Solitary Intra-Abdominal Castleman's Disease, Hyaline Vascular Type: Case Report

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Abstract

Objective

To report a case of solitary intra-abdominal Castleman disease and highlight the importance of this entity to clinicians in the management of these patients.

Case presentation and intervention

A 20 year old gentleman who presented with a recurrent intra-abdominal retroperitoneal mass. Previous biopsies obtained from a laparotomy showed Castleman disease- vascular hyaline type. Patient did not respond to chemotherapy and the mass was gradually increasing in size. Surgical intervention was the only option and the patient underwent complete resection of the retroperitoneal mass. Final histopathological examination confirmed the initial diagnosis of Castleman disease.

Conclusion

Castleman's disease is a fairly rare benign tumor of lymphoid origin. It should be included in the list of differential diagnosis of retroperitoneal masses. Unicentric Castleman disease should be treated surgically when feasible and carries better prognosis compared to multicentric disease.

Key words:

Retroperitoneal mass- Castleman's disease- lymph node- unicentric-multicentric.

Introduction

Castleman's disease rarely presents as an isolated retroperitoneal mass radiographically indistinguishable from retroperitoneal malignancies, it was first described by Dr.Benjamine Castleman in Boston in 1956 (1). It is also known as giant lymph node hyperplasia and angiofollicular lymph node hyperplasia. Although it is not officially a cancer, the "multicentric" form of this disease acts like lymphoma. The expected location is mediastinum and rarely peritoneum (2). We report a case of solitary intra-abdominal Castleman's disease.

Case report

A 20 year old gentleman presented initially outside Kuwait in December 2003 with intra-abdominal mass. The patient had a laparotomy and the mass was biopsied according to the history obtained from the patient. The mass was in the retroperitoneal area inferior to left kidney. The initial diagnosis was made as T cell non Hodgkin's lymphoma which was not confirmed by immunohistochemistry. He received 6 cycles CHOP three weekly from Feb'04 to May'04. In June'04, it was decided that the mass was persistent despite of chemotherapy and the patient was re-explored and an attempt of resection of the mass was performed.

Histopathological examination of the specimen established the final diagnosis of Castleman's disease, Hyaline vascular type. The patient was under regular follow up by his physician abroad for few years and annual postoperative CT scans of the abdomen showed residual mass measuring 4cm, this mass was gradually increasing in the size. Most recent scan in July'08 showed a 6.4x6.8x7cm mass in the left lumbar/iliac fossa. The mass was irregular at the lateral margin of the left psoas muscle displacing the bowel loops anteriorly. He presentated to our center at the Kuwait cancer control center in July30,08, he was completely asymptomatic and this was the only palpable abdominal mass clinically which was about 10cm at left lumbar region. Intravenous urogram was performed and

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showed displacement of the left ureter. In view of recurrence/ progression of this local mass and diagnosis of Unicentric Castleman's disease, the oncology board decided that complete surgical resection of this mass was the best option.

Laparotomy and resection of the retroperitoneal mass was done in Aug'08. A left ureteric double J stent was placed preoperatively and a midline incision was used. Adhesiolysis was performed in order to mobilize the left colon up to the splenic flexure. 8x6cm mass was felt below the lower pole of the left kidney extending down to the pelvis on the lateral aspect of the psoas muscle deeply impacted into paraspinal space. The mass was infiltrating the psoas muscle posteriorly, margins were grossly free as the mass was encapsulated except for the posterior aspect. Excision of the mass from the lateral aspect was

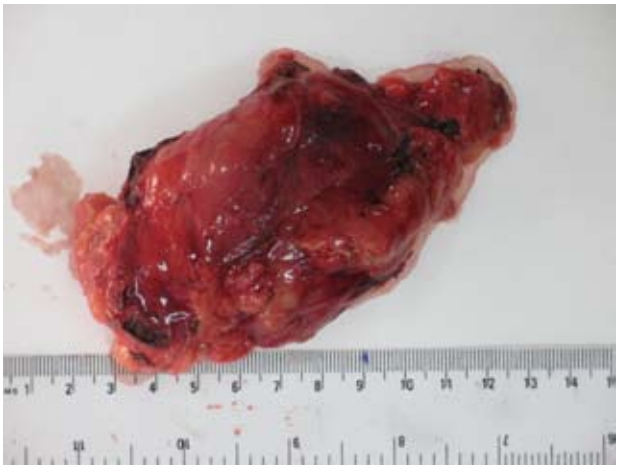

Fig. 1 : Magnification 40x

done using the ligasure, minimal bleeding was encountered and controlled. Patient tolerated the procedure well. Histopathology revealed a left retroperitoneal mass measuring 11x7x3cm and weighing 88.5gms with smooth outer surface (Figure 1).

Microscopically, sections revealed a lymph node with effacement of nodal architecture by blood bands of hyalinized tissue (Figure 2). Follicular hyperplasia with atrophic germinal centers rimed by concentric onion skin layers of small lymphocytes were seen (Figure3). Diagnosis was made as Castleman's disease. The patient's case was discussed in an oncology meeting and the consensus was to follow the patient up as the mass was grossly completely excised.

