Case Report

Localized Castleman’s Disease Presenting as a Vascular Right Iliac Fossa Mass

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Introduction

Castleman's disease is also known as angiofollicular lymphoid hyperplasia, giant lymph node hyperplasia, lymphoid hamartoma, and follicular lymphoreticuloma. It is a rare disease, characterized by hypertrophy of the lymph nodes and histologically by angiofollicular lymph node hyperplasia. Benjamin Castleman and his colleagues first mentioned it in 1956, in a group of patients with localized mediastinal lymph node hyperplasia. Since then, more than 400 cases have been reported in the international literature.

Case report

A 34-year-old woman presented with symptoms of upper respiratory tract infection and acute gastroenteritis. She had no history of altered bowel habits prior to this admission but admitted to a recurrent right iliac fossa pain and weight loss over a period of 6 months. There was no history of note. On examination, she was afebrile with a pulse rate of 80/minute and blood pressure of 110/70 mmHg. She was pale and had clubbed fingers. Cervical lymph nodes were not palpable. Abdominal palpation revealed a firm and slightly tender mass over the right iliac fossa. This mass measured about 5 cm and moved with respiration. The liver and spleen were palpable 5 cm below the costal margin but were non-tender. Otherwise, the systemic examination was unremarkable.

Her haemoglobin was 5.6 g/dL, leukocyte count was 10,200/mm³, and platelet count was elevated at 625,000/mm³. The erythrocyte sedimentation rate was elevated to 145 mm/hr. Full blood analysis revealed microcytic hypochromic anaemia, but haemoglobin electrophoresis did not show any haemoglobinopathies. Liver function tests showed hypoalbuminaemia (21 g/L) but normal levels of bilirubin and trans-
aminases. Her renal function and coagulation profile were within normal limits.

Chest radiography showed normal cardiac size with normal lung markings. Ultrasound showed a mass in the right iliac fossa, with a moderately enlarged liver and spleen. Duplex ultrasound showed that the mass was hypervascular. Computerized tomography (CT) scan confirmed a right iliac fossa mass measuring 5 x 5 cm with internal calcification (Figure 1). The mass was not enhanced by contrast agent and was not related to the bowel. There was streakiness of the mesentery with inter-loop fluid collection, indicating an inflammatory reaction. The para-aortic nodes were also enlarged. The liver and spleen did not show any focal lesions.

Our provisional diagnosis was a soft tissue sarcoma; a visceral angiogram with preoperative tumour embolization was planned. Mesenteric arteriogram showed that the arterial supply was mainly from the superior mesenteric artery and had prominent venous drainage (Figure 2). Blood was also supplied by the lumbar arteries. Tumour embolization was not performed for fear of causing bowel ischaemia.

We proceeded to a laparotomy. Intraoperatively, we noted incidental malrotation of the bowel with a short mesentery and mobile caecum and ascending and descending colons. The hepatic and splenic flexural attachments were absent. The right iliac fossa mass appeared to be encapsulated and was enclosed within the leaves of the jejunal mesentery, about 20 cm from the duodenojejunal flexure. The mass was lobulated and comprised of multiple enlarged and matted lymph nodes, the largest measuring 4 cm (Figure 3). There were multiple tiny lymph node-like nodules studded in the mesentery. The mass, together with the enclosing mesentery, was resected along with 15 cm of jejunum, and an end-to-end anastomosis of the bowel was created. Postoperative recovery was uneventful. Histopathology showed localized plasma cell Castleman's disease. The patient was advised to return for lifelong follow-up with periodic ultrasound examination of the abdomen for possible tumour recurrence.

**Discussion**

Castleman's disease is classified into two types: localized, in
which the disease is situated at one site only, and multicentric or systemic, in which several sites are involved.2-3 Histologically, there are two main types of Castleman's disease, hyaline vascular and plasma cell.2-5 Hyaline vascular Castleman's disease is much more common (90%) than plasma cell disease.2 Multiple germinal centres, surrounded by circumferentially arranged layers (onion skin pattern) of small lymphocytes, with a prominent vascular stroma and occasional plasma cells, characterize hyaline vascular disease. Sheets of dense plasma cells and a less vascular stroma surrounding the germinal centres characterize plasma cell disease.3

