A case of Castleman’s disease mimicking a multicystic ovarian tumour

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We report on a case of solitary Castleman’s disease that had an unusual presentation. A 29-year-old Filipino woman who had a history of intermittent right lower abdominal pain for several years was admitted to the Princess Margaret Hospital because of a sudden exacerbation of the abdominal pain. Ultrasonography had previously detected a multilocular right ovarian cyst of approximately 6.5 cm in diameter. Intra-operative findings, however, revealed a retroperitoneal presacral cystic tumour of approximately 7 cm in diameter, which was unrelated to the ovaries. The tumour was removed and found to be well defined and measured 7 x 5 x 4 cm. The cut surface revealed homogenous light-brown tissue at the periphery. The central part showed cystic spaces of 1- to 3-cm diameter. Histological examination of the tumour led to the diagnosis of hyaline vascular–type Castleman’s disease. Cystic spaces were visible within the infarcted tissue; extensive cystic changes in Castleman’s disease are unusual and may have caused the diagnostic difficulty.

Key words: Diagnosis, differential; Giant lymph node hyperplasia; Retroperitoneal neoplasms

Introduction

Castleman’s disease, also known as angiofollicular lymphoid hyperplasia, was originally described as a solitary lesion in the mediastinum. Although the tumour-like mass is located in the mediastinum in two thirds of patients with this disease, extrathoracic sites can also be affected. Such sites include regions that contain lymph nodes—for example, the axillae, neck, groin, mesentery, and retroperitoneal spaces. Tissues which do not contain lymph nodes are rarely also involved. We report on a case of Castleman’s disease that presented as a right pelvic multilocular mass resembling a multilocular ovarian cyst.

Case report

A 29-year-old Filipino woman was admitted to the Princess Margaret Hospital in May 1994 because of a sudden increase in the severity of right lower abdominal pain. The patient had been healthy previously, but had a history of intermittent right-lower abdominal pain for several years. She had previously undergone an ultrasound examination of the pelvis, which revealed a right ovarian cyst of approximately 6.5 cm in diameter.

Physical examination on admission to hospital showed that the patient’s general condition was satisfactory. Abdominal examination did not reveal any tenderness or mass. Per rectal examination, however, suggested the presence of a right pelvic mass of approximately 5 cm in diameter. Ultrasonography revealed a multilocular cyst of 5.1 x 5.2 x 6.3 cm that contained hyperdense areas, in the right pelvic cavity.

Emergency surgery was performed and revealed a well-defined retroperitoneal presacral cystic tumour of approximately 8 cm in diameter. The results of an intra-operative frozen section biopsy of this tumour showed that the patient had Castleman’s disease. The ovaries and fallopian tubes were unremarkable. The tumour was completely excised and there were no postoperative complications.

Gross examination of the tumour showed an outer fibrous capsule that measured 7 x 5 x 4 cm. The cut surface revealed homogenous light-brown tissue mainly at the periphery. The central part of the cut surface
contained cystic spaces of 1- to 3-cm diameter (Fig 1). Histological examination confirmed the diagnosis of Castleman’s disease of the hyaline vascular type (Fig 2). The tissue section contained follicle-like lymphoid aggregates, which were surrounded by concentric layers of small lymphocytes. Instead of containing activated germinal centres, the lymphoid aggregates were penetrated by capillaries that contained varying degrees of hyalinization. The tissue between the abnormal follicular structures was also rich in small blood vessels that had undergone hyalinization. Cystic spaces were present within the infarcted tissue. The patient has not had any recurrence of Castleman’s disease during 3 years of follow-up.

**Discussion**

Castleman’s disease is a rare condition that was first described by Castleman et al.[1] The disease is also known as lymph nodal hamartoma, angiofollicular mediastinal lymph node hyperplasia, angiomatos lymphoid hamartoma, follicular lymphophoreticuloma, and benign giant lymphoma. Another alternative name is angiofollicular lymphoid hyperplasia, a term which originally described a solitary lesion in the mediastinum—the most frequent site of involvement.

The classic symptom of Castleman’s disease is a usually benign solitary lesion, although multicentric disease can also occur.[3-6] A significant proportion of patients with the multicentric form of disease have a poor prognosis, often a rapidly progressive course of disease, and a fatal outcome. In both the solitary and multicentric forms of the disease, there are two principal histopathological types: the hyaline vascular variety, which occurs in the majority of patients, and the much rarer plasma cell type.

Cystic change in Castleman’s disease is extremely rare.[7,8] The case reported here posed a diagnostic curiosity. The retroperitoneal, presacral location of a cyst is uncommon, and predominant cystic changes further led to the misdiagnosis of multicystic ovarian tumour. The close association between cystic and infarcted tissue implies an aetiological link. Such marked cystic and retroperitoneal changes in a case of Castleman’s disease have not been reported in the English literature. Although rare, Castleman’s disease could be included in the differential diagnosis in any young woman who presents with a multilocular pelvic mass.

**References**