A high index of suspicion should be kept for this disorder, prompting early diagnosis and treatment in order to prevent brain damage.

Final diagnosis

Primary nephrogenic diabetes insipidus with intracranial calcification

Keywords: nephrogenic diabetes insipidus, hypernatraemic dehydration, intracranial calcification

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Excess adipose and heartburn

WDB Clements, P Rice, C Harvey

A previously well 80-year-old woman presented with a three-month history of increasing epigastric discomfort, abdominal bloating, 14 pounds weight gain and heartburn. On examination she was generally well, although the upper abdomen appeared diffusely distended. There was no discrete mass or organ enlargement palpable, although the upper abdomen was dull to percussion. Haematological and serum biochemical parameters were all within normal limits and urinalysis was normal. Her chest X-ray showed a large hiatus hernia. Computed tomography (CT) scan of the abdomen was performed (figures 1 and 2).

Figure 1 CT scan of abdomen

Figure 2 CT scan of abdomen

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Questions

- 1 What abnormality is shown on the CT scan?
- 2 What is the most probable diagnosis?

Answers

QUESTION 1

The CT scan shows a large homogenous mass arising from the retroperitoneal area. The left kidney is contrast enhanced and normal, however the right kidney is not present in the normal site, inferring its absence or displacement by the mass. In this case it was displaced superiorly into the subhepatic space of Morrison (figure 2). The mass effect of the rapidly growing neoplasm and consequent increase in intra-abdominal pressure may have resulted in hiatal herniation.

QUESTION 2

This is a large retroperitoneal tumour. The homogeneity of the mass and attenuation value of the CT scans suggest that it is of fatty origin. In this case it was a benign lipoma weighing 15 kg (figure 3) which, in this site, is more uncommon than its malignant variant, the liposarcoma. Upon removal of the tumour the patient's symptoms of heartburn regressed and she remains well after two years.

Discussion

Retroperitoneal tumours are a rare group of tumours with deceptive histopathological patterns and unpredictable clinical behaviour, all of which present diagnostic, therapeutic and prognostic problems. They constitute 0.07–0.2% of all neoplasia and 65–86% are malignant,^{1,2} 25% are dysontogenetic (ie, teratoma, dermoid and other cysts), 15% arise from neural elements (ie, benign and malignant neurinoma, neuroblastoma, benign and malignant paraganglioma), and the remainder are of mesenchymal origin. Lipomas and fibrolipomas constitute one third of this group the other two thirds are sarcomas (ie, lymphosarcoma, reticulosarcoma or rhabdomyosarcoma).

The origin of lipomata are unknown. They may represent a congenital anomaly of adipose tissue. It has also been noticed that fat forming mesenchymal cells have the potential to differentiate into a variety of complex tissues, a fact that may be related to the observation that tumour fat is unavailable for nutritional purposes in extremely emaciated patients with huge retroperitoneal lipomas.³ In Dercum's

disease, an autosomally dominant inherited condition, lipomata are multiple, with a propensity for the limbs. They are frequently painful, necessitating excision, and have the potential for malignant transformation in a small number of cases.

Generally, lipoma are considered innocent tumours which only rarely recur after incomplete excision and which exhibit a very low incidence of myxomatous or sarcomatous transformation. Virchow, in 1857, was the first to describe malignant change in a lipoma and the collective series of Von Wohlfendorff in 1921 demonstrated a significant incidence of malignancy in lipomata arising in the retroperitoneum. Since then, other authors have concurred with this finding.⁴

There is little doubt that the retroperitoneal lipoma is a distinctive entity as regards its presentation, histological and clinical behaviour. Their tendency for rapid unrestrained growth is attributed to the lack of resistance offered by adjacent retroperitoneal structures. Consequently, they may reach an enormous size before they are detected. These tumours are usually well encapsulated and exert their clinical effects through pressure on neighbouring structures which may result in the patient's demise through cachexia, ileus or uraemia. In a recent series reported by Serio *et al* palpable abdominal mass with vague pain was the commonest presenting feature (87%), followed by gastrointestinal symptoms (7%), urinary symptoms (7%) and arterial hypertension (4%). Although the presentation is usually indolent with the duration of symptoms ranging from 1–72 months (median 6.5 months) patients may present as an emergency with intestinal obstruction, vascular occlusion or intraperitoneal haemorrhage.⁵

Retroperitoneal lipomata occur most commonly in the 5th and 6th decades of life with a marked female preponderance (3:1), hence the suggestion that the climacteric and incumbent hormonal changes may influence dormant undifferentiated mesenchymal tissue. In support of this theory, it is well recognised that fatty tissue proliferates and spreads both during and after the climacteric. These tumours arise most commonly from the perinephric fat and less frequently from the mesenteric or pararectal fat, therefore 80% are present in the abdominal cavity and 20% in the pelvis. Despite being well encapsulated, between 10 and 35% of cases encompass vital retroperitoneal structures. It is not uncommon for nephrectomy to be necessary to effect complete excision and the complexity and extent of the surgery is reflected in the high morbidity and mortality rates in early series. It was also noted that, where resection was performed, up to 50% recurred within a short space of time. The high local recurrence rates led to the hypothesis that these tumours are multicentric, which is supported by the operative finding of smaller satellite tumours adjacent to the main tumour.

With the development of CT scanning and magnetic resonance imaging, complemented by advances in surgical technology and peri-

Figure 3 The excised lipoma

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?