Castleman's Disease—A Rare Diagnosis in the Retroperitoneum

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Abstract Abdominal masses have always been a dilemma for the surgeons and this problem is more so in retroperitoneal masses. In our case, the patient presented with retroperitoneal mass in the left lumbar region with no other specific local or systemic symptoms. The diagnosis of Castleman's disease was established only after histopathological examination of the mass after resection. Castleman's disease is an angiofollicular lymph node hyperplasia presenting either as a localized or a systemic disease. In our case, the patient presented with the localized form of the disease and as it is a rare presentation we are presenting this case.

Keywords Castleman's disease · Retroperitoneal mass · Lymph node hyperplasia

Introduction

Abdominal masses are a clinical enigma and pose a dilemma for the surgeon. Pre-operative diagnosis is always difficult due to varied presentations of the tumors and this problem is more so in retroperitoneal masses.

Castleman's disease (CD) is a rare disorder that presents either as a localized disease or a systemic disease. It represents a morphologically distinct form of 'angiofollicular lymph node hyperplasia'.

Case Summary

A 23 year old male presented with complaints of pain in the abdomen for 15 days and a lump to the left of the umbilicus since 15 days. Pain was dull aching in nature, not associated with vomiting. Patient had no urinary complaints. On examination, vitals were stable, a lump of 8×5 cm was palpable to the left of the umbilicus, firm in consistency with smooth surface and well defined superior, inferior and lateral margins. Rest of the systemic examination was within normal limits. All routine investigations were within normal limits, HIV 1 & 2 were negative. 24 h urine catecholamine levels were normal.

Ultrasonography of the abdomen showed a large well defined, echo poor, solid mass lesion adjacent to the aorta on the left side, approaching the left kidney. CT Scan showed a 8.7×7.7×4.8 cm solid homogenous benign mass primarily of retroperitoneal origin.

The patient underwent exploratory laparotomy. A highly vascular mass was seen lying adjacent to the aorta on the left side extending up to the upper pole of the left kidney (Fig. 1). The mass was excised en bloc and sent for histopathological examination, which revealed 'Localized CD of the hyaline vascular type'.

The post-operative period was uneventful. The patient is free of disease at 1-year follow-up.

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Fig. 1 Hypervascular mass approaching the aorta

Discussion

CD was first reported by Symmers in 1921 and the pathology characterized by Benjamin Castleman in 1956 as a benign lymph node hyperplasia [1, 2]. Keller et al. identified two pathologic types of CD in 1972 [3].

The Hyaline vascular (HV) type, as in our case, presents as a pathological and extensively hypertrophied lymph node. Macroscopically it appears as an encapsulated homogenous mass with an orange-yellowish colour. Microscopically, is characterized by giant lymph follicles centered on a central vessel with marked hyalinization, as seen in our case (Fig. 2). Follicles are surrounded by circumferentially arranged layer of small polyclonal B-lymphocytes, in an onion skin feature [4]. It may present with mass effect symptoms such as anorexia, vomiting, weight loss, urinary retention, abdominal or lumbar pain or a palpable mass. Treatment is complete surgical excision and no recurrences have been reported after complete resection till date, as in our case [5]. The prognosis is excellent with a 5-year survival rate of nearly 100% [6].

Fig. 2 Histopathological appearance of hyalinized type of CD

Regressive follicle with hyalinised blood vessel in the center.

The interfollicular area with extensive expansion and vascular proliferation & dendritic cells.

The localized form of CD arises predominantly in the mediastinum, where it was first described by Castleman [2]. Other sites include neck, abdomen, axilla, inguinal region and in virtually all lymph node areas [5–8]. Testa et al. have reported 315 cases of localized CD; 65% were in the mediastinum, 16% in the neck, 12% in the abdomen, 3% in the axilla and 4% in diverse locations [8]. Bucher et al. reported 195 cases of localized CD, arising in the abdomen and retroperitoneum of which 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 peri-renal region, as in our case [6].

The plasma cell (PC) type presents with systemic symptoms such as fever, anaemia, weight loss, elevated ESR levels, hypergammaglobulinemia or hypoalbuminemia. The prognosis is poor with a median survival of 30 months [6].

The histology of both the types are not always clearly separated and mixed HV-PC types can also occur [5].

The etiology of CD is related to chronic human herpes virus 8 (HSV8) also known as Kaposi's sarcoma-associated herpes virus (KSHV) and is not neoplastic as confirmed by the fact that lesions are made of polyclonal proliferation [5]. It seems that CD is the result of a chronic low grade inflammatory process triggered by latent infection with HSV8, which leads to lymphoid system hyperplasia and also stimulates secretion of IL-6 [5].

Conclusion

Localized CD should be considered when facing a solid hypervascular abdominal or retroperitoneal mass. A better knowledge of this disorder and its characteristics would help the surgeon to avoid unnecessarily extensive resection for this benign disorder when dealing with abdominal or retroperitoneal tumors.

References

1. Symmers D (1921) Primary hemangiolymphoma of the hemal nodes: an unusual variety of malignant tumour. *Arch Intern Med* 28:467–474
2. Castleman B, Iverson L, Menendez VP (1956) Localized mediastinal lymph-node hyperplasia resembling thymoma. *Cancer* 9:822–830
3. Keller AR, Hochholzer L, Castleman B (1972) Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer* 29:670–683
4. Herrada J, Cabanillas F, Rice L, Manning J, Pugh W (1998) The clinical behavior of localized and multicentric Castleman disease. *Ann Intern Med* 128:657–666
5. Larroche C, Cacoub P, Godeau P (1996) La maladie de Castleman. *Rev Méd Interne* 17:1003–1013
6. Bucher P, Chassot G, Zufferey G, Ris F, Huber O, Morel P (2005) Surgical management of abdominal and retroperitoneal Castleman's disease. *World J Surg Oncol* 3:33–38
7. Fields S, Bar-ziv J, Portnoy O, Sasson T, Sherman Y, Libson E (1995) Radiologic spectrum of localised Castleman's disease. *Isr J Med Sci* 31:660–669
8. Testa P, Pigne A, Voinnesson A, Vieillefond A, Paillas J (1980) Les hyperplasies lymphoïdes angiofolliculaires (maladie de Castleman) Première localisation meso-sigmoïdienne. *Chirurgie* 106:156–160