A Rare Form of Isolated Mesenteric Castleman’s Disease Presenting as an Abdominal Mass (Isolated Mesenteric Castleman’s Disease)

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Abstract
Castleman’s disease is a rare disorder characterized by proliferation of the lymphoid tissue. The most frequent location of the disease is the mediastinum. The location of the disease in the mesentery is rare and it is usually associated with the generalized form of the disease. We report a case of a 22-year-old woman with isolated mesenteric Castleman’s disease of the lymphoid variant, which presented as a palpable abdominal mass. The final diagnosis was reached after exploratory laparotomy and resection of the tumor. The described case is the first reported case of Castleman’s disease of the lymphoid subtype, located in the mesentery.

Key words
Castleman’s disease - mesentery - hyaline - vascular type - lymphoid subtype

Introduction
Castleman’s disease (giant lymph node hyperplasia or angiofollicular lymphoid hyperplasia) is a rare proliferative disease of unknown origin, first described by Castleman in 1954 (1). In the majority of the reported cases, the disorder is located in the mediastinum, whereas the mesenteric involvement is very unusual (2). The disorder is divided in two histological types: a) the hyaline-vascular type, which accounts for more than 90% of cases and is usually asymptomatic, and b) the plasma cell type, which is almost always associated with systemic manifestations. Although the former type is the most frequent among the patients suffering from the disease, its variant, called lymphoid subtype, has very rarely been reported in the mediastinum and other extrathoracic sites and never in the mesentery.

We present a case of isolated mesenteric Castleman’s disease of the lymphoid variant, which presented as a palpable abdominal mass. The described case is the first reported case of Castleman’s disease of the lymphoid subtype, located in the mesentery.

Case report
A 22-year-old woman presented at the emergency unit of our department complaining about acute abdominal pain located in the left lower quadrant and hypogastrum. The clinical examination revealed the presence of a palpable, mobile and slightly tender mass in the lower abdomen. Laboratory data showed slight hypochromic microcytic anemia (hemoglobin: 9.2 g/dl, hematocrit: 30.4 %, MCV: 59, MCH: 18.8), leucocytosis with polymorphonucleosis (14,100 leucocytes/mm3: 79.6% PMN cells) and elevated erythrocyte sedimentation rate (45mm).

X-rays of the chest and abdomen showed no pathologic findings. The ultrasonography of the abdomen revealed a 5.5cm x 6cm sharp defined, solid mass located between the intestinal loops. The CT of the abdomen confirmed the presence of a well circumscribed, encapsulated mass measuring 5.7 x 5 x 6cm, which seemed to arise from the mesentery and was adherent to the intestine. The mass enhanced moderately after the intravenous injection of contrast (Fig.1). In addition, CT showed multiple enlarged...
CT of the abdomen showing the presence of a well circumscribed, encapsulated mass arising from the mesentery and adherent to the intestinum. The mass was moderately enhanced after the intravenous injection of contrast medium.

Fig. 2 Small bowel follow-through contrast study showing a displacement of the intestinal loops by an extrinsic mass.

Regional mesenteric lymph nodes at the periphery of the tumor. In order to investigate further the precise origin of the tumor, a small bowel follow-through contrast study was made, which showed that the small bowel loops were displaced by an extrinsic mass, which had no connection to the intestinal wall. No significant obstruction of the lumen was evident (Fig. 2).

In view of the above described findings the patient was operated. The exploratory laparotomy revealed a mass located in the root of the mesentery, with no connection to the intestinal wall as well as regional enlarged lymph nodes (Fig 3). The biopsy of the mass revealed its lymphoid origin and excluded malignancy. The segment of the mesentery that contained the mass and the enlarged lymph nodes was resected along with a 30-cm segment of small intestine. Macroscopically, the mass measured 7 x 5 x 4 cm in size, was egg-shaped, hard and surrounded by a thick fibrous capsule. The cut surface was homogeneous, brown and firm (Fig 4).

The histopathological examination showed that the mass consisted of hyperplastic follicles scattered in a mass of lymphoid tissue. The follicles had a characteristic expansion of their mantle zone and small germinal centers. Inside the germinal centers, marked vascular proliferation and hyalinization were observed (Fig 5).
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stoma was characterized by moderate hyperplasia of postcapillary venules and an admixture of plasma cells and eosinophils. The immunohistochemical examination of the plasma cells was positive for κ and λ chains, proving the polyclonality of immunoglobulin. The histopathological examination of the five adjacent enlarged lymph nodes, which were found in the specimen showed similar microscopic changes.

The histopathological diagnosis was angiofollicular lymph node hyperplasia (Castleman’s disease) of a variant of the hyaline vascular type, described as lymphoid subtype.

After definite establishment of the diagnosis, the patient underwent CT of the thorax in order to exclude the presence of enlarged lymph nodes in the mediastinum. As this was normal, we defined our case as an isolated Castleman’s disease of the mesentery.

