Exophytic renal angiomyolipoma

A case report

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Summary

A case of a large exophytic renal angiomyolipoma is presented. Despite the features on excretory urography suggestive of an extrarenal mass, computed tomography provided information concerning the origin and pathological nature of the lesion.


Renal angiomyolipoma (AML) is a benign tumour (hamartoma) composed of fat, smooth muscle and blood vessels, in varying proportions. AMLs are commonly found in association with tuberous sclerosis (TS), presenting in 80% of these patients as small multiple often bilateral intrarenal lesions. However, 75% of tumours are solitary and unilateral lesions in patients with no evidence of TS; in TS they tend to be large and symptomatic. Twenty-five per cent of AMLs predominantly grow extrarenally, extending into the perirenal space. Although never extending beyond the perirenal fascia, their growth may be exuberant.

A case of a large exophytic renal AML demonstrating the role of computed tomography (CT) in the pre-operative diagnosis is presented.

Case report

A previously healthy, 47-year-old woman presented with severe, colicky left lumbar pain radiating to the left hypochondrium, low grade pyrexia and anaemia. There was no associated dysuria or haematuria. Examination revealed a large tender fluctuant mass distending the left hypochondrium. Excretory urography demonstrated good functioning kidneys with normal pelvicaliceal systems, and inferomedial displacement of a slightly enlarged left kidney. Crenation of the lateral cortical margin of the left kidney was noted (Fig. 1). Ultrasonography showed a large complex pararenal retroperitoneal mass with extensive hyperechoic areas consistent with fat density. CT demonstrated a large pararenal mass confined to the perirenal space, arising from the lateral renal cortex. The mass was of heterogeneous density with large areas of fat density (Fig. 2a). CT after the administration of contrast medium showed a tortuous vascular network coursing throughout the fatty tumour component, with homogeneous contrast enhancement of the soft tissue density component (Figs 2b and 2c). There was no evidence of invasion of adjacent structures.

Surgery revealed a large fatty retroperitoneal tumour arising from the lateral border of the left kidney, displacing it posteromedially. The tumour was displacing the splenic flexure of the colon anteromedially, and the spleen and stomach superomedially and anteriorly. The tumour was resected with preservation of the kidney. It consisted of mature fat cells, spindle and muscle cells with many blood vessels. No identifiable lipoblasts or mitotic figures were noted. These features were in keeping with a diagnosis of AML. The postoperative course was uneventful.

Discussion

Benign renal tumours represent less than 5% of all renal neoplasms. AML is an uncommon benign tumour noted in only 27 cases in a pathological series of 8,501 renal tumours.

A gradual increase in size and haemorrhagic complications account for symptoms in AML, 96% of which necessitate medical attention. The most common presenting symptom is loin pain found in 75% of cases, followed by loin mass in 41%, haematuria in 30% and hypotension in 18%. Clinical and operative differentiation between AML, renal carcinoma, and renal or perirenal sarcomas, particularly lipo-
Fig. 2. (a) Renal CT before administration of contrast medium demonstrates a large heterogeneous mass with significant fat density arising from the lateral margin of the left kidney. (b) Renal CT after contrast medium administration shows a tortuous vascular network coursing throughout the fatty tumour component (arrow), with homogeneous contrast enhancement of the soft tissue component. (c) Magnification of Fig. 2b — intact perirenal fascia with no invasion of adjacent visceral structures.

sarcoma, may be difficult, but is essential because the latter conditions require a radical surgical approach in most instances, whereas AML does not. Thus pre-operative diagnosis based on radiography and histology is essential to management. In only 10% of patients is the characteristic fat density of AML discernible on plain films, often only with the aid of CT. Such radiolucency may, however, also be noted in retroperitoneal liposarcomas.

Excretory urography does not permit differentiation between renal carcinoma and AML, as both present as mass lesions replacing renal parenchyma, while displacing and distorting the pelvicaliceal system. Both exophytic renal AML and perinephric liposarcoma can present as a mass displacing the kidney, with normal pelvicaliceal systems on excretory urography.

The signs found on angiography, although suggestive of AML, are unreliable in differentiating between a benign and malignant lesion.

Most renal AMLs demonstrate a predominance of fat, thus ultrasonography, particularly in large lesions demonstrating dense echogenicity, should suggest a diagnosis of AML. However, in a minority of cases, where fat is not the major component, haemorrhage obscures the lesion, or growth is mostly extrarenal, ultrasonography may not help.