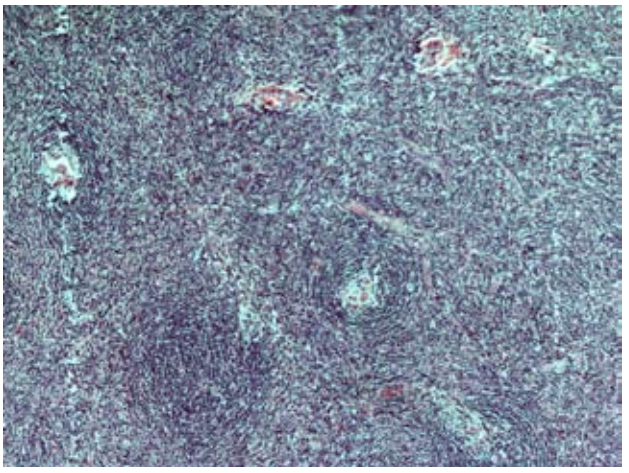

Fig. 2 : Magnification 40x

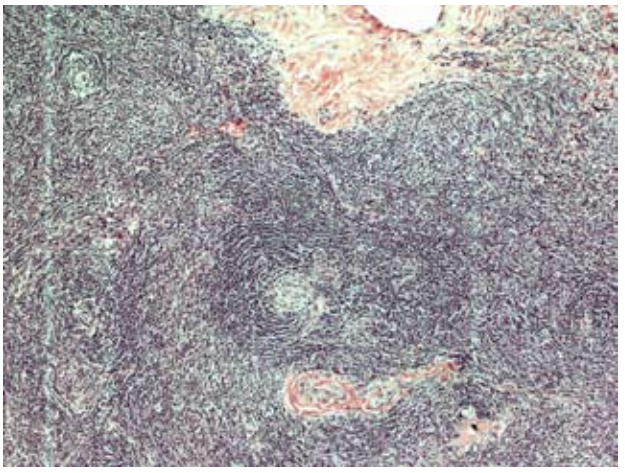

Fig. 3 : Magnification 40x

Discussion

Castleman's disease is a rare disorder characterized by non-cancerous tumors that may develop in the lymphoid tissue at a single site or throughout the body [3]. The two main forms of Castleman's disease are localized and multicentric forms. The localized or unicentric castleman disease only affects a single lymph node and is not widespread. These abnormally large lymph nodes may press on other tissues inside the chest or abdomen as it has been reported to present as a solitary pulmonary nodule which was treated by resection [4]. Other case reports published on solitary Castleman's disease was affecting the meninges (intracranial solitary Castleman's disease) which was also treated by surgical excision [5]. People with localized Castleman's disease are usually cured when the lymph node is surgically removed. It represents a morphologically distinct form of a rare atypical lymphoproliferative disorder rather than a neoplasm. Most cases occur in the mediastinum,

but cervical, axillary, inguinal, shoulder and vulvar regions are also possible locations [6].

Seven percent of cases are in the retroperitoneal space, and 2% have a pararenal location [7].

The multicentric Castleman disease affects more than one group of lymph nodes and may also affect other organs containing lymphoid tissue [8] .

The pathogenesis of Castleman's disease is unclear. Chronic low grade inflammation and immunodeficiency state, and autoimmunity have been proposed as likely mechanisms, viral infection such as HIV and HHV-8 may be an inciting agents [9]. So far, studies have shown that HHV8 is found in most people with multicentric castleman disease who are HIV positive. But only one case of unicentric castleman disease has been associated with HHV8 [10].

Another potential contributor to the disease may be a type of protein produced by immune cells called interleukin-6 (IL6). It is possible that HHV8, or some other unidentifiable factor, may stimulate overproduction of IL6, leading to an overgrowth of lymphatic cells [11]. The microscopic subtypes of Castleman disease include a hyaline vascular type or a plasma cell type based on how the lymph node tissue appears under the microscope, 90% of the solitary tumors are of the hyaline-vascular type [6]. Less often, a combination of both types may occur. The hyaline vascular type is more common and tends to be localized while the plasma cell type tends to be multicentric.

Treatment depends largely on the type of Castleman's disease present. Surgery is often used to obtain a tissue biopsy to diagnose Castleman disease. Surgery is also very effective treatment for localized disease. The type of surgery depends on where the disease is located. Radiation therapy has sometimes been used instead of surgery to effectively treat localized disease.

The solitary type is usually benign, responds well to surgical excision and is curative. Widespread Castleman's disease follows aggressive, often fatal clinical course, and the prognosis is poor, such patients need multimodality therapy, such as steroids, immunosuppressions, antineoplastic agents and surgery.

Treating multicentric Castleman disease is generally more difficult because the disease is very rare, has varied non specific symptoms and signs and spontaneously goes into remission at times, doctors have found it difficult to identify the best treatment. In addition, there are no clinical trials that offer definitive evidence in favor of any therapy. Most therapies are palliative and the lymphoma that develops from Castleman disease usually grows fast and is hard to treat.

Conclusion

Castleman's disease is a benign tumor of lymphoid origin. It should be considered in the differential diagnosis of any retroperitoneal mass. Surgery is the recommended treatment for patients with localized Castleman disease. Removing the abnormal lymph nodes appears to cure the disease. Dealing with multicentric disease can be challenging and those patients have far worse prognosis than the localized ones.

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