

operative anaesthetic care, we can obtain a better definition of the tumour mass pre-operatively and accept the challenge of demanding surgical procedures. Retroperitoneal lipomata are uncommon and have an indolent presentation. Lipomas occurring in this site are

a different proposition than those in other sites and require meticulous pre-operative evaluation and an exacting surgical approach if recurrence and potential malignant transformation is to be avoided.

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Pelvic mass in 46-year old man

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A 46-year-old man was referred from the Urology department with a pelvic mass discovered on abdominal ultrasonography performed to study dysuria and abdominal pain. Findings from physical examination were unremarkable, without evidence of hepatosplenomegaly or lymphadenopathy. Results of laboratory investigations, electrocardiogram and chest radiography were normal. The serum and urine immunoelectrophoretic patterns showed no monoclonal components. An enzyme-linked immunosorbent assay test for human immunodeficiency virus was negative.

Abdominal ultrasonography revealed a round solid mass in the right iliac fossa. Thoracic and abdominal contrast-enhanced computed tomography (CT) showed a 5-cm pelvic mass perfectly limited and localised between bladder and iliac vessels (figure 1), without other abnormalities. This mass was punctured. A percutaneous transabdominal fine-needle aspiration biopsy specimen showed small mature lymphocytes and some macrophages. Because lymphoma was considered a likely diagnosis, laparotomy was performed, with removal of the pelvic mass. The rest of the abdomen was normal. The postoperative course was uneventful. Pathologic studies were diagnostic (figure 2).

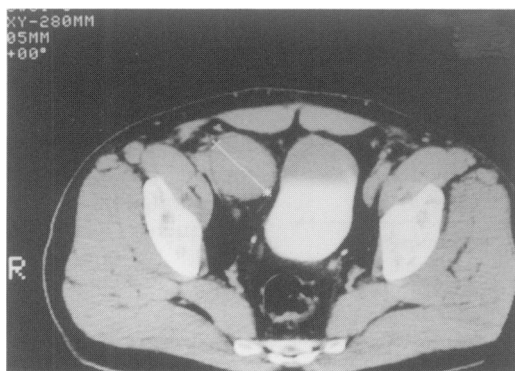


Figure 1

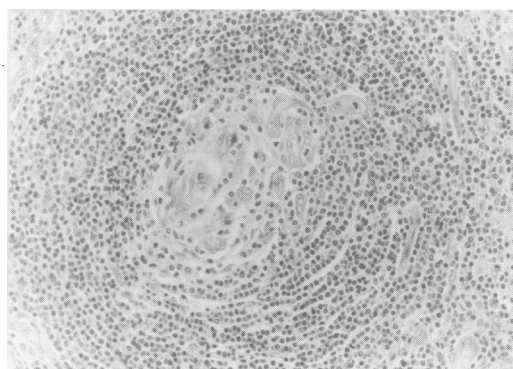


Figure 2

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Questions

- 1 CT scan shows a well-demarcated pelvic mass with a maximum diameter of 5 cm. Suggest three of the more likely diagnoses.
- 2 What abnormality is demonstrated in this histological section?

Answers

QUESTION 1

The differential diagnosis of pelvic masses includes primary or secondary pelvic tumours (bladder carcinoma, seminoma, germinal tumours, neurogenic tumours, benign teratoma, liposarcoma, smooth muscle tumours, fibrous malignant histiocytoma, haemangiopericytoma), inflammatory processes (actinomycosis, pelvic tuberculosis), idiopathic retroperitoneal fibrosis, and lymphoproliferative disorders.

QUESTION 2

Castleman's disease (hyaline vascular type). The lymphoid follicles are surrounded by circumferentially arranged capillaries penetrating the germinal centres. These centres contain deposits of hyaline and the interfollicular stroma has a major proliferation of capillaries, some small lymphocytes and occasional plasma cells.

Discussion

Castleman's disease was first described in 1956, in a patient with a mediastinal mass. Mass histology showed an angiofollicular lymph node hyperplasia. Since then, a number of cases have been described, mainly in the mediastinum (64%) and in the abdomen (14%).^{1,2} To date, only 10 cases have been described in the pelvis (table).³⁻¹² The main features of Castleman's disease are summarised in the box.

The incidence of pelvic Castleman's disease is similar in both sexes. It appears in most patients before the age of 31, although the age range is broad. Abdominal pain is frequently the first symptom manifested by female patients, although dysuria is more commonly reported by male patients. The mass is between 4.5 cm and 15 cm.

