CASE REPORT

Retroperitoneal Castleman's disease: benign in the midst of malignancy

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SUMMARY

A 32-year-old asymptomatic woman presented with localised mass on the left side of the abdomen for 8 months. An abdominal ultrasound and CT scan revealed a solid heterogenous mass located just below the left kidney in the retroperitoneum. Provisional diagnosis of soft tissue sarcoma was made. Following this, surgery was planned and open biopsy (frozen section) was performed, which demonstrated localised hyaline-vascular type of Castleman's disease, which was then excised completely. Thus, intraoperative diagnosis avoided extensive resection and radiation therapy, which reduced the patient's morbidity and expenses. Postoperative period was uneventful and no recurrence was seen even after 3 years of follow-up.

BACKGROUND

Castleman's disease (CD), or angiofollicular lymph node hyperplasia, is an uncommon condition distinguished by the development of benign lymph node masses. Symmers in 1921 first described it and later in 1956 Castleman, *et al* characterised it as benign lymph node hyperplasia.^{1 2} CD is classified into unicentric CD (UCD) and multicentric CD (MCD). MCD is marked by the release of cytokines such as interleukin (IL) 6 from the involved lymph nodes, which induces systemic inflammatory symptoms and has a worse prognosis. UCD is a benign disease, seen mostly in the mediastinum and rarely recognised in the retroperitoneum. Retroperitoneal CD accounts for only 7% of all 400 cases reported so far, so preoperative diagnosis of retroperitoneal CD is very challenging.³ We should emphasise on this benign disease and keep it as an important differential in the list of retroperitoneal masses that are commonly malignant and need aggressive management. Proper preoperative workup and intraoperative biopsy help us in reaching the diagnosis and precludes extensive resection, thus decreasing associated morbidity, need of radiotherapy and agony of the patient related to malignancy.

CASE PRESENTATION

A 32-year-old Asian woman, homemaker, presented with a mass in the left upper abdomen since 8 months. Her medical and family history was not significant. On systemic review, the patient was of average built with no associated pallor, lymphadenopathy or pedal oedema. There was a palpable lump (11×8 cms) in the left upper abdomen, which was firm and non-tender.

INVESTIGATIONS

Laboratory tests showed normal blood profile (Haemoglobin 12.5 g/dL), biochemistry, C reactive protein, lactic dehydrogenase, α -fetoprotein, β -human chorionic gonadotropin; and viral markers (HIV-1, HIV-2, hepatitis B virus and hepatitis C virus) were negative. Abdominal ultrasound showed a mixed echogenic solid mass next to the left kidney. CT revealed a well-defined, 9×7.5×8.5 cm heterogenous, hypervascular mass in the anterior pararenal fascial plane with stellate areas of nodular calcification and multiple feeding vessels. The mass had displaced the small bowel anteriorly, the pancreatic tail and left kidney superiorly with moderate left hydronephrosis and was extending up to the L5 vertebra inferiorly (figure 1). The chest CT was normal.

DIFFERENTIAL DIAGNOSIS

A preoperative diagnosis of malignant retroperitoneal tumour was made on the basis of the imaging findings and surgery was planned.

The important points in favour of this provisional diagnosis were its hypervascularity on radiological imaging and the higher frequency of malignancy in retroperitoneal tumours. The most commonly seen retroperitoneal tumours are

- Soft tissue sarcomas—liposarcoma, leiomyosarcoma, neurofibrosarcoma, rhabdomyosarcoma and undifferentiated sarcoma.
- These are seen as hypervascular heterogeneous masses with necrosis on CT scan.
- Vascular tumours—haemangiosarcoma and lymphangiosarcoma.
- They are usually cystic with low density on ultrasonography (USG) and CT scan.

Figure 1 Contrast-enhanced CT of the abdomen showing heterogenous, hypervascular mass in the left anterior pararenal fascial plane with few internal stellate areas of nodular calcification.

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Figure 2 Encapsulated, tan, elastic soft oval tumour sized 10×8×9 cm.

- ▶ Lymphoma that presents as multiple adenopathy with homogeneous enhancement and a well-defined margin without necrosis on CT scan.
- ▶ Urological tumours such as seminoma, prostatic cancer and teratoma may present as metastatic retroperitoneal lymphadenopathy.

TREATMENT

Preoperatively, angioembolisation was performed to minimise intraoperative bleeding. Moreover, a cystopanendoscopy was performed to assess ureteric involvement, which was found to be absent and ureteric stenting was carried out to decompress the renal system.

Laparotomy was performed, which revealed a 15×10 cm large, firm, irregular yellowish brown mass in the left retroperitoneum situated anterior to the left kidney and ureter, adherent to paraspinal muscles posteriorly, extending from the left renal vein to the common iliac bifurcation. Multiple feeders were seen arising from the aorta and the lumbar arteries which were ligated. Before excision, a frozen section was taken, which showed lymphoid lesion with no evidence of malignancy. Tumour was excised en bloc and sent for pathological evaluation. The total blood loss was 500 mL and no blood transfusion was given. On gross examination, the tumour was encapsulated, tan, soft, elastic and oval shaped and sized 10×8×9 cm (figure 2). The cut surface was dark red with a central white zone of fibrosis and calcification (figure 3).

