Case Report

A case of renal involvement in juvenile xanthogranulomatosis ♠

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ABSTRACT

Juvenile xanthogranuloma (JXG) is a type of non-Langerhans cell histiocytosis that rarely involves other than the skin. Here, we present detailed ultrasound (US) findings, including a contrast study, of a rare JXD renal lesion. A 42-year-old woman with JXG had chronic kidney disease. Ultrasound showed multiple cystic masses with fine internal septa in both kidneys. Contrast-enhanced US revealed early staining and late washout consistent with the internal septa inside the masses and led us to suspect cystic renal cell carcinomas in both kidneys. Left nephrectomy was performed for diagnostic purposes. Microscopic examination revealed a foamy component with Touton-type giant cells by histiocytosis; CD68 and S100 were positive, and CD1a was negative, leading the diagnosis of JXD. The US findings of extracutaneous lesions on JXA are variable and can be cystic, and when arising in the kidney may resemble cystic renal cell carcinoma.

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Case report

Juvenile xanthogranuloma (JXG) is a type of non-Langerhans cell histiocytosis that typically occurs in infants and children. Lesions are usually asymptomatic, red-yellow papules and nodules on the skin. Although, extracutaneous involvement is uncommon, the most frequent extracutaneous sites are the subcutaneous soft tissue, central nervous system, liver, spleen, lung, eye, oropharynx, and muscle [1]. Here, we present detailed ultrasound (US) findings, including a contrast study, of a rare JXD renal lesion.

A 42-year-old woman was diagnosed with JXG at the age of 18 years due to central enuresis and pituitary JXG involvement. Subsequently, numerous JXG lesions developed on the skin, esophagus, conjunctiva, and hip joints. The patients also had chronic kidney disease (CKD stage 4).
Fig. 1 – (A) Ultrasound image of the left renal lesion shows a well-defined cystic mass protruding outside the kidney and containing fine, thickened internal septa, with posterior echo enhancement. (B) Superb microvascular imaging shows blood flow consistent with fine septa (arrowhead). (C) Contrast-enhanced ultrasound shows early staining (arrowhead) and late washout of the fine septa in the mass (arrow) compared to background renal parenchyma, which is consistent with Bosniak category type 4.

Fig. 2 – (A) Coronal and (B) axial T2-weighted MR imagings show multiple cystic lesions with numerous internal septa in both kidneys (arrows), the lesions protrude into the renal pelvis. Left-dominant bilateral renal atrophy is also seen.

An US using an Aplio i500 system with a 14 MHz linear transducer (Canon Medical Systems, Japan) showed cystic masses of 36 mm, 31 mm in the right kidney, and 16 mm in the left kidney, which protruded into the renal pelvis. The masses contained fine, thickened internal septa (Fig. 1A), and the blood flow was consistent with the septa (Fig. 1B). In addition, multiple simple cysts in both kidneys as well as left-dominant bilateral renal atrophy were observed. Contrast-enhanced US was performed because contrast-enhanced computed tomography and magnetic resonance imaging (MRI) could not be performed due to her CKD. Contrast-enhanced US revealed early staining and late washout consistent with the internal septa inside the masses (Fig. 1C).

MRI showed multiple cysts in both kidneys that were increased compared to the previous study, with multiple fine septa inside them (Fig. 2), and contrast-enhanced US findings led us to suspect cystic renal cell carcinomas in both kidneys. Left nephrectomy was performed for diagnostic purposes after a renogram confirmed that the left kidney was nonfunctional. Macroscopic examination of the left nephrectomy specimen revealed multiple cystic lesions of different sizes and multiple fibrous septa (Fig. 3A). Microscopic examination revealed a foamy component with Touton-type giant cells by histiocytosis; CD68 and S100 were positive, and CD1a was negative, leading us to the diagnosis of JXD (Figs. 3B and C).

Discussion

The few reported extracutaneous JXD lesions found on US have been variable and described as: homogeneous solid, sharply circumscribed, relatively avascular subcutaneous lesion [2], solid and cystic adrenal lesion with indistinct borders and blood flow within the solid area [3], and as a heterogeneous, well-defined exophytic renal lesion with mild blood flow [4]. To the best of our knowledge, this is the first re-
port of extracutaneous JXD lesions that were scrutinized with contrast-enhanced US, and extracutaneous JXD lesions of our case were depicted as multifocal and multicystic lesions with strong enhancing effects, which were difficult to distinguish from cystic renal carcinomas on imaging.

Conclusion

The US findings of extracutaneous lesions on JXA are variable and can be cystic, and when arising in the kidney may resemble cystic renal cell carcinoma.

Patient consent

A written consent was obtained from the patient for publication of this case and any accompanying images.

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