Cystic nephroma with heterotopic bone formation: a pathological and radiological pitfall

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Abstract

This case illustrates heterotopic bone formation in a patient with cystic nephroma (CN) of the kidney. Renal CN may be difficult to discern clinically and radiologically from 'Multilocular Cystic Renal Cell Neoplasm of Low Malignant Potential (MCRCNLMP)' or a clear cell renal cell carcinoma with extensive cystic change. In both of the latter, dystrophic calcification or ossification is not uncommon. The formation of heterotopic bone in CN/MEST is rare and should not be confused with sarcomatous transformation on histology. Radiologically, the presence of central calcification increases the suspicion of a cystic renal cell carcinoma, a potential diagnostic pitfall.

Keywords cystic nephroma; heterotopic bone formation; mixed epithelial and stromal tumour

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Introduction

When considering the radiological findings of a cystic renal neoplasm in which there are areas of calcification, the primary suspected diagnosis is a renal cell carcinoma, either a multilocular cystic clear cell renal cell neoplasm of low malignant potential (MCRCNLMP) or clear cell renal cell carcinoma with extensive cystic change. MCRCNLMP (previously referred to as multilocular cystic renal cell carcinoma) is composed of numerous cysts, the septa of which contain groups of clear cells indistinguishable from grade 1 clear cell renal cell carcinoma. MCRCNLMP is a subtype of renal cell carcinoma with >200 patients described in the literature, of which no cases have recurred or metastasised.[1] Clear cell renal cell carcinoma with extensive cystic change shows expansile nodules of clear cells which alter the septal configuration.

We present a case of cystic nephroma (CN), a tumour which is considered by many to be on a morphological spectrum with mixed epithelial and stromal tumour (MEST). CN and MEST are benign tumours and radiological mimics of renal cell carcinoma. There are rare case reports of aggressive variants of CN/MEST showing sarcomatous transformation. Dystrophic calcification within these tumours is rare, with only a single case reported in the literature.[2] The formation of heterotopic bone with bone marrow has not been previously reported. This finding could potentially be interpreted as sarcomatous transformation histologically or increase the suspicion of renal cell carcinoma radiologically, and this forms the basis of this illustrative case report.

Case report

A 74-year-old lady was investigated for lower limb oedema and underwent abdominal ultrasound, which revealed a large complex left renal cyst. Subsequent contrast-enhanced computer tomography (CT) demonstrated a 7 × 5 × 6 cm multilocular encapsulated cystic mass arising from the interpolar region of the left kidney with heterogeneous septal contrast enhancement and two foci of calcification (Figure 1). Multiple lung nodules were also identified. The radiological findings were highly suggestive of cystic renal cell carcinoma and the patient underwent a laparoscopic radical nephrectomy. The postoperative course was uneventful, with no evidence of recurrence 5 months after surgery.

Macroscopic findings

A radical left nephrectomy was received. Sectioning revealed a 58 mm diameter multilocular cystic tumour. The cysts were filled with clear yellow fluid; they were smooth walled with no papillary excrescences. The tumour was circumscribed, there was no haemorrhage or necrosis and no evidence of extrarenal extension. The tumour was focally solid with a gritty calcified texture.

Microscopic findings

Histopathology revealed a multiloculated cystic lesion. The cysts were lined by an attenuated flattened cuboidal to columnar and in places hobnail epithelium. The intervening stroma formed septae of varying thickness and cellularity. The stroma was

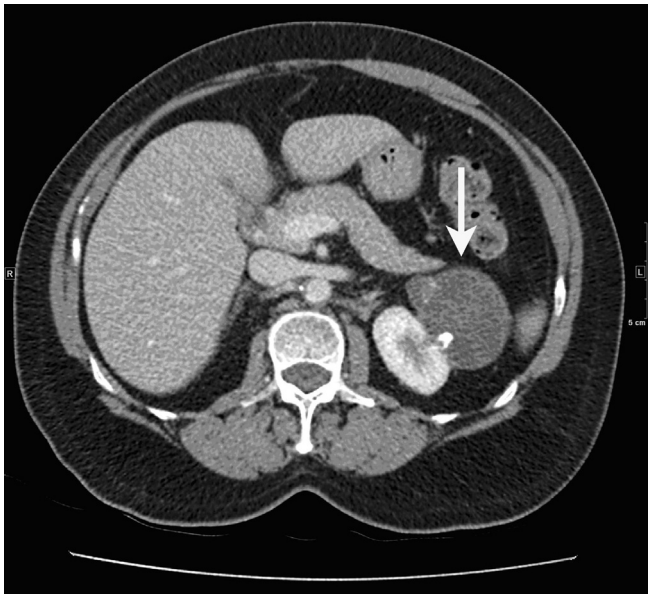

Figure 1 Axial contrast enhanced CT image shows the multiloculated left renal cyst with a dense focus of calcification.

focally hyalinised and in areas showed ovarian-like appearances. Smooth muscle bundles were also identified. A focus of heterotopic bone formation, including marrow spaces, was identified, this focus measuring 9.5 × 7 × 4 mm (Figure 2). There was no evidence of tumour extension into the renal sinus or perinephric fat.

