Mesenteric Castleman’s Disease: Case Report and Literature Review

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A 41-year-old man was discovered to have a pelvic mass during investigation for hypertension 2 years ago. Otherwise, he was asymptomatic, and abdominal magnetic resonance imaging revealed a highly vascular solid mass superior to the urinary bladder. A 131I-meta-iodobenzylguanidine whole body scan was reported as normal, which ruled out the possibility of extra-adrenal pheochromocytoma. No definitive preoperative diagnosis could be established. Surgical resection of the tumour revealed mesenteric Castleman’s disease, hyaline vascular type. Such a diagnosis should be considered for any abdominal vascular mass. [Asian J Surg 2010;33(3):150–3]

Key Words: Castleman’s disease, hyaline vascular type, mesenteric tumour

Introduction

Castleman’s disease (CD) is a giant lymph node hyperplasia or angiofollicular lymphoid hyperplasia of unknown origin,1 which has attracted attention because of its association with human immunodeficiency virus (HIV) and human herpes virus 8 (HHV8).2 The disease was first described by Castleman in 1954,3 and in the majority of the reported cases, the disorder is located in the mediastinum, whereas mesenteric involvement is very unusual.1 The neck, retroperitoneum, pancreas, pelvic cavity and axillary and inguinal lymph nodes are also reported sites of involvement.

It is associated with a number of malignancies, including Kaposi’s sarcoma, non-Hodgkin’s lymphoma, Hodgkin’s lymphoma and POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M-protein, skin manifestation). It is encountered in patients aged between 30 and 40 years and rarely occurs in children.2 CD should be included in the differential diagnosis of every mass located in the mesentery. In the present case, the final diagnosis was established after explorative laparotomy and resection of the tumour. To the best of our knowledge, only 23 cases of mesenteric Castleman’s disease have been reported in the English-language literature.3

Case report

A 41-year-old man with hypertension presented to the outpatient clinic with a history of lower abdominal mass (6 cm × 7 cm) (Figure 1) that was discovered incidentally two years earlier upon investigations for hypertension. Colonoscopy was performed twice over 2 years on both occasions. There was a pressure effect on the rectum without any mucosal abnormality, and he was advised to have the abdominal mass resected, but he refused. The patient was asymptomatic with a normal appetite, no vomiting or abdominal pain, with regular bowel habits and no weight changes. He had no urinary, respiratory or cardiovascular symptoms, no constitutional symptoms, had not
undergone previous surgery. He was not a smoker and his only medication was antihypertensive drugs (amlodipine and carvedilol).

Upon physical examination, his blood pressure was 150/90 mmHg; he had suprapubic fullness of about 10 cm × 10 cm with a smooth surface, which was nonpulsatile, firm in consistency and relatively immobile; otherwise his examination was unremarkable. Abdominal magnetic resonance imaging revealed a large heterogeneous solid mass superior to the urinary bladder, which measured about 7 cm × 8 cm × 9 cm in maximum dimension (Figures 2A and 2B). The mass showed avid enhancement with intravenous contrast agent and multiple necrotic foci. The mass appeared to be vascular, and took multiple branches from the right common iliac artery. We excluded preoperative biopsy because early workup revealed borderline high catecholamine levels that raised the possibility of extra-adrenal pheochromocytoma, in addition to the high vascularity of the mass. An endocrinologist and nephrologists were consulted preoperatively.

We requested a 131I-meta-iodobenzylguanidine whole body scan to rule out extra-adrenal pheochromocytoma. Metanephrine, vinyl mandelic acid and catecholamines were within normal limits. Preoperative and intraoperative precautions for possible diagnosis of extra-adrenal pheochromocytoma were taken. Positron emission tomography was not considered because the course of the disease over > 2 years was benign.

The patient underwent laparotomy. The mass was found to be 10 cm × 10 cm and highly vascular. It occupied the whole mesorectum, encroached on the sigmoid mesentery and stretched the rectum in front of it. Separation of the mass from the rectum was not possible, therefore, traditional anterior resection was carried out followed by reestablishment of bowel continuity by a primary stapled colorectal anastomosis (Figure 3). The patient had a smooth postoperative course and was discharged home on postoperative day 5 in good condition.

The pathology report came back as rectosigmoid mesenteric Castleman’s disease, hyaline vascular type. The bowel mucosa, submucosa and muscularis propria were intact. Microscopy showed the attached mass to be enlarged lymph nodes, showing diffuse follicles with extensive vascular proliferation and hyalinisation of the germinal centres and perifollicular areas (Figure 4A). Plasma cells were seen focally (Figure 4B), but were all polyclonal, as confirmed by immunohistochemical staining. The lesion was completely excised and no other lymph node involvement was seen.
HIV and HHV8 tests were negative. The patient was readmitted 2 days after discharge with partial adhesive intestinal obstruction that settled with conservative treatment. The patient was followed up at 2 months and 6 months without any problems.

Discussion

CD is a benign condition of unknown aetiology. It is a lymphoproliferative disorder that has attracted attention because of its association with HIV and HHV8, and several malignancies including Kaposi’s sarcoma, non-Hodgkin’s lymphoma, Hodgkin’s lymphoma and POEMS syndrome. The histological findings can be divided into two types: hyaline vascular and plasma cell types. The hyaline vascular type accounts for > 90% of cases and is characterised by small hyaline follicles and intrafollicular capillary proliferation. The plasma cell type is less common and has a spreading arrangement of plasma cells in the involved lymph nodes. Clinically, the hyaline vascular type is usually asymptomatic, whereas patients with the plasma cell type might present with systemic manifestations including fever, anaemia, elevated C-reactive protein and erythrocyte sedimentation rate, and hypergammaglobulinaemia.

A correct preoperative diagnosis of mesenteric CD was not made; localised CD requires surgical removal of the tumour for both diagnosis and therapy. Recurrence of the localised disease is rare after complete surgical resection, but it has been reported in patients with incomplete surgical removal. Limited success with radiotherapy (30–45 Gy) and chemotherapy has been reported. It is generally accepted that the disease is benign and surgery is curative. Complete surgical excision of the lesion after laparotomy or even laparoscopic surgery is curative, and there have been no reported cases of recurrence after total excision of a solitary mass.

CD can be divided into two further forms: the more common localised solitary form and the less frequent 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 widespread form of the disease is characterised by disseminated lymphadenopathy and is almost always associated with systemic symptoms. The therapeutic approach for the widespread form remains controversial because many treatment regimens have been proposed including surgery, chemotherapy, corticosteroids, or combinations of these. In most cases, diagnosis of mesenteric CD is confirmed only after resection and histopathological examination of the specimen.
References