

Castleman's Disease: Two Case Reports, with Review of Literature

*Abdel Hadi Breizat,*¹ Amal S. Al-Tahieneh¹*

Abstract

Castleman's Disease (CD) is a rare disorder characterized by a benign atypical proliferation of lymphatic tissue, and can occur anywhere along the lymphatic channels in either localized or multicentric(systemic) forms. Most cases occur as a mediastinal mass, although about one quarter of cases presented as neck and abdominal masses whether nodal or extra nodal. We present two patients, one is a 58- year old man with localized cervical mass, who was treated successfully by local excision. The other is a 30-year old female complaining from a retroperitoneal mass abutting the caudate lobe of the liver, mistaken as liver haemangioma, which was then successfully removed laparoscopically. More than three years of follow-up followed with no recurrence noticed.

Keywords: Castleman's disease, cervical mass, retroperitoneal mass

(J Med J 2009; Vol. 43 (4):355- 360)

Received

June 11, 2008

Accepted

November 30, 2008

Case 1: A 58-year- old man with a history of asymptomatic left neck mass that had slowly grown over 7 months. On examination, a hard non-tender mobile mass of 5 x 7cm diameter, limited anteriorly by the lateral border of sternomastoid muscle posteriorly by the medial border of trapezius muscle and inferiorly by the upper border of the left clavicle; and the mass was not related to the thyroid gland.

Laboratory Investigation; including complete blood picture, ESR, serum biochemistry (urea, uric acid, calcium, phosphorus, cholesterol, liver enzymes, alkaline phosphatase, total proteins) all gave results within normal range. Chest radiography, abdominal ultrasound, and thyroid scanning were also normal. Computerized tomography (CT) of chest, abdomen & pelvis were normal.

CT scanning of the neck showed a well-circumscribed deeply-seated soft tissue mass 7 x 5x 5 cm in dimensions. It contains few small central hypo dense area arising from left side of the neck extending from Cervical 2 vertebra, down to the level of C5 (figure 1). The mass displaces the major vessels of the neck posteriorly and medially. Fine needle aspiration of the lesion suggested Hodgkin's disease.

Excisional biopsy of the lesion with the adjacent lymph node revealed on histological examination a reactive process, with a marked plasma cell infiltrate, which stained for kappa and lambda indicating poly clonal plasma cells, with occasional giant cells which were topographically suspicious of Hodgkin's disease but were negative for leukocyte common antigen (LCA), CD30, and CD15A diagnosis of CD of plasma cell type was made, then the patient was discharged and followed up closely.

1. Department of General Surgery, Al Basher Teaching Hospital, Ministry of Health, Amman, Jordan.

* Correspondence should be addressed to:

Abdel Hadi breizat

E-mail: dr_a_breizat@yahoo.com

Figure (1): CT scan neck, deeply seated soft tissue mass, extending from C2-C5.



Figure (1): CT scan neck showed a deeply seated soft tissue mass, extending from C2-C5.

Case 2: A 30-year old female patient presented with one year history of mid back and lower abdominal pain. The back pain was continuous, while the abdominal one was colicky in nature. A history of weight loss of 7 kg over the last 6 months was reported. On examination, she was confirmed as a pale female patient with a non-jaundiced normal head, neck, chest and abdominal examination. Lab investigation revealed a haemoglobin: 10.6 g/Dl, ESR: -105-127mm/h. blood film showed mild hypochromic, microcytic red cells, with hypersegmented neutrophils, and normal other parameters. In addition to normal biochemical investigations, C-reactive protein was positive. Abdominal US revealed a mass separated from the liver, near the caudate lobe (figure 2).

Figure (2): A mass near the caudate lobe of the liver.



Figure (2): MRI and a suspicious segment 1 liver lesion

Liver isotope scan revealed an area of decreased tracer uptake in the left lobe of the liver, labeled RBC; it did not reveal any tracer concentration in cold area a conclusion of a space occupying lesion negative for hemangioma .

A diagnostic laparoscopy done revealed a localized non-hepatic mass, retroperitoneal just underneath the caudate lobe of the liver. That was successfully removed laparoscopically.

Histopathology; an oval 4x3.5x3.5 cm firm well-circumscribed mass, microscopically: a huge lymph node with marked follicular hyperplasia. The sinuses were infiltrated by sheets of plasma cells with many Russell's body; with its features being those of giant lymph node hyperplasia of plasma cell type (CD). Figure 3(A, B, C)

Figure (3A): Follicular hyperplasia.