Immunohistochemistry showed the epithelium lining the cystic spaces to be strongly positive for pan-cytokeratin (AE1/AE3). The ovarian-like stroma stained positively for oestrogen receptors, progesterone receptors and CD10. The smooth muscle was highlighted by smooth muscle actin. Within the marrow-like spaces, CD45 showed positivity for lymphoid cells (Figure 2). The morphological findings in conjunction with the immunohistochemical phenotype of the lesion were considered diagnostic of CN with the unusual feature of "heterotopic bone", comprising bone trabeculae with marrow spaces containing haematolymphoid cells.

Discussion

CN is a benign cystic tumour of the kidney, which was originally named "cystic adenoma", in 1892 by Edmunds.[3] The histopathological profile and radiological characteristics of CN have been

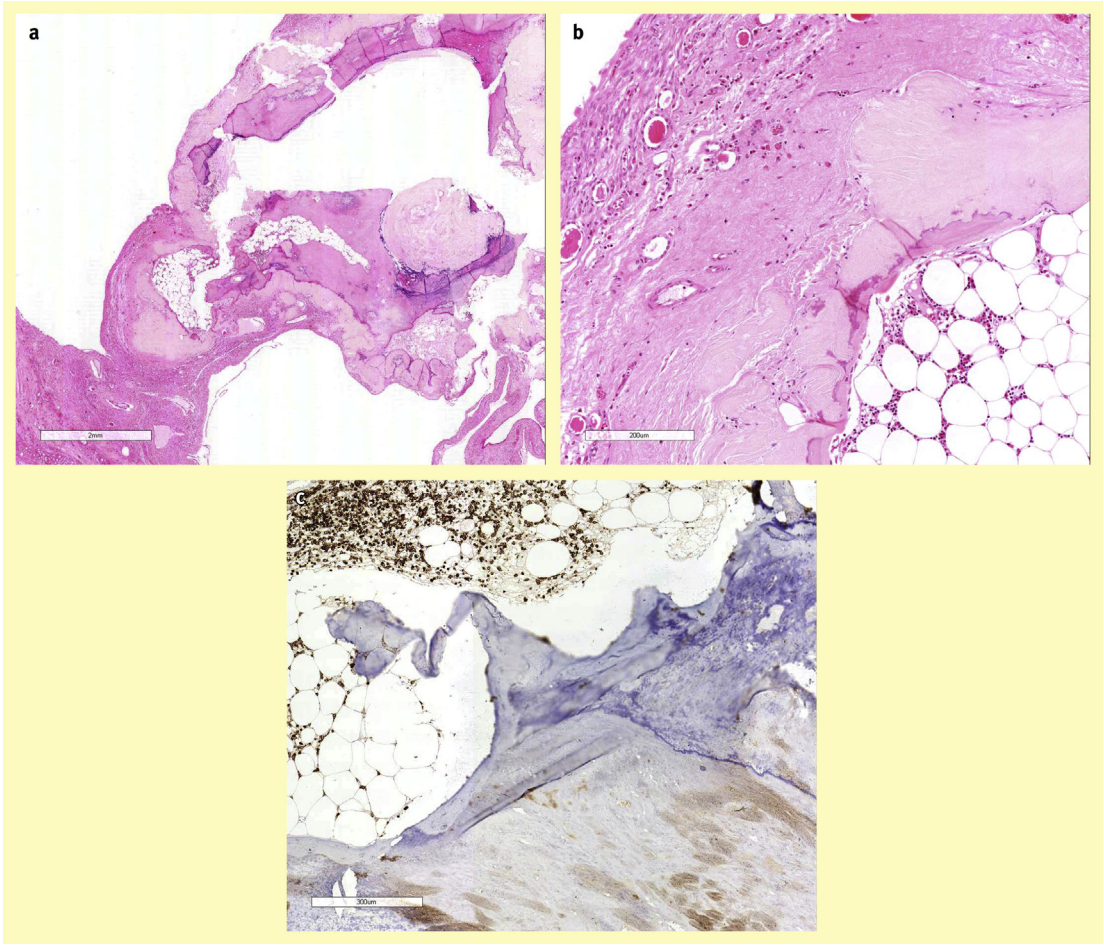

Figure 2 (**a**) Shown at scanning magnification (×2), the area of bone centrally, adjacent to more typical CN, with the cystic spaces top left and bottom right. (**b**) Higher (×10) magnification, cyst wall left edge, underlying stroma, central bone matrix and right marrow space with haematolymphoid cells (**c**) CD45 (LCA) showing strong positivity within the cellular lymphoid component of the marrow space with adjacent bone.