Symptomatic adrenal myelolipoma: therapeutic considerations


Dipartimento di Chirurgia, Università di Pisa, Via Roma 67, 56100 Pisa, Italia

Four cases of symptomatic myelolipoma of the adrenal gland are presented. Three were treated at laparotomy and one with a laparoscopic approach. On the basis of these four cases we discuss the aetiology, pathogenesis, clinical manifestations, diagnosis and treatment of this rare disease. We also report the follow-up of a patient with bilateral myelolipoma, who underwent monolateral excision.

We conclude that myelolipoma, if correctly diagnosed, can be treated conservatively with careful follow-up, limiting surgery to symptomatic cases.

Key words: myelolipoma; adrenal gland; adrenal masses.

Introduction

Myelolipoma is a rare benign non-functioning tumour of the adrenal gland. It is characterized histologically by the presence of mature adipose tissue containing foci of haematopoietic bone marrow cells. Before the introduction of CT and US these tumours were rarely diagnosed during life. In 1977, 190 cases were reported, most of which had been found incidentally during autopsy. Myelolipoma only becomes symptomatic when large. In our department from 1988 to 1994, 28 incidental adrenal masses were surgically removed. Four cases were symptomatic adrenal myelolipomas. In this study we report details of the findings, diagnosis and surgical strategy of the four cases, one of which was bilateral.

Case reports

Case 1

LF, a 43-year-old woman, had intermittent pain for 8 months in the left upper quadrant radiating to the flank. Her clinical history was not significant and physical examination was negative.

Laboratory studies and results of urinalysis were normal. Serum electrolytes and urea were within normal values. Plasma cortisol, urinary alpha-ketosteroids, catecholamines and urinary VMA were also within normal ranges.

An abdominal ultrasound scan revealed a 10-cm diameter highly echogenic mass in the left adrenal region.

CT revealed a soft tissue mass in the region of the left adrenal gland.

Surgery was performed through a transverse left abdominal incision. The left adrenal mass was exposed after incision of the colo-epiploic ligament and mobilization of the stomach, spleen and pancreatic tail.

The mass measured 11 cm in diameter, was spherical and weighed 90 g. The cut surface revealed a yellow solid tumour with sporadic haemorrhagic areas. Histologically the tumour consisted almost entirely of fat with foci of typical bone marrow elements consisting of precursors of the erythroid and granulocytic series.

The patient was discharged 7 days after surgery.

The patient shows no sign of disease 6 years postoperatively.

Case 2

FA, a 48-year-old man, complained of lumbar tenderness and abdominal discomfort for 5 months. Physical examination of the abdomen was negative. The laboratory and endocrine studies were normal. Ultrasound scan revealed two hyperechogenic masses, of about 5 cm and 2 cm in diameter, respectively, in the right and in the left adrenal gland.

Abdominal CT revealed two circumscribed inhomogeneous fat masses, with low density, in the same area.

Exploration was performed with a transverse upper abdominal incision; only the mass on the right was removed because its dimensions were larger. The tumour was easily dissected from surrounding structures and blood loss was minimal. The total weight of the mass was 38 g. Pathological diagnosis was adrenal myelolipoma. Convalescence was normal, and the patient was discharged a week postoperatively.

The patient was seen regularly for follow-up and 5 years after treatment is healthy and free of symptoms. The left myelolipoma which had not been surgically treated has not undergone volume alterations on ultrasound and tomographic evaluation.

Case 3

DP, a 57-year-old man, was admitted to hospital with a complaint of intermittent lumbar and subcapsular pain
which increased with respiration. Physical examination and laboratory assays of blood and urine did not disclose any abnormality. Determination of adrenal hormones was normal. US revealed a capsulated tumour of 6 cm in diameter. CT scan showed a 4.5 × 3.5 cm mass with regular edge, extremely low in density and with an irregular 1.5 cm area within. A transverse upper abdominal incision was performed and a large (7 × 5 × 3.5 cm) mass was excited in its entirety with the adrenal gland. The weight of the mass was 100 grams. Histological examination confirmed the diagnosis of adrenal myelolipoma with aspects of nodular hyperplasia of the cortex and irregular areas of calcification. The patient's post-operative recovery was uneventful and he is healthy and free of symptoms one year after surgery.

Case 4

TR, a 44-year-old man, was referred with lower abdominal pain radiating to the lumbar region of about 1 year's duration. Physical examination, blood samples and urine assays of adrenal hormones were normal. The ultrasound showed a 10-cm hyperechogenic mass on the upper pole of the right kidney (Fig. 1). CT revealed a mass between the liver and the right kidney that was heterogeneous in density and with little contrast enhancement for the presence of fatty tissue (Fig. 2). The tumour was removed using a laparoscopic approach. The patient represented the 27th case of laparoscopic adrenalectomy in our experience.

The patient and the trocars were placed as Gagner suggested. The operating table was in the anti-trendelenburg position and inclined at 45°. Five trocars were positioned as follows: the first 10 mm trocar at the lower edge of the umbilicus to insert the laparoscope; The second 10 mm trocar in the subcostal position on the right mid-clavicular line in order to use the retractor with five digits. A third 10-mm trocar was positioned at the intersection of the umbilical line and the median right axillary line to insert the Kelly clamp and Babcock forceps. A fourth 10-mm trocar was placed at the same site to insert electrified scissors and a 5-mm trocar at the intersection of the right anterior axillary line and the last rib to insert the aspiratory and washing tube. By cutting the right triangular ligament and opening the posterior peritoneum we accessed the right adrenal region. Intraoperative ultrasound facilitated the investigation of the adrenal vascular pedicle and the relationships to the inferior cava.