Figure(3B): Sheets of plasma cell infiltrate.

Figure (3C): Few giant cells.

Patient had uneventful postoperative period, c reactive protein was returned negative; and anaemia corrected; Haemoglobin 12.6gm/dl and was followed between the haematological department (were bone marrow aspirate and biopsy done and was normal). More than three years of follow-up followed with no recurrence and good health.

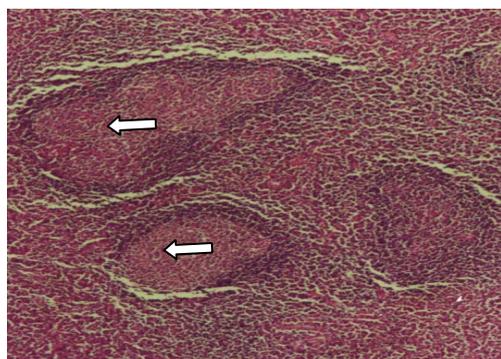


Figure (3, A): Follicular hyperplasia.

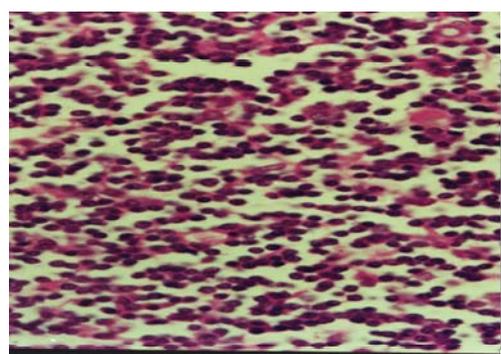


Figure (3B): Sheets of plasma cell infiltrate.

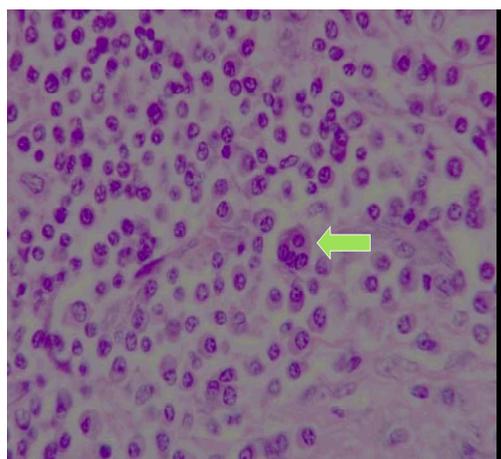


Figure (3C): Few giant cells.

Introduction

Castleman's disease (CD), is an uncommon lympho-proliferative disorder, also known as angiofollicular lymph node hyperplasia. It was first reported by Symmers in 1921.¹ B. Castleman first characterized this pathology in 1956 as a benign lymph node hyperplasia resembling a thymoma.² It can be found anywhere along the lymphatic chain, although the head and neck are second only to the mediastinum. The mediastinum is affected in 70% of cases, and only 6% affect the neck,³ while the abdomen is affected in 12%, and the least common is the axilla 4%.⁴ Based on the biological behavior, CD is classified into two clinical subtypes: a localized (or unicentric) and a multicentric subtype. Localized disease manifests as a solitary mass, which may be well circumscribed or infiltrative. It is associated with lymphadenopathy confined to one lymph node or nodal area and usually follows a benign course. Multicentric disease carries a worse prognosis, and subsequent infection or malignancy that may lead to death.⁵ The cause and pathogenesis of this disease is thought to be due to chronic Human herpes virus 8(HSV8);⁶ as HSV8 has been found in lymphoid cells in cases of systemic form, or plasma cell (PC) type of (CD). Its nature is not neoplastic as confirmed by the fact that the lesion are made of a polyclonal proliferation. Histologically, CD have been categorized into two subgroups, the hyaline-vascular subtype is more common (90% of cases) and is usually asymptomatic and less frequent than the plasma-cell subtype which is often associated with systemic manifestations.⁷

Discussion

In the head and neck, CD most commonly presents as asymptomatic solitary mass located deep to the sternomastoid muscle with a duration varying between 2 months to 10 years.⁸ In the presented neck mass, it was 7 months until patient presented, when neck discomfort increased. There is no sex predilection in CD, and the age ranged between 8 years and 66 years with the youngest patient being diagnosed 6 weeks

after birth.^{9,10} CD of the head and neck is often a diagnostic challenge, which can be attributed to a paucity of signs and symptoms, the absence of specific diagnostic markers and the ability of CD to mimic other neoplasms of the head and neck.