CT, through its ability to detect fat density in a renal lesion, will provide an accurate means of distinguishing an intrarenal AML from carcinoma in most instances. Distinction between exophytic renal AML and liposarcoma is more difficult, since both demonstrate fat density on CT, 35% of liposarcomas arise within the perirenal space, and liposarcomas present a spectrum of histological grades of malignancy. CT identifies the perirenal fascia clearly, thus allowing distinction between an exophytic AML and a liposarcoma arising outside the perirenal fascia. In poorly differentiated liposarcoma, renal invasion is commoner, and the tumour presents with varying amounts of fibrofatty tissue. These lesions may resemble exophytic AMLs and differentiation will depend upon the detection of invasion of the tumour through the perirenal fascia. In well-differentiated lesions growth is by local expansion, they have a predominance of fat, and when arising within the perinephric space, do not demonstrate renal invasion. In these cases distinction between them is possible. Intermediate grade liposarcomas demonstrate significant fat density, and may invade the renal parenchyma; thus differentiation from AML will depend upon the presence of extension beyond the perirenal fascia, which may not always be present despite renal invasion.

Despite the sometimes florid extrarenal growth of some AMLs, as in this case, in most instances CT allows accurate discrimination between such lesions, renal carcinoma and perirenal liposarcoma.

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REFERENCES

Cervical osteophytes and respiratory failure

An unusual case of upper airway obstruction

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Summary

An unusual case is presented in which large cervical osteophytes caused upper airway obstruction. The presenting features of acute-on-chronic respiratory failure and cor pulmonale were alleviated by permanent tracheostomy.

Narrowing of the upper airway, which may occur at any level between the nose and the carina, may lead to respiratory distress, obstructive sleep-induced apnoea and cardiovascular complications.

Case report

A 63-year-old man presented to the emergency unit of Groote Schuur Hospital with a 3-month history of increasing inspiratory difficulty, orthopnoea and peripheral oedema. He had been a heavy smoker in the past and had been hoarse for the past 17 years.

He was unable to speak, was distressed, stridulous, and cyanosed, with a respiratory rate of 30/min. His pulse rate was 100/min; blood pressure 90/40 mmHg; jugular venous pressure elevated 6 cm above the clavicle. He had oedema of the sacrum and pedal oedema extending up to his knees. Chest movement was poor and a tracheal tug was noted. Auscultation revealed inspiratory and expiratory wheezing, and bilaterally reduced air entry. A loud P2 sound and evidence of tricuspid incompetence were also noted. Abdominal examination revealed a tender pulsatile hepatomegaly.

An ECG showed sinus tachycardia, right axis deviation and inferior ischaemia. Examination of arterial blood gases revealed:

- pH 7.29;
- Partial pressure of CO2 9.9 kPa;
- Partial pressure of O2 6.4 kPa;
- Base excess (BE) +6.2;
- and standard bicarbonate 29 mmol/l.

The serum sodium level was 114 mmol/l and the serum potassium level 6.0 mmol/l.

A chest radiograph revealed cardiomegaly with right atrial enlargement, prominence of the pulmonary vasculature, some hyperinflation of the lung fields and no evidence of pneumonic consolidation. Endotracheal intubation relieved his respiratory distress and he was transferred to the respiratory intensive care unit for further management. Therapy included oxygen-enriched humidified air via the endotracheal tube and a T-piece, intravenous and nebulised bronchodilators, physiotherapy to clear secretions, and diuretics. He improved clinically on this therapy with resolution of the clinical signs of right ventricular failure and clearing of secretions.

Indirect laryngoscopy revealed an oedematous supraglottis; the posterior pharyngeal wall was prominent and impinging on the arytenoids. An elective tracheostomy was performed before further evaluation. Subsequent fibre-optic bronchoscopy showed that the vocal cords were also oedematous but otherwise normal and mobile, but the subglottic airway was markedly narrowed and did not allow passage of the 5 mm bronchoscope. Lateral radiographs of the cervical spine (Fig. 1) showed the presence of large osteophytes extending from C1 to C6 and compressing the upper airway.

Lung function testing and flow volume loops performed via the tracheostomy tube and subsequently via the tracheostomy tube