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Research Article

Xanthogranulomatous Pyelonephritis in the Paediatric Population: Ten Years of Experience in a Tertiary Centre

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Abstract

Introduction: Xanthogranulomatous Pyelonephritis (XGP) is a rare infective disease involving functional failure of the affected kidney. Although its certainty diagnosis is anatomopathological, it can be suspected by imaging. Ultrasound (US) and Magnetic Resonance Imaging (MRI) have been described as diagnostic methods. Nephrectomy persists as the treatment of choice in these cases.

Aim: To describe our series of patients with PXG, the imaging techniques used in the preoperative period and the intervention performed.

Methods: Retrospective study of patients under age of 14 from 2010-2020 who underwent nephrectomy due to suspected diagnosis of XGP by imaging. Demographic data, clinical variables, imaging explorations used, surgical technique and histological findings are collected.

Results: Five patients were analysed with a median age at surgery of 3.3 (1.26 – 6.95) years. Proteus spp was identified in lithiasis culture. Lithiasis was staghorn in 50% of them. In two patients the preoperative diagnosis test was MRI alone, and in two it was Computed Tomography (CT). In one patient, US was the only test performed before surgery. In all patients, renal function failure was observed. Five nephrectomies were performed, one of them laparoscopically. A pathological diagnosis of pseudotuberculosis pyelonephritis was identified in one child.

Conclusion: XGP is a rare disease. MRI and US are useful in its diagnosis with no need of CT. Nephrectomy persists as the treatment of choice in these cases.

Keywords: Paediatrics; Diagnosis; Pyelonephritis; Lithiