PYELONEPHRITIS XANTHOGRANULOMATOUS IN CHILDHOOD: CASE REPORT AND LITTERATURE REVIEW

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PYELONEPHRITE XANTHOGRANULOMATEUSE CHEZ L'ENFANT: A PROPOS D'UN CAS

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RÉSUMÉ

Pre-requis : La pyélonéphrite xanthogranulomateuse est une forme particulière d'infection rénale chronique rarement rencontrée chez l'enfant.

But : Illustrer un cas pédiatrique de pyélonéphrite xanthogranulomateuse en insistant sur l'apport de l'imagerie et sur les différents diagnostics différentiels qui peuvent se poser.

Observation : Nous rapportons le cas d'une pyélonéphrite xanthogranulomateuse polaire inférieure du rein droit chez un petit garçon âgé de 2 ans en insistant sur la difficulté diagnostique.

Conclusion : La pyélonéphrite xanthogranulomateuse est une situation pathologique rare chez l'enfant qu'il faut évoquer devant certaines masses rénales de l'enfant.

MOTS-CLÉS

Pyélonéphrite xanthogranulomateuse, enfant, infection

SUMMARY

Background: Xanthogranulomatous pyelonephritis (XGP) is a specific form of chronic inflammatory kidney disease rarely seen in children. The Symptoms are often vague and non-specific

Aim: the aim of this paper is to return the particularities of imaging features in xanthogranulomatous pyelonephritis, insisting on differential diagnosis with renal tumors, especially in case of no renal stone or tract obstruction evidence.

Case: We report a case of xanthogranulomatous pyelonephritis in a 2-year-old boy involving the lower renal pole which demonstrates the diagnostic difficulties encountered in this disease

Conclusion: Xanthogranulomatous pyelonephritis is a rare condition in children and should be included in the differential diagnosis of a child presenting a renal mass.

KEY-WORDS

Pyelonephritis, xanthogranulomatosis, children, infection

آلتهاب الكلية الحويضة الحبيبومي عند الطفل

الباحثون : دويرة . و - لواتي . هـ - جراية . هـ - بن حسين . ل - مورماش . ي - ساحلي . س - حمزاوي . م - بالأغة . ا.

آلتهاب الكلية و الحويضة الحبيبومي هو نوع من تعفن الكلية المزمن النادر عند الطفل.

تشتمل دراستنا على حالة طفل مصاب بهذا المرض عمره سنتين و نؤكد من خلال هذه الحالة على صعوبة التشخيص و على دور التصوير . نستخلص أنه يتحتم علينا أن نفكر في هذا المرض كلما تواجدت كتلة في الكلية عند الطفل.

الكلمات الأساسية : آلتهاب الكلية و الحويضة الحبيبومي - طفل - خمج.

Xanthogranulomatous pyelonephritis (XGP) is a specific form of chronic inflammatory kidney disease rarely seen in children. The disease presents either in the diffuse form or less commonly as a focal process which is almost impossible to differentiate from renal malignancy. XGP is usually associated with urinary tract obstruction, infection and/or renal stones. Symptoms are often vague and non-specific. In reviewing the literature we have found only 265 cases in children since 1960 (1). We report a case of focal xanthogranulomatous pyelonephritis in a 2-year-old boy involving the lower renal pole which demonstrates the diagnostic difficulties encountered in this disease. We also review the published reports of xanthogranulomatous pyelonephritis in pediatric patients.

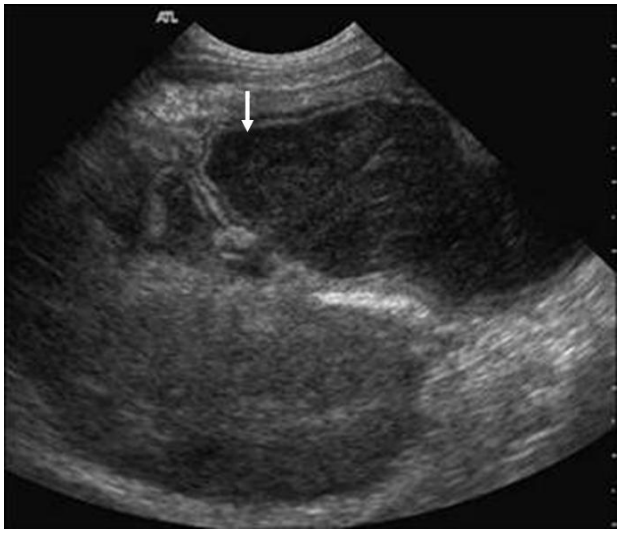
CASE REPORT

A two-year-old male patient presented with fever and abdominal pain of fifteen days duration. There was no history of trauma or weight loss. He was a well developed, normal appearing child in mild distress.