Diagnostic imaging methods such as ultrasound, CT scanning and MRI are helpful in delineating the extent of the mass but they cannot identify CD because of the lack of tumor-specific signs.¹¹ Gallium scintigraphy is considered a sensitive tool for the diagnosis and detection of the hyaline-vascular type of CD, but its utility in detecting the plasma cell variant is debated.¹²

Histological evaluation is the only way to make a definitive diagnosis.¹³ Every effort should be made to remove the lesion totally, since it is curable and no recurrences have been reported after its total excision in the literatures.^{14,15}

Total excision is accomplished in our neck mass patient, close follow up through more than 4 years proved no recurrence.

Most intra-abdominal lesions are located in the pelvis, mesenteric and perinephric regions but such lesions have been described throughout the abdomen (16). The histology of plasma cell type (PC) is not specific of systemic CD and can be found in autoimmune disease, AIDS and in lymph nodes draining carcinoma, so it is imperious to exclude this condition before diagnosing CD of PC type.¹⁷ This implies that serologic testing for HIV should be performed whenever a diagnosis of CD is contemplated, and in our cases the strict pre and postoperative work up with the follow up was important to exclude other pathology, for being a localized plasma cell type.

Pascal Bucher et al.¹⁸ review in 2005, 195 cases of localized CD have been reported in the world literature, arising in the abdomen and retroperitoneum. Of these 195 cases, 122 (63%) were in the retroperitoneum and 73 (37%) in the abdominal cavity. Of the 122 lesions localized in the retroperitoneum, 24 (20%) were in the perirenal region, in the presented case it is a

retroperitoneal lesion related to caudate lobe of the liver and this position was peculiar to our patient not mentioned in other literatures. As mentioned above, en-bloc resection is the standard therapy & laparoscopic excision was proved successful, and may be this is the only case which is laparoscopically removed retroperitoneal CD. A long term follow up in these patients is necessary, and the association of CD with Kaposi sarcoma that can appear as late as 8 years can be explained by the common pathogen in the two disorders, Human herpes virus (HSV8),^{6,16} and it has been now 3 years with no recurrence.

Conclusion

Castleman's disease (CD) is a rare lymphoid disorder which could present in two forms: the localized and the systemic. The localized form has a unique indolent lymph node hyperplasia; which can be found in the abdomen, retroperitoneum or any lymph node basin; as a solitary mass. Localized CD is radiologically nearly undistinguishable from malignant neoplasms. A good preoperative work-up and an open biopsy during surgery, for abdominal and retroperitoneal mass if no diagnosis has been established, can help to avoid extensive resection when facing this benign disorder. Complete surgical excision is curative; laparoscopic resection proved successful especially after a complete resection. The prognosis is excellent. Long term follow up is required in these patients in spite of the benign nature of the disorder.

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مرض كاسلمان: تقرير حالتين مرضيتين مع مراجعة أدبية

عبد الهادي بريزات، أمل الطحاينة

قسم الجراحة العامة، مستشفى البشير التعليمي، وزارة الصحة، عمان، الأردن

الملخص

مرض كاسلمان هو مرض حميد نادر الحصول يصيب الأنسجة اللمفاوية عادة ويتميز بتكاثر غير نموذجي للخلايا اللمفاوية قد يكون في موقع واحد او ينتشر في مناطق مختلفة تشمل المسارات اللمفاوية ضمن العقد اللمفية وخارجها. تحصل معظم الحالات داخل المنصف الصدري على الرغم من أن ربع الحالات قد تصيب الرقبة والبطن. نعرض هنا حالتين الأولى رجل في الثامنة والخمسين من العمر كان يعاني من وجود كتلة في الرقبة، والأخرى امرأة في الثلاثين من العمر كانت تعاني من كتلة أسفل الكبد خلف الغلاف البريتوني. تمت معالجة الحالتين بنجاح؛ الأولى بعملية استئصال كامل للكتلة والثانية رفعت الكتلة عن طريق منظار البطن. تمت متابعة المريضين دون رجوع الورم لأكثر من ثلاث سنوات.

الكلمات الدالة: مرض كاسلمان، كتلة الرقبة، كتلة وراء الغشاء البريتوني