Xanthogranulomatous Pyelonephritis in Childhood : Diagnosis Difficulties and Success of Conservative Treatment

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La pyélonéphrite xanthogranulomateuse chez l'enfant : Difficultés diagnostiques et succès du traitement conservateur

LA TUNISIE MEDICALE - 2010 ; Vol 88 (n°06) : 427 - 429

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RÉSUMÉ

Prérequis : La pyélonéphrite xanthogranulomateuse est une forme rare de pyélonéphrite chronique chez l'enfant. Son diagnostic préopératoire est difficile et sa prise en charge thérapeutique reste non codifiée.

But : Les auteurs rapportent une forme focale pseudo-tumorale et discutent ses modalités thérapeutiques.

Observation : Garçon âgé de 2 ans admis pour une volumineuse masse douloureuse et fébrile du flanc droit évoluant depuis 15 jours. Le bilan biologique a montré une hyperleucocytose à 12.800 par mm3 et une protéine C réactive élevée à 50 milligrammes par litre. L'examen cyto-bactériologique des urines était négatif. L'échographie et la tomodensitométrie ont montré une masse multiloculaire, du pôle inférieur du rein du rein droit, mesurant 80 millimètres de diamètre, prenant le contraste en périphérie avec épaississement péri rénal. Le rein controlatéral était normal. L'origine tumorale ou inflammatoire n'a pu être précisée, motivant l'exploration chirurgicale. Par une para-rectale, découverte d'un volumineux abcès du pôle inférieur du rein droit, contractant des adhérences intimes avec les structures avoisinantes. Une biopsie avec mise à plat de la collection, toilette et drainage ont été réalisés. L'étude histologique a posé le diagnostic de pyélonéphrite xanthogranulomateuse. Le prélèvement per-opératoire était positif à Escherichia coli. Une antibiothérapie adaptée a permis la régression complète et la conservation du rein. Le recul est de 2 ans sans récidive.

Conclusion : La pyélonéphrite xanthogranulomateuse doit être discutée devant toute masse lombaire fébrile. Le traitement conservateur peut amener la guérison et doit être recommandé dans les formes focales de l'enfant.

Mots-clés

Masse rénal, pyélonéphrite xanthogranulomateuse, enfant

SUMMARY

Background : Xanthogranulomatous pyelonephritis is an uncommon form of chronic pyelonephritis rarely seen in children. Preoperative diagnosis is difficult and management is still obscure.

Aim : The authors report on a focal pseudotumoral case and discuss therapeutic modalities.

Case : A 2-year-old boy was admitted for a 2-weeks history of fever and right flank pain. Physical examination revealed a right lumbar mass. White blood cell count was 12.800/mm3 and C reactive protein elevated (50 mg/l). The urine culture was negative. Ultrasonography and computerized tomography demonstrated a focal multilocular cystic mass of the lower polar of right kidney, measuring 80 millimeters, with peripheral enhancement and perirenal involvement. At surgical exploration, a huge abscess of the lower polar of the kidney with dense adhesions to adjacent structures were noted. Excision of the collection and local drainage were performed. Pathologic examination of biopsy specimen confirmed the diagnosis of xanthogranulomatous pyelonephritis. Intraoperative culture was positive for Escherichia coli. Successful treatment of the lesion was achieved with adjunctive antibiotic therapy. The postoperative course was uneventful over a follow-up period of 2 years.

Conclusion : Xanthogranulomatous pyelonephritis should be considered in the differential diagnosis of a renal mass. A first-line conservative treatment must be strongly recommended in pediatric focal cases.

Key-words

Kidney, renal mass, xanthogranulomatous pyelonephritis, child

Xanthogranulomatous pyelonephritis is an uncommon form of chronic pyelonephritis rarely seen in children. Preoperative diagnosis is difficult and management is still obscure. The authors reported on a focal pseudotumoral case and discuss therapeutic modalities.