Using a sterile plastic bag we extracted the mass whole thus avoiding fragmentation in the peritoneal cavity. The mass measured 10 × 9 × 4 cm and weighed 85 g. The histological diagnosis was myelolipoma (Figs 3, 4). The patient was discharged 3 days post-operatively after a satisfactory recovery.

Discussion

Adrenal myelolipoma affects both males and females, with a higher occurrence around the fifth and sixth decades of life, although in the literature there are cases in the pediatric and geriatric ages. In most cases, myelolipoma is unilateral, rarely bilateral. In a review of 59 well-documented cases of surgically removed adrenal myelolipoma only one patient had bilateral tumours.

The size of these tumours vary from a diameter of only a few mm to more than 15 cm. On average, when diagnosed, their diameter is 5 cm, although tumours of 34 cm and weighing 5500 gr have been reported. This neoplasia exists in 0.08-0.2% of autopsy reports and it represents about 8% of adrenal incidentalomas. They are usually asympto-
FIG. 2. Abdomen CT: voluminous, heterogeneous mass of the right adrenal gland (7.5 × 9, 5 × 7.5 cm) with little contrast enhancement for the presence of fatty tissue (Case 4).

FIG. 3. Section from adrenal myelolipoma (× 65) (Case 4): mature fat and hematopoietic tissue displacing adrenal cortical cells.

FIG. 4. Section from adrenal myelolipoma (× 65) (Case 4): hematopoietic tissue containing cells at different stage of maturation.

Symptomatic adrenal myelolipoma

Svmptomatic and have no endocrine activity. Non-specific abdominal pain is the most common symptom, secondary to mechanical compression from large tumour bulk, hemorrhage or intratumoral necrosis. There are also reports of myelolipomas presenting with unusual epigastric pain. Rare symptoms can include haematuria and hypertension. In the literature a large number of cases of an association between myelolipoma and endocrine diseases have been reported such as pathological obesity, hermaphroditism, pseudohermaphroditism, polyendocrine insufficiency, Addison's disease, Nelson's syndrome, Cushing's disease and Conn's syndrome.

Endocrine investigations in order to differentiate functioning from non-functioning tumours of the adrenal gland include serum cortisol, serum catecholamines, 24-hour urinary VMA, metanephrines, free cortisol, 17-hydroxysteroids, 17-ketosteroids and plasma aldosterone levels. The aetiopathogenesis of this disease is unknown, although a large number of theories have been put forward: ectopia of myeloid tissue, embolism of bone marrow elements, hamartosis, metaplasia, degeneration of cortical cells, hematopoietic proliferation of substances formed by tumoral necrosis.

Grossly, myelolipoma is a yellowish, roundish, well-circumscribed mass, even if it does not have a true capsule. Microscopically, myelolipomas consist of mature adipose tissue with scattered islands of hematopoietic cells, more frequently belonging to the granulocyte series, but also to the astrocytes; lymphocytes and megalokaryocytes are rare. Patients with myelolipoma do not have hematopoietic diseases and have normal bone marrow.

Before ultrasound and CT, myelolipomas could be identified only when a standard abdominal X-ray showed a suprarenal, transparent mass. Now the imaging allows more and
more frequent diagnosis of these unusual adrenal tumours. The sonographic and tomographic appearance of myelolipoma depends on the structural characteristics of the tissues that it is composed of. These lesions can contain adipose and myeloid tissue in different proportions. The tumours with a higher adipose content are more correctly identified with US and CT. On the contrary, myelolipoma containing less fat and more myeloid tissue, hemorrhagic areas and sporadic calcifications are more difficult to differentiate from other adrenal neoplasms. More often these lesions are solid masses with well-defined edges. They are hyperechogenic on US and low in density on CT; very rarely the masses are composed of myeloid tissue which would show up as hypechoegenic on US and have CT densities well within the range of fat before contrast enhancement.

Images after contrast enhancement give only an improvement in detecting the edges of the lesion in respect to the adjacent organs, the liver and the kidney, but they do not provide any other information. In spite of the sensitivity of US in showing adrenal masses greater than 2 cm, 2,5 echo-pattern of myelolipoma is not pathognomonic, thus it is necessary to verify it with CT or MRI. CT is the most precise procedure for detecting adrenal myelolipomas. MRI sagittal and coronary sections are more precise than CT to detect these tumours. MRI can distinguish between adrenal haemorrhages, adenomas, metastases and pheochromocytomas. On MRI, myelolipomas usually have an increased intensity relative to the liver on both short and long TR/TE sequences depending on the amount of fat. 19 They may be inhomogeneous or low in intensity, depending on the percentage of other tissue components present in the tumour. The differential diagnosis of a suprarenal fatty mass includes myelolipoma, renal angiomyolipoma expanding from the upper pole and retroperitoneal lipoma, which are entirely fat-containing masses, whereby myelolipomas are a mixture of fat and other tissue. Liposarcomas of the adrenal gland or retroperitoneum are large lesions and should not be confused with smaller and well-defined myelolipomas.

Angiomyolipoma are very vascular tumours and can be diagnosed using angiography. The differential diagnosis between liposarcoma and myelolipoma is often impossible. Green reported a case of adenocarcinoma metastatic to the adrenal gland simulating myelolipoma. The lesion infiltrated the number of patients who are subjected to a long follow-up because of non-functioning adrenal masses.

References

report of two cases with a review of the literature. Surgery 1986; 99: 293–301.

Accepted for publication 11 April 1995