells (CD21, CD35) and myeloid cell markers (CD33, CD13, myeloperoxidase). CD45 and HLA-DR are generally positive. CD4 is often positive. The ki-67 index is variable [2, 6].

Treatment of gastric SH includes surgery (total or subtotal gastrectomy), radiotherapy, chemotherapy, monoclonal antibody therapy, immunomodulators, and new targeted agents. In previous studies, CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) chemotherapy has been reported to be efective for HS [15, 16].

Resistance to treatment is high, and the majority of patients have metastatic disease at diagnosis, with unsatisfactory treatment outcomes. Despite this context, the long-term prognosis is good if the lesion is small and localized.

HS prognosis was previously not promising [3, 10, 13]. A recent survey demonstrated a median overall survival of 6 months among 158 patients [17]. As in our patient's case (survival less than 3 months).

A literature search identified 4 cases. Three of these patients underwent surgery, while the remaining patient received adjuvant chemotherapy. It has been shown that adjuvant chemotherapy can be effective in prolonging survival [18].

CONCLUSION

Gastric histiocytic sarcoma is very rare. Distinguishing this type of tumor from other gastric tumors by simple radiological evaluation is difficult, underscoring the importance of immunohistochemical study. Because of its rarity, it is crucial to collect and evaluate additional cases in order to make progress in the therapies and prognoses associated with this diagnosis.

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