Abdominal examination revealed a soft and lax abdomen with a tender mass having ill-defined margins in the right lumbar region. The rest of physical examination results were unremarkable.

Results of laboratory studies demonstrated a normal haemoglobin level (127 g/l) and a high count of white blood cells (12800e/mm3). Urinalysis did not reveal any white or red blood cells or bacteria. A renal ultrasound scan revealed an exophitic heterogeneous mass with large cystic areas in the lower right renal pole replacing normal cortico medullary differenciation. It was largely extended to the perinephritic area and adherent to the ascending colon and its meso (figure1).

Figure 1 : US renal scan: an exophitic heterogeneous mass with large cystic areas ( ) in the lower right renal pole replacing normal cortico medullary differenciation



A computed tomography (CT) scan (figures 2 (a, b, c) was obtained demonstrating normal functioning kidneys and the right renal mass described below. This process was almost cystic, with ill-defined borders, inhomogenous density and irregular enhancement concerning the tissular component and the septa. It had a large perinephritic extension with an important thickening of Gerota's fascia and infiltration of the lateral wall of the abdomen, the mesenteric fat and the psoas muscle. There was no evidence of hydronephrosis or calculi. The inferior calyceal cavities were displaced by the process with no evidence of pyeloureteral junction syndrome or obstruction. Due to the renal contour bulging around the exophytic mass and the adjascent adenopathy, a neoplastic process such as a Wilms' tumor could not be ruled out preoperatively. A surgical exploration was then performed.

Figure 2 : (a) Precontrast CT scan shows an iso- or slightly hypodense exophytic mass ( ) with irregular borders and an inhomogenous density of the lower pole of the right kidney

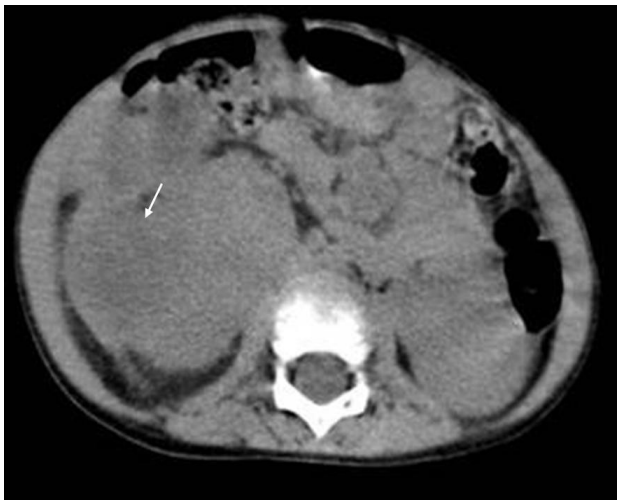
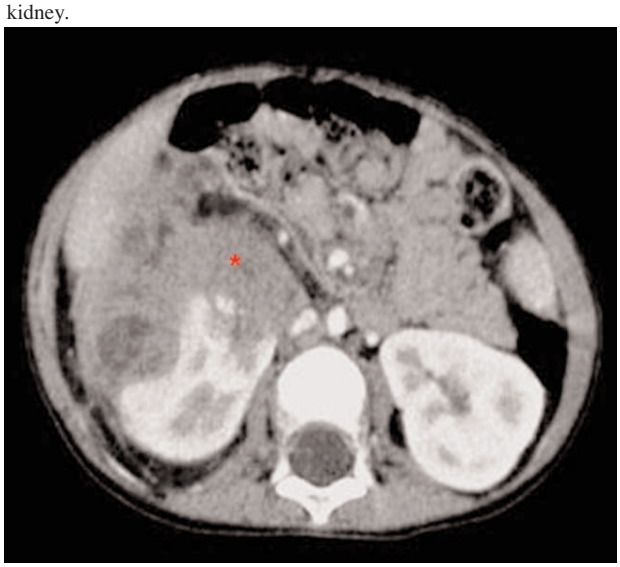


Figure 2 : (b,c) Postcontrast CT scan reveals irregular enhancement of the exophytic mass (*) with some internal nonenhancing foci and the nonenhancing hypodense lesion (*) in the medial portion of the left kidney.