Figure 3 Cut surface—dark red with a central white zone of fibrosis and calcification.

Figure 4 Microscopic picture showing central perforating vessels surrounded by lymphocytes (H&E stain ×400 magnification).

Histopathological studies exhibited hyaline-vascular follicles with highly vascularised interfollicular areas and numerous mature lymphocytes. No malignant cells were seen. Immunohistochemistry revealed that the tissue was positive for B-cell marker CD79a and negative for T-cell marker CD3. These histological findings were in accordance with a localised type of CD (figure 4).

OUTCOME AND FOLLOW-UP

On account of the benign nature of this disorder, the patient was not subjected to radiotherapy, which is usually given in the management of retroperitoneal soft tissue tumours. The patient had smooth postoperative recovery and there was no recurrence even after 3 years of follow-up.

DISCUSSION

CD is a non-clonal disorder of lymph nodes, mostly seen in adults, with age 30 years or less but may also be seen in children. CD is classified into UCD and MCD.

It has two histological subtypes: the hyaline-vascular type (85–90%) and the plasma cell type though a mixed type also occurs.⁴ The hyaline-vascular type is generally unifocal, asymptomatic and fortuitously diagnosed on USG or CT scans. Larger ones present with compressive symptoms such as breathlessness, cough, icterus, postprandial uneasiness, anorexia, emesis, neuropathy and voiding difficulty.⁵ It is predominantly seen in the third decade in otherwise normal individuals. Histologically, lymph nodes are substituted by follicles with hyalinisation of its wall and concentric whirls of mantle lymphocytes in an onion skin pattern. Surgical resection is the benchmark treatment, with nearly 100% 5-year disease-free survival.

The plasma cell type (10%) is commonly related with the multifocal form. Histologically, it presents as hyaline-vascular type with sheets of plasma cells. It is primarily seen in the sixth decade of life with systemic features⁶ as pallor, debility, pyrexia, weight loss, oedema, generalised lymphadenopathy, hepatosplenomegaly, pleural effusion and worsening renal function. It may be infrequently associated with amyloidosis or POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, M protein and Skin changes). The pathogenesis of plasma cell type is a persistent low-grade inflammation caused by HHV-8 infection in the lymph nodes, leading to IL-6 secretion and hyperplastic reaction of the lymphoid system. This condition is mostly seen in immunocompromised patients and is associated

with poor prognosis.⁷ It is usually managed by a combination of steroids, rituximab, chemotherapeutic agents and radiotherapy.

CD is a very rare differential of retroperitoneal tumours and is mostly indistinguishable from other diseases despite preoperative radiographic workups and intraoperative finding, which makes its prior diagnosis difficult. CD is seen as a homogeneously hypoechoic mass on USG and non-specific enhancing homogeneous mass with star-shaped microcalcifications on CT, both of which are not diagnostic. Although, MRI is superior as it shows better soft tissue delineation, but is also not definitive for the diagnosis of CD.⁸

In our case, the provisional diagnosis of soft tissue sarcoma was made due to retroperitoneal location of the tumour and hypervascular and heterogenous appearance on CT. As retroperitoneum hosts a wide range of pathologies, including a variety of rare benign and malignant tumours, the incidence of malignant neoplasm of the retroperitoneum is four times that of benign tumours. Sarcomas encompass one-third of retroperitoneal tumours, with two histopathological subtypes prevailing, specifically liposarcoma (70%) and leiomyosarcoma (15%). Other retroperitoneal lesions incorporate primary lymphoproliferative tumours (Hodgkin's and non-Hodgkin's lymphoma) and epithelial tumours (renal, adrenal, pancreas) or might illustrate metastatic disease from known or unknown primary sites (testicular tumours, prostatic carcinoma, melanomas). Benign

tumours can cause consternation and are usually an incidental finding during an evaluation for unrelated symptoms. They may be mentioned on intuition of being a sarcoma. The most common benign pathologies encountered in the retroperitoneum include benign neurogenic tumours, paragangliomas, fibromatosis, renal angiomyolipomas and benign retroperitoneal lipomas.⁹

Preoperative fine-needle aspiration cytology has no role in diagnosis because of its low specificity and risk of tumoural seeding (1/40 000–1/1000).¹⁰ So, definitive diagnosis can only be made by open biopsy at the time of surgery, as in our case, which came out to be localised type of CD which was then excised completely. Thus prior diagnosis avoids the need for extensive resection and radiation therapy, which furthermore reduces patient's morbidity, expenses and associated torment related to malignancy.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- Castleman's disease is a rare entity and uncommonly seen in the retroperitoneum.
- It is a benign tumour in the midst of malignant neoplasms, which are usually found in the retroperitoneum.
- Definitive diagnosis can only be made by intraoperative frozen section, which avoids the need for extensive resection and radiotherapy.
- It allays the agony of the patient associated with the diagnosis of malignancy.
- The patient has a 100% disease-free survival after complete resection.

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