In 1972, Keller *et al*¹ recognised two distinct histologic types which differ in their clinical implications. The hyaline vascular (HV) form, which accounts for approximately 90% of

Castleman's disease

- masses in diverse locations
- characterised by massive growth of lymphoid tissue
- two distinct histologic subtypes: hyaline vascular (HV) and plasma cell (PC)
- symptoms are usually local and due to compression
- HV form can be cured by surgery, whereas PC needs chemotherapy

cases, is typically present in asymptomatic patients with a localised mass. It is characterised by a predominance of giant lymph follicles, prominent vessels with marked hyalinisation and concentric layers of lymphocytes arranged around radial vessels and occasional plasma cells. These cells show a polyclonal pattern on immunohistology. Surgery may be curative, although local recurrence is possible. Rare cases have been complicated by the development of vascular neoplasm or progression to malignant lymphoma.¹¹ Thus, long-term patient follow-up is necessary. All cases described in a pelvic location have been the HV type.³⁻¹²

Histologically, the plasma cell (PC) type contains a massive accumulation of polyclonal plasma cells in the interfollicular area. However, a monoclonal plasma cell component has been demonstrated in a few cases.² Patients with multicentric Castleman's disease are older and have constitutional symptoms, anaemia and polyclonal hypergamma-globulinaemia. They have a poor prognosis, despite treatment with corticosteroids and chemotherapy. However, a clear separation of the two variants is not always possible, as cases with 'mixed' histologic features have been described.

Although the histology of Castleman's disease is characteristic, it is not specific. 'Castleman's-disease-like' changes are observed in some illnesses: rheumatoid arthritis, Sjögren's syndrome, neoplasm and acquired immunodeficiency syndrome,² which need to be excluded. Our patient did not have any of these diseases. Pre-operative diagnoses may be difficult, especially if the mass is localised in an unusual place, as in the case of our patient.

Although Castleman's disease is a very infrequent lymphoproliferative disease, it should be considered when a pelvic mass appears.

Final diagnoses

Castleman's disease (HV form).

Keywords: pelvic mass, Castleman's disease, angiofollicular lymphoid hyperplasia

Table Pelvic Castleman's disease

Ref no	Age (years)	Sex	Size (cm)	Clinical manifestations
3	28	M	7 × 5 × 4	at autopsy
4	29	F	4.5	incidental
5	50	M	7.2	urinary
6	37	F	9	abdominal pain
7	21	F	6	incidental
8	19	F	15	abdominal pain
9	17	F	8 × 5 × 4	abdominal pain
10	21	F	8.2 × 1.5	abdominal pain
11	31	M	10	haemolytic anaemia
12	31	M	10 × 8 × 9	urinary

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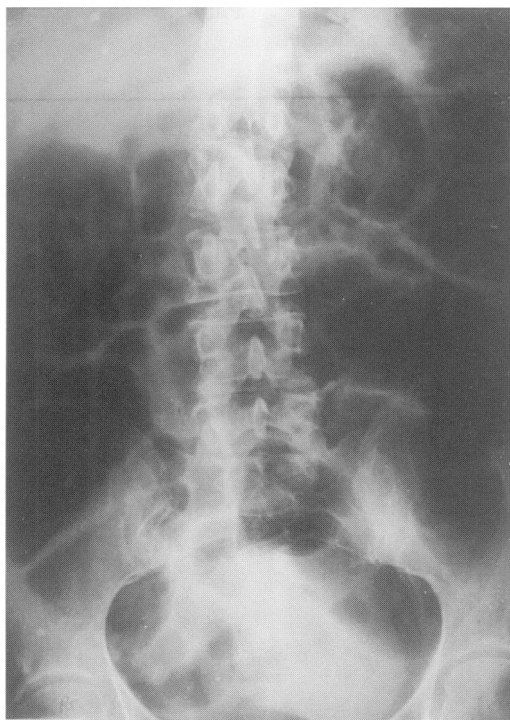
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Hypothyroidism presenting as acute abdomen

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A 47-year-old woman presented as an emergency with a 12-day history of generalised abdominal pain, most severe in the lower abdomen, constipation, abdominal distension and loss of appetite. She had undergone subtotal thyroidectomy for hyperthyroidism 10 years earlier.

On examination she was afebrile, dehydrated, and was haemodynamically stable. Abdomen was distended, there was rebound tenderness in the lower quadrants with tinkling bowel sounds. There was a leucocytosis but her urea and electrolyte levels were within normal limits. Her plain X-ray abdomen is shown in the figure.



Questions

- 1 What is the most probable diagnosis?
- 2 What medical condition may underlie this clinical presentation?

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Figure X-Ray of abdomen (supine)