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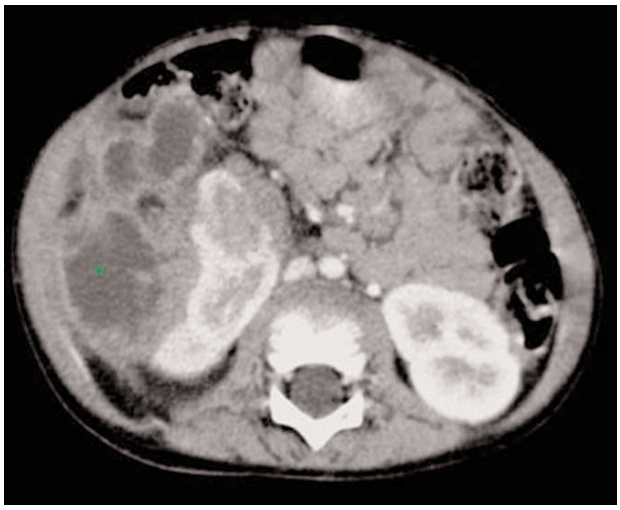
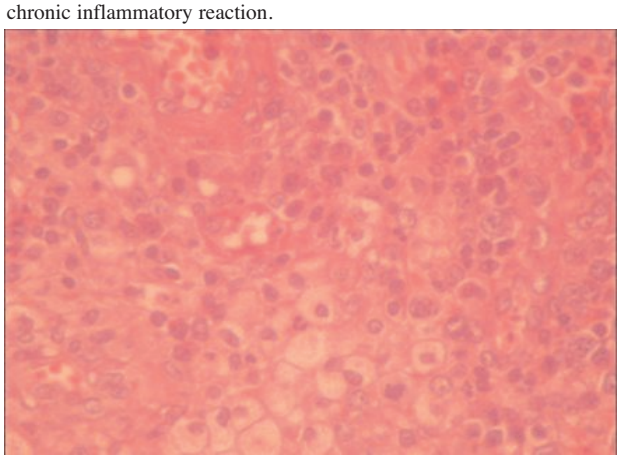


At surgery, the intraoperative findings were well correlated with the imaging data. In fact, there was a renal abcess adherent to the surrounding tissues: it was adherent to the ascending colon and its meso, the psoas muscle, the lateral abdominal wall and the posterior peritoneum. The disease process was confined to the lower pole of the kidney. A intraoperative pathological exam did not find neoplastic cells and a limited resection was then performed to remove the abcess. The bacteriological examination revealed an Escherichia coli. The definitive histological examination (figure 3) revealed mixed acute and chronic inflammatory cell infiltrate with giant cells and lipid-laden macrophages (foam cells) concluding to a XGP. The child had an uncomplicated postoperative recovery and was treated with parenteral antibiotics with remarkable outcome.

Figure 3 : Microscopic examination reveals foamy cells mixed with polymorphonuclear neutrophil cells demonstrating the acute inflammatory reaction and plasmocyte with lymphocyte cells for the chronic inflammatory reaction.



DISCUSSION

XGP is an unusual form of pyelonephritis rarely occurring in children with only 265 cases reported in the literature since 1960 (1). Several interrelated aetiological features are thought to be responsible for the pathogenesis of xanthogranulomatous pyelonephritis. They include calculus or non-calculus urinary obstruction, ineffectively treated urosepsis, chronic renal ischaemia causing localised alterations in renal metabolism, lymphatic obstruction, deterioration of the lipid metabolism, and finally an impaired immune response. The most common offending organisms are E. coli and Proteus mirabilis (3, 4).

It may present as either a segmental or a diffuse chronic inflammatory infectious process characterized by a high proportion of foam cells. In the rare localised form, the lesion can be confined to one pole, as was seen in our patient. More commonly, a diffuse process is seen involving the whole kidney, leading in most cases to a decrease in renal function. Xanthogranulomatous pyelonephritis is further divided into 3 stages: stage I (nephric XGP), corresponds to inflammation confined to the kidney; stage II (perinephric XGP), the inflammation involves both the kidney and perirenal fat; stage III (paranephric XGP), the inflammation involves the kidney, perirenal fat, and the retroperitoneum (5).

