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Xanthogranulomatous Pyelonephritis and the Bear's Paw Sign

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This report is based on medical experiences at the Neuquén Provincial Hospital.

The term “bear paw sign” was first proposed by Parker and Clark in 1989. 1 This sign is characteristic of xanthogranulomatous pyelonephritis and is evident by intravenous contrast-enhanced CT urography. It represents an enlarged kidney with multiple low-attenuation masses scattered throughout the renal parenchyma, representing dilated calyces filled with debris and collections of xanthomas. This, combined with hypo enhancement of the surrounding renal parenchyma, represents a resemblance to a bear paw, where the necrotic areas within the dilated collecting system mimic the toe pads of a bear’s paw (**Figure 1 A - Figure 1B**).

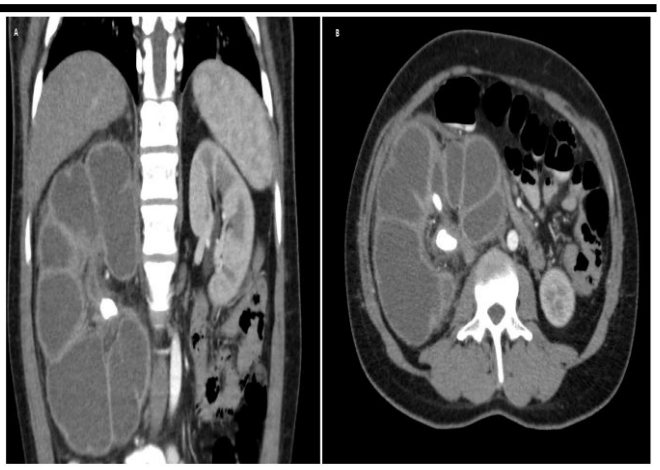

Figure 1: Coronal oblique (A) and axial (B) CT urography in the arterial phase of a 28-year-old male patient with a history of recurrent pyelonephritis. Note the enlargement of the right kidney associated with severe hydronephrosis, with a stone in the renal pelvis. This is accompanied by thinning and hypoenhancement of the renal parenchyma, as well as an increase in the density of perinephric fat. These findings illustrate the bear's paw sign characteristic of xanthogranulomatous pyelonephritis, subsequently confirmed by pathology.

The first case of xanthogranulomatous pyelonephritis was first described by Schlagenhaufer in 1916, although at that time it was called “staphylomycosis” due to its similarities with actinomycosis and the presence of staphylococci on microscopic examination. It was not until 1935 that Oberling used the term "xanthogranulomatous" in reference to the yellowish color of the affected kidney and the granulomatous inflammation visible under a light microscope [1]. In this sense, this disease is the result of obstruction, usually lithiasic, and chronic infection of the affected kidney,

producing a suppurative granulomatous reaction in which the renal parenchyma is replaced by lipid-filled macrophages [2].

There are several theories regarding etiology, including renal lithiasis (which accounts for 80% of cases), urinary tract obstruction, and recurrent urinary tract infections (30% to 40% caused by Escherichia coli and Proteus mirabillis). However, the precise cause is unknown. Since radiological and clinical findings are inconclusive, most cases are diagnosed during surgery [3].

The lesion is almost always limited, but when it extends to the parenchyma, it can be classified into the following stages: phase I, only the parenchyma is affected; phase II, the involvement includes the parenchyma and adjacent adipose tissue; and phase III, the lesion extends to the perinephric adipose tissue and retroperitoneum. When the lesion is affected bilaterally, the prognosis is poor [4].

It represents only 0.6% of all cases of chronic pyelonephritis,4 and its differential diagnosis should be based primarily on fibrolipomatous replacement, a condition in which the kidney undergoes replacement of its parenchyma with fibroadipose tissue, resulting from chronic infections. In the latter case, fibroadipose proliferation originates in the renal sinus, enveloping the kidney and causing its atrophy. This condition may coincide with xanthogranulomatous pyelonephritis, since they share triggering factors such as obstructions and recurrent infections [5].

In conclusion, imaging plays a significant role in assessing the extent and associated complications. Without treatment, xanthogranulomatous pyelonephritis is a potentially fatal disease, but morbidity and mortality are significantly reduced when detected and treated promptly.

1. Author Contribution Statement

I acknowledge that the authors listed below jointly contributed to the conception and design of the study, the analysis and interpretation of the data, and the writing of the manuscript entitled Xanthogranulomatous Pyelonephritis and the Bear's Paw Sign". All authors reviewed the manuscript and approved the final version submitted for publication.

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