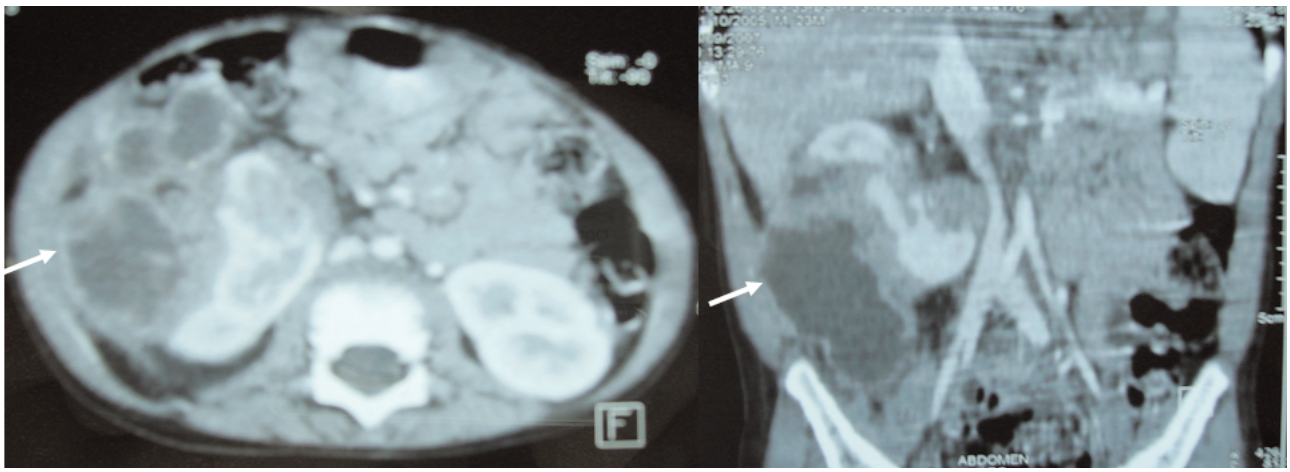
CASE

A 2-year-old boy was admitted for a 2-week history of fever and right flank pain. There is no previous history of urinary symptoms or weight loss. Physical examination revealed a voluminous right palpable lumbar mass. White blood cell count was 12.800/mm3 and C reactive protein was elevated (50 mg/l). Hemoglobin was normal (12.7 g/dl). The urine culture was negative. Ultrasonography showed an anechoic lesion in the lower pole of right kidney, measuring 80 millimeters. There is no urolithiasis or calcifications. Computerized tomography demonstrated a focal multilocular cystic mass with peripheral enhancement and perirenal involvement (Figure 1). The controlateral kidney was normal. The differential diagnosis included neoplastic process, such as Wilm's tumor, or inflammatory disease, such as renal abscess and xanthogranulomatous pyelonephritis. The patient underwent surgical exploration through a para-rectal approach. During surgery a huge abscess of the lower pole of the right kidney with dense adhesions to adjacent structures and purulent cystic fluid were noted. Excision of the collection with kidney preservation and local drainage were performed. Pathologic examination of biopsy specimen confirmed the diagnosis of xanthogranulomatous pyelonephritis. Intraoperative culture was positive for Escherichia coli. Successful treatment of the lesion was achieved with adjunctive antibiotic therapy for 2 weeks. The postoperative course was uneventful and the patient was asymptomatic over a follow-up period of 18 months.

DISCUSSION

Xanthogranulomatous pyelonephritis (XGP) is an uncommon form of chronic pyelonephritis rarely seen in children. It is characterized by destruction of renal parenchyma with replacement by granulomatous tissue containing lipid-laden macrophages [1]. Its etiopathogenesis is poorly understood but is presumed to be related to urinary tract obstruction, renal calculi, and chronic infections commonly due to E. coli and Proteus. Other etiologic factors include alterations in lipid metabolism, altered immune response and disturbances of leukocyte function, malnutrition, lymphatic obstruction, venous occlusion, arterial insufficiency and necrosis of pericalyceal fat [1-3]. Preoperative diagnosis is difficult because of its non-specific presentation. It can be suspected on a constellation of clinical and laboratory findings combined with imaging features [1-3]. XGP is classified as diffuse (92%) or focal (8%) and divided into 3 stages depending on the extension of inflammation: stage I (nephric XGP), stage II (perinephric XGP), stage III (paranephric XGP), as observed in our case [2]. In the focal form, ultrasonography shows a localized hypoechoic mass, often misdiagnosed as renal tumor, like in our case [1, 3, 4]. Usually indicated, CT scan demonstrates global renal parenchyma replacement by multiple low attenuation rounded areas with a strongly enhanced rim and demonstrates the extent of perirenal involvement [1-3]. There are few data about magnetic resonance imaging (MRI) findings in the focal form. The lesion has slightly low signal intensity on T2 and is isointense with the renal parenchyma on T1. MRI and especially the fast T2 sequences seem to be very useful with the absence of hyperintensity in the differentiation of XGP from tumoral masses [4]. Imaging features not consistent with Wilms' tumor include the absence of sharp definition and

Figure 1 : CT scan demonstrated a focal multilocular cystic mass with perirenal involvement



encapsulation of the mass or ill-defined margins with inflammatory infiltration of the perinephric fat. Intrarenal calcifications are rare in Wilms' tumor [2]. For our patient, the correct diagnosis was made after surgical exploration. Management of paediatric XGP is still obscure due to the limited number of cases. In the localized form, segmental resection of the affected kidney has been reported to be effective [1-3]. Recently, some authors report cases of successful treatment, with debridement and antibiotics [1-5]. Our therapeutic attitude permitted a good outcome and supports conservative management. Once the diagnosis is made, the prognosis is excellent after appropriate treatment.

CONCLUSIONS

Xanthogranulomatous pyelonephritis should be considered in the differential diagnosis of a renal mass. Magnetic resonance imaging seems to be the most valuable radiological investigation and percutaneous biopsy may be needed in selected cases. A first-line conservative treatment must be strongly recommended, in paediatric focal cases.

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