On laboratory examination, anemia is a common finding. Leukocytosis is present in 50–75% of cases of XGP. An elevation in alkaline phosphatase and liver enzymes is seen in less than half of patients. Urinalysis is significant for pyuria in 90% of cases Urine cultures are positive in 50–80% of patients, with Gram-negative organisms (P. mirabilis and E. coli) being the most common pathogens (6). One plausible explanation for the higher than expected percentage of sterile urine cultures is that obstruction prevents contaminated urine from reaching the bladder. In our case there was no evidence for calyceal contamination (7).

The most commonly reported symptoms are fever, abdominal and/or flank pain, weight loss, malaise, anorexia, and lower UTI symptoms (3). Pyuria is frequently seen. However, the diagnosis must not be discounted if pyuria is not present.

The most common physical examination findings include a palpable mass and flank tenderness. Hypertension can also be seen. No single radiologic feature is diagnostic (8). Voiding cystourethrography is required not only for evaluation of congenital anomalies associated with the involved kidney, but also for the possible concurrent congenital anomalies of the opposite kidney (3). Ultrasound can show enlargement of the entire kidney with multiple hypoechoic areas representing hydronephrosis and/or calyceal dilatation with parenchymal destruction, as well as calculi .A focal mass can be less commonly revealed, as it was in our case.

Computed tomography scan is usually indicated because conventional radiographs and/or ultrasound can yield ambiguous results (10). Typical CT findings include global renal enlargement with multiple low attenuation rounded areas completely replacing renal parenchyma. This is consistent with dilated calyces or focal areas of destruction filled by pus or debris (“bear paw sign”) (9, 11). Extra-renal involvement

(perinephric, psoas, bowel, diaphragm, posterior abdominal wall) is common (9,11). A kidney affected by XGP can be mistaken for a malignant process, such as a Wilms' tumor. The differential diagnosis of XGP includes Wilms' tumor and other neoplastic disorders (neuroblastoma, leukemia, lymphoma, clear cell carcinoma, and retroperitoneal sarcoma) and inflammatory processes (renal or perirenal abscess, pyonephrosis, renal tuberculosis, focal or diffuse nephritis, and fungal infection). Ultrasound and CT features not consistent with Wilms' tumor include the absence of sharp definition and encapsulation of the mass or ill-defined margins with inflammatory infiltration of the perinephric fat (11).

However, these characteristics might not all be present as it was the case of our children. The confirmation of this diagnosis always depends on histological examination. Microscopic findings include a mixed acute and chronic inflammatory cell infiltrate with giant cells and lipid-laden macrophages (foam cells); interstitial fibrosis is also present along with variable degrees of tubular atrophy and/or dilation can also be seen. Misinterpretation of “foamy cells” as “clear cells” consistent with renal adenocarcinoma is the most important diagnostic challenge histologically. No correlation between the histologic features of XGP and the extent of clinical course of the disease has been established (2, 11). Treatment of this disease is a surgical excision of the diseased tissue with or without antibiotic therapy. In most cases this will mean a total nephrectomy (8, 11). However, in the relatively rare cases where the localised form of the disease is found, which involves only a single pole of the kidney, it might be possible to carry out a partial nephrectomy. When performing surgery, it is very useful if the correct preoperative diagnosis has been made because surgery in this condition can be extremely difficult as a result of the inflammatory process extending beyond the boundaries of the kidney (9,10).

Drainage of any perirenal and/or renal abscess with adjunctive antibiotic treatment is strongly recommended before definitive surgery to decrease the complications in the diffuse form of the disease.There are rare reports of successful treatment of focal XGP with antibiotics (1).

CONCLUSION

Xanthogranulomatous pyelonephritis is a rare condition in children and should be included in the differential diagnosis of a child presenting with fevers, malaise, weight loss, flank tenderness, or a renal mass. Many reports on this disease conclude that preoperative diagnosis is not easy and this viewpoint seems to have been generally accepted, given the vague manifestations of the disease. However, this is a diagnosis that can and should be made preoperatively because of the increasing awareness of the condition. The characteristics of the condition that might help in a preoperative diagnosis of xanthogranulomatous pyelonephritis include: disease is usually unilateral (although very rare bilateral cases have been reported); renal function is absent or grossly impaired on the involved side; large, often numerous, renal calculi are present; anaemia, raised ESR, and leucocytosis are often present. Once the diagnosis is made, the prognosis is excellent after appropriate surgical resection. Early consultation by urology will help guide management. In most cases, nephrectomy is required.

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