Patients with hyaline vascular Castleman's disease are usually asymptomatic and the lesion tends to be localized.2 Patients are usually young adults, less than 35 years old.3 The lesion is often discovered incidentally, for example, on routine pre-employment chest radiography. Sometimes, when the lesion is large and compresses adjacent structures, the patient presents with abdominal or thoracic pain. The more common sites are the mediastinum, abdomen, and pelvis.3 Most patients with plasma cell Castleman's disease present with constitutional signs and symptoms of fever, malaise, anaemia, thrombocytosis, and increased erythrocyte sedimentation rate caused by B-cell hyper-reactivity.2 Only 10% of these patients have localized disease.3

Multicentric Castleman's disease (MCD) is a severe form of the disease. Patients are usually older, in their fifth or sixth decades of life, with multiple lymph-node involvement and associated hepatosplenomegaly.3,6 Some patients have POEMS syndrome, which consists of peripheral neuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin lesions.7,8 Histologically, MCD may present as a combination of both hyaline vascular and plasma cell morphology.3 Medical opinions differ as to whether MCD is a distinct type or a disseminated variant of a localized disease.4-6

Castleman's disease is associated with paraneoplastic pemphigus (an autoimmune bullous mucocutaneous disorder), benign lesions such as pseudotumour, and malignancies such as lymphoma and Kaposi's sarcoma.9-14 The aetiology of Castleman's disease is not well understood. Viral infection by human herpes virus 8 or Epstein-Barr virus, and dysregulation of interleukin-6 (IL-6) secretion have been implicated as possible causal factors of this disease. No genetic or toxic factor has thus far been identified.15-17

Differential diagnosis of this rare disease includes reactive lymph node hyperplasia, malignant lymphoma, HIV infection, and autoimmune diseases such as rheumatoid disease or Sjögren's syndrome. It is therefore important to obtain proper clinical, histological, and immunohistochemical analysis. The diagnosis of Castleman's disease is based on histological examination of the specimen in combination with immunohistochemical labelling.18,19

The treatment of choice for localized Castleman's disease is complete surgical resection.2,3,20 There is a low recurrence rate with subtotal resection. The prognosis is very good with no relapse in almost all cases if the lesion is completely removed.3 Radiotherapy is an alternative option if the patient is a poor surgical candidate, but the result varies.3,20 Long-term follow-up is required to detect the possible development of malignancy, such as non-Hodgkin's lymphoma.14 The treatment of MCD involves surgery, radiotherapy, steroids, and combination chemotherapy, such as cyclophosphamide, vincristine, and doxorubicin.3,4,20,21 Other treatment modalities include interferon-α, retinoic acid, and anti-IL-6 antibodies.3,22,23 However, the prognosis is currently still poor for this type of disease, with a reported median survival of only 26 months.4

The disease in our patient was confined to one site only, in a small area of the jejunal mesentery. Histopathological and immunohistochemical staining showed plasma cell Castleman's disease. The prognosis for this patient with localized Castleman's disease is favourable as the lesion was completely resected and the patient is currently asymptomatic with no disease recurrence more than 1 year after the operation.

Abdominal CT scan showed a mass that was not enhanced by contrast agent, indicating that it might not be a vascular lesion. This contradicted the initial duplex ultrasound finding of a hypervascular mass, which was interesting because the tumour had arisen from within the two leaves of the mesentery. Retrospectively, the duplex ultrasound scan finding of hyper-vascularity was believed to be due to the mesenteric vessels coursing the surface of the tumour.

This rare and interesting tumour caused a diagnostic dilemma due to the discrepancy between findings on duplex ultrasound and CT scan. Mesenteric angiogram revealed that the feeder artery arose from the superior mesenteric artery, which further confused the picture. This tumour was uniquely wrapped by the leaves of the mesentery, which accounted for the investigative findings. This had not been previously described in the literature.

In conclusion, Castleman's disease is a rare non-neoplastic lymphoproliferative disorder that relies on proper clinical and histological evaluation for its diagnosis. Other more common diseases such as lymphoma and autoimmune diseases must be ruled out. Localized Castleman's disease has a very good prognosis with a complete cure by total surgical resection.
MCD, on the other hand, still has a poor outcome despite the availability of multiple modalities of therapy.

References