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Unusual case of repeated hematemesis: A Case Report and Review of the Literature

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ABSTRACT

Castleman's disease is a rare pathology of unknown etiology with different clinical manifestations, which creates both diagnostic and therapeutic dilemmas. It's abdominal localization is rarely described. There are no characteristic signs in imaging, and the diagnosis of certainty is histological. The management of Castleman's disease and its prognosis differ depending on whether it is localized or pluricentric. The localized form usually develops favorably after surgical excision. We present a particular case located in the mesentery, revealed by repeated hematemesis.

Introduction

Castleman's disease is a rare pathology, with uncertain cause. It is defined as a lymphoid angiofollicular hyperplasia classically found in the mediastinum. Its extra thoracic location, particularly mesenteric, is exceptional. The advent of new radiological modalities allowed early diagnosis and treatment of these entities. [1]

From a case of our service, we will discuss the different diagnoses to evoke in front of a mass of the epiploic space and we will recall the semiological elements of castelman disease and the modalities of its care and prognosis.

Case report

We report a case of a 20-year-old man, with no past medical history, presented to the hospital for abdominal pain with repeated hematemesis.

The clinical examination revealed an epigastric sensitive mass.

An intial abdominal CT scan done before consulting in our department showed an epigastric mass having an intimate contact with the stomach reminiscent of a GIST and fibroscopy revealed gastric varices.

An additional abdominal CT angiography with gastric filling was requested (Figure 1,2) and revealed the presence of a well limited mass developed at the expense of the lesser omentum, with regular contours, measuring 38 UH of spontaneous density, intensively and heterogeneously enhances in the arterial phase with homogenization at portal and late phase, measuring 74x47x65 mm. This mass get in touch with the small gastric curve and the left liver lobe with persistence of a greasy separation border. It is associated with lymphadenopathies of the small curvature, large gastric tuberosity and peri esophageal, probably producing compression of the left gastric vein, with as a result a development of collateral venous circulation peri-gastric and peri-esophageal protruding into the gastric lumen that we were able to better individualize with fibroscopy.

We have concluded to an hyper vascular mass of the small omentum with loco regional lymph nodes and collateral venous circulation bulging in the gastric lumen that may be related to a Castleman's disease.

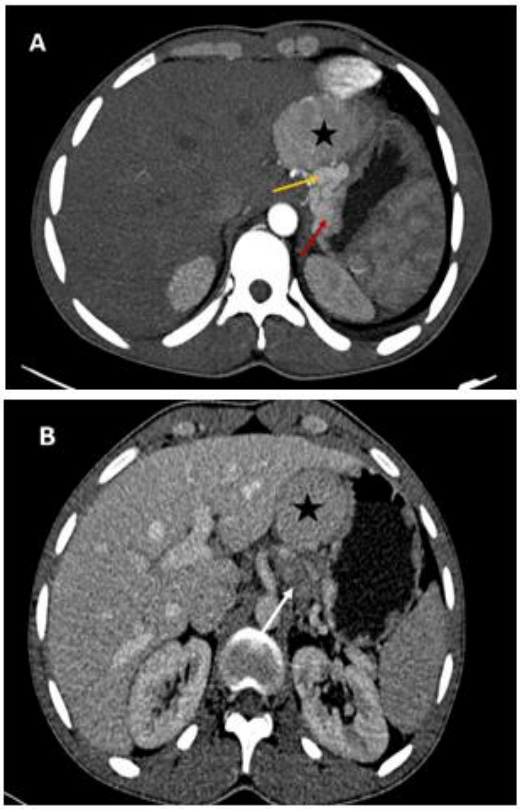

Figure 1. Axial abdominal CT angiography (in the arterial (A) and portal phase (B)) revealed the presence of a well limited mass (asterisk) developed at the expense of the lesser omentum, with regular contours, intensively and heterogeneously enhances in the arterial phase with homogenization at portal phase, producing a compression of the left gastric vein (yellow arrow) and gastric varices (red arrow). We also notice the presence of loco regional lymph nodes (white arrow)

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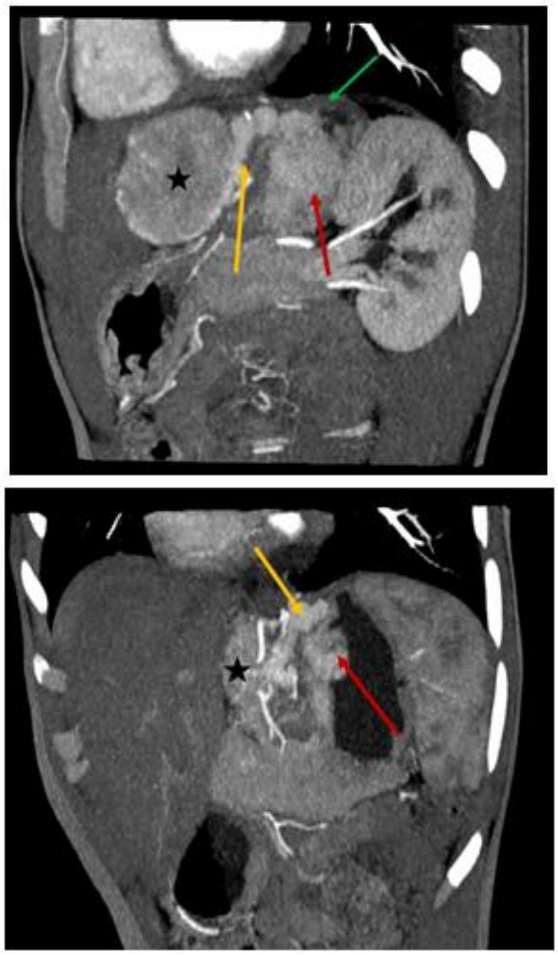

Figure 2. Coronal reformation of an abdominal computerized tomography showing the exact location of the lesion (asterisk) producing compression of the left gastric vein (yellow arrow) with varicose gastric varices (red arrow) protruding into the gastric lumen (green arrow)

The therapeutic attitude was a surgical resection of the mass (Figure 3) with anatomopathological study that confirms the diagnosis of Castelman's disease.

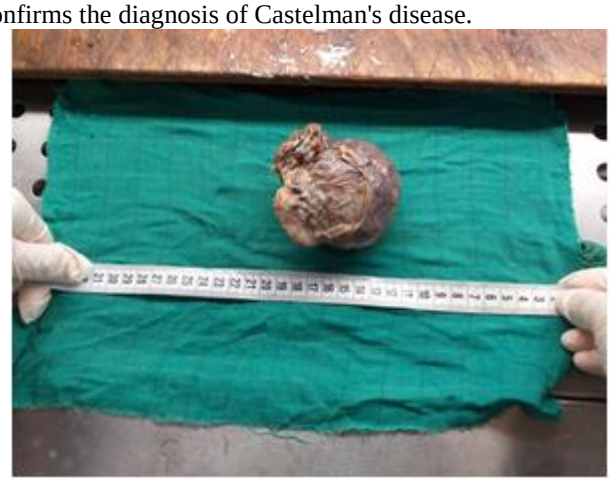

Figure 3. Gross appearance of the mass after surgical resection

Discussion

Castleman's disease, also called angiofollicular hyperplasia, was individualized by Castleman in 1956. This benign atypical lymphoproliferation is considered as a prelymphomatous state. [1, 3]

The origin of Castleman's tumors is unknown. Immune dysregulation, responsible for excessive proliferation of B lymphocytes in lymphoid organs has been suspected [1, 3].

It comes in two forms, localized and multifocal. The localized form, predominant in young adults, is most often asymptomatic. The main locations have been described in the mediastinum and in the chest wall, even if the mesentery, adrenal or pancreas may be also concerned. [1, 5, 6]

Clinically, general signs may exist such as asthenia, anorexia and fever. Biological abnormalities often founded are anemia, thrombocytopenia, elevated sedimentation rate of red blood cells, polyclonal hypergammaglobulinemia. However, there is no specific biological test. [6, 7]

The abdominal localization of CD is rarely described in the literature, and often leads to confusion with other pathologies, as Gastrointestinal stromal tumor (GIST) which was the first diagnosis we thought of, in view of the appearance of the lesion and its intimate contact with the stomach, but which we eliminated after performing a CT scan with gastrografin ingestion that showed no passage of contrast agent [4, 8].

In addition, the presence of associated lymph nodes makes this diagnosis unlikely since nodal involvement is absent in GIST.

Moreover, accessory spleen, wandering spleen, lymphoma, tuberculosis of the mesentery, solid pedicled liver mass, and some other hypervascular lesions represent the other differential diagnosis. [4,8]

In Imaging, It appears as a voluminous mass with irregular margins, no invasing of adjacent vessels and nerve structures, associated with regional lymphadenopathy. [4,7,8]

An intense and early enhancement in the arterial phase that persists in the late phase, as our case, is the most typical sign. This type of enhancement can be explained by a focal vascular proliferation associated with abnormal dilation of the capillary vessels. [4,7,8]

Several studies report that on CT and MRI, tumors larger than 5 cm show an heterogeneous enhancement with a non-vascularized central area in the early phase. [4] However, in our case, we found an homogenous enhancement during the arterial and late phase.

However, no characteristic signs in imaging allow making the diagnosis of Castleman's disease. The diagnosis of certainty is histological and, in 90% of cases, the examination shows a form of "vascular hyaline" characterized by abnormal lymphoid follicles and hypervascularisation of the interfollicular zone. [2,4]

In the case of single location, as in our case, the treatment of choice is surgical removal. In its multicentric form the treatment is based on corticosteroid therapy and immunosuppressive treatments. [3, 6]

Long-term follow-up is required regarding the risk of malignant transformation. [2]

Conclusion

Castleman's disease is a rare pathology that corresponds to a single or multi-centric ganglionic hypertrophy. The diagnosis can be difficult, especially in single location, because of the lack of clinical and radiological specificity. The intense arterial phase enhancement of the masse associated to the presence of loco regional lymph nodes should guide us towards this diagnosis, which is essentially based on the histological examination after surgical excision that will guide the therapeutic management.

The prognosis is different depending on whether it is a localized or multicentric form of the disease.

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