The postoperative course of the patient was uneventful and she was discharged on the 10th postoperative day. Ten months after the operation the patient is free of signs of recurrence, while the anemia has totally been resolved.

Discussion

Castleman’s disease is a rare pathologic process, with undefined precise incidence and unknown etiology, characterized by lymph node hyperplasia. Although it is most commonly seen in adults, it can also occur during childhood (3). In the vast majority of the cases, the disease is located in the mediastinum. However, extrathoracic sites have also been described, including the mesentery, axilla, neck and retroperitoneum.

The disease is divided histologically in two major types: the hyaline vascular type and the plasma cell type (4). The plasma cell type is seen in less than 10% of the patients with Castleman’s disease and is characterized by a diffuse interfollicular plasma cell proliferation with minimal vascular component. The hyaline-vascular type, which is the most common type (90%) is characterized by large follicles, showing marked capillary proliferation and hyalinization in a mass of lymphoid tissue. At the periphery of these follicles there is a concentric layer of lymphocytes that comprise the mantle zone. Rarely does the mantle zone show prominent hyperplasia, while the interfollicular area presents minimal to moderate vascular proliferation and inconspicuous germinal centers. These subtype of the hyaline-vascular type is called lymphoid subtype, and has been described in the mediastinum as well as other sites, such as the retroperitoneal space and the inguinal region, but not in the mesentery so far (4,5). Our case belongs to this unusual variant and to our knowledge this is the first case of mesenteric Castleman’s disease of the lymphoid subtype described in the literature.

Patients with the hyaline-vascular type of Castleman’s disease are usually asymptomatic and the disease is discovered incidentally, in contrast to the patients with the plasma cell type, which are often symptomatic at the time of diagnosis (2). The clinical manifestations of the disease are not specific and may include fever, anemia, fatigue, weight loss. Laboratory examinations may reveal anemia, elevated erythrocyte sedimentation rate or hypergammaglobulinemia. In our case, the patient presented with a palpable abdominal mass without any other clinical sign of a symptomatic disease. However, the laboratory data showed mild anemia, leucocytosis with polymorpho-nucleosis and increased sedimentation rate.

Castleman’s disease can be divided into two further forms: the most common localized-solitary form and the less usual multicentric form. Approximately 80% of the cases of the solitary form belong to the hyaline-vascular type and the remaining 20% to the plasma cell type. The usual location of the solitary mass is the mediastinum (70%), whereas the mesentery is very rarely involved. The widespread form of the disease is characterized by disseminated lymphadenopathy, is almost always associated with systemic symptoms and is dominated by the plasma cell type (6).

Irrespective from the histopathological type of the disease, the localized form always shows a benign behavior. Complete surgical excision of the lesion - after laparotomy or even laparoscopic approach - provides cure of the disease, since there are no reported cases of recurrence after total excision of a solitary mass (7,8).

On the contrary, the multicentric form of the disease follows a more aggressive course and is associated with poor prognosis. The therapeutic approach of the widespread form remains controversial, as many treatment regimens have been proposed, including surgery, chemotherapy, corticotherapy or combination of these (9,10). An accurate staging of the disease, including a very thorough clinical examination for the detection of suspicious lymph nodes in the axilla, neck or groin and CT of the thorax should be performed in any case in order to exclude the presence of extra-mesenteric disease.

The development of mesenteric Castleman’s disease has to be differentiated from other benign or malignant mesenchymatous lesions of the mesentery, such as stromal tumors, leiomyomas, leiomyosarcomas, fibromas etc. Castleman’s disease has to be differentiated also from other inflammatory lesions that may result in mesenteric lymphadenopathy such as terminal ileitis, tuberculosis etc. Finally, the differential diagnosis included also tumors arising from the intestine or other adjacent organs.

The preoperative diagnosis of the disease is still very difficult, even with the newest medical imaging techniques (11). Malara et al (12) described in detail the sonographic, computed tomographic and angiographic features of the mesenteric Castleman’s disease. According to them, the mesenteric Castleman’s disease presents as a homogeneous, hypoechoic mass on ultrasound, while the color Doppler sonography reveals a vascular tumor with a low resistance flow pattern. The CT findings confirm the homogeneity and hypervascularity of the mass and angiography demonstrates a hypervascular mass with hypertrophied feeding vessels and a homogeneous capillary blush, at least in the hyaline-
vascular type (13). Unfortunately, these radiological features are not specific for Castleman’s disease, as they can be observed in every lymphomatous tumor, benign or malignant or other mesenteric masses (14).

In most of the cases, the diagnosis of mesenteric Castleman’s disease is confirmed only after the resection and histopathological examination of the specimen.

In conclusion, Castleman’s disease has to be included in the differential diagnosis of every mass located in the mesentery, especially when the lesion shows the imaging features mentioned above and the patient presents with the previously described abnormal laboratory findings.

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References