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Xanthogranulomatous pyelonephritis in a child presenting with cutaneous fistula

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Background: Xanthogranulomatous pyelonephritis is a chronic inflammatory disease associated with various presentations, but rarely in children. Recognition of a nephrocutaneous fistula in a child may be considered as a sign of renal tuberculosis in developing countries, particularly accompanying a family history.
Case: We report a case of xanthogranulomatous pyelonephritis (XPN) with nephrocutaneous fistula in a child. Considering resistance to antibiotics and the tuberculosis in family history, it was previously misdiagnosed as renal tuberculosis. Nephrectomy was unavoidable. Yellow colored nodules in macroscopic examination mimicked renal cell carcinoma, but histopathologic diagnosis was XPN.
Conclusion: XPN should be kept in mind in cutaneous fistula possibly originating from kidney, especially in immune-compromised patients with renal calculi.
Key words: Xanthogranulomatous pyelonephritis, cutaneous fistula, childhood.

Introduction

Xanthogranulomatous pyelonephritis (XPN) is an uncommon but well-recognized form of chronic inflammatory disease of kidney. It rarely appears in childhood, but a few cases even in neonatal period were reported.[1–9] Patients have variable clinical manifestations. We report an unusual case of XPN presented with nephrocutaneous fistula in a child.

Case

A 6-year-old girl was admitted because of right flank pain and erythematous subcutaneous nodule at the same location, which spontaneously drained a few days before. She had an episode of painless macroscopic hematuria and nocturia four months ago lasting a few days. Her godfather had died of pulmonary tuberculosis the year before. She seemed to be malnourished as she had a short stature and low body weight. Her family had a poor social and economical status.

Laboratory investigation revealed anemia (hemoglobulin 11.2 gr/L), leucocytosis (10.5X103/mikrol), and elevated erythrocyte sedimentation rate. Blood and urine cultures were negative. Ultrasonography revealed atrophy in right kidney parenchyma and ill-margined calcifications in medulla. A fistula tract from lower pole of right kidney to skin was visualized. IVP demonstrated no function in right side, but left kidney and lower urinary tract seemed to be normal.

Despite the intensive antibiotherapy there was no clinical improvement. There were no response to antituberculosis therapy including rifampycin, streptomycin and INH. The patient underwent right nephrectomy and fistula tract excision.

Macroscopic examination revealed total destruction of renal parenchyma and yellow colored

nodular masses around black staghorn calculus in major calyx (Figure 1). In histopathological examination, diffuse lipid-laden histiocytes and lymphocytes forming granulomas were detected (Figure 2). No evidence of tuberculoid granulomas or renal tumor was observed.

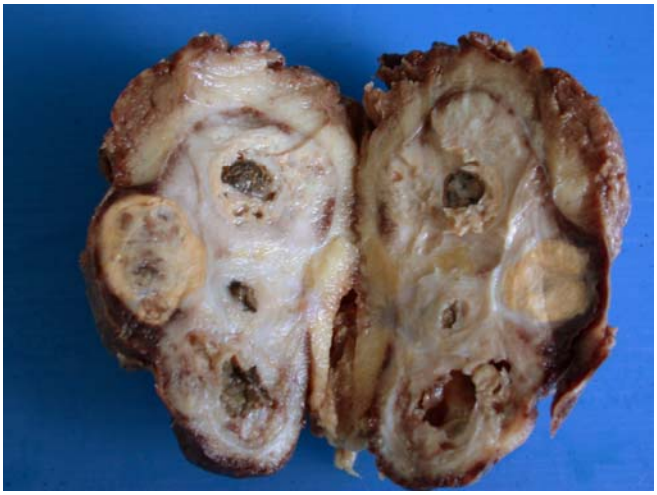

Figure 1. Macroscopic appearance of the bisected kidney. Yellow–tan colored necrotic lesions around staghorn calculi located in major calyx was remarkable. A yellow colored solid nodule protruding into the perinephric tissue was mimicking renal cell carcinoma.

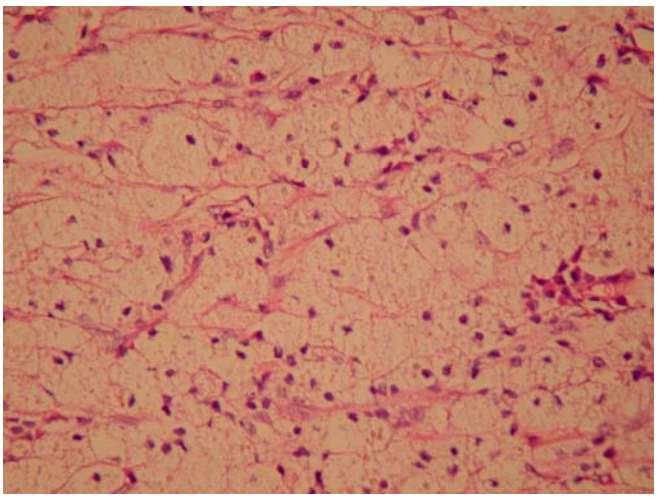

Figure 2. Microscopy revealed diffuse infiltration of lipid-laden histiocytes and scattered lymphocytes forming granulomatous foci (Hematoxyline and eosin x100).

Discussion

The xanthogranulomatous pyelonephritis is an uncommon form of chronic inflammatory disease of the kidney. About 500 cases are described in adults and about 80 cases in children, from the first description in 1963. It rarely appears in childhood, but a few cases even in neonatal period were reported.[9] Urinary tract obstruction, congenital urological anomalies, nephrolithiasis and recurrent urinary infections are mostly accompanying disorders. Its manifestations mimic those of neoplastic disease like Wilms' tumor or renal cell carcinoma.[5,6,10] More commonly, diffuse involvement of kidney leading to dysfunction is seen. Total nephrectomy is unavoidable if there is resistance to antibiotic therapy and non-functional kidney.

Preoperative diagnosis is possible if the disease is considered in differential diagnosis, especially drug resistant and recurrent pyelonephritis in presence of renal calculi or kidney originated fistulas.

Fistula formation is an unusual presenting sign in XPN. Gastrointestinal system, adjacent urinary organs and skin are the most commonly injoined structures. Nephrobronchial fistulas and lung abscess were also reported.[11,12] Cutaneous fistula occurs mostly into ipsilateral flank region, but unusual localizations such as knee were previously reported.[13,14]

HIV infected patients seem to be predisposing to fistula formation. Immunodeficiency or ineffective medical care in pyelonephritis must be the main causes of fistula formation in XPN.[10,11,13] Bingöl-Koloğlu et al. stressed on poor social-economic condition and malnutrition as characteristics of their 17 cases.[1]

Our experience supports that this entity must be considered even in childhood when a cutaneous fistula, possibly originating from kidney is encountered. Malnourishment and staghorn calculi are considered as the main predisposing factors in our case. Long standing urinary infection and hematuria must have been neglected in this social background. Medullary calcifications in preoperative ultrasonography seemed to be clues of staghorn calculi.

Nephron sparing surgery would be appropriate if focal involvement is confirmed by imaging analyses. But preoperative diagnosis is challenging in XPN. In one of the largest XPN series, Bingol-Kologlu et al.

reported that only two of the 17 patients were correctly diagnosed based on CT findings.[1] Preoperative diagnosis was unclear even if different imaging methods were used.[15]

Nephrectomy and primary excision of fistula is curative in diffuse renal destruction.[10–14] Percutaneous drainage of abscess and preoperative antibiotic therapy are recommended for uncomplicated surgical interventions. Long-term antibiotherapy had not resolve the discharge via the fistula, but facilitated the nephrectomy in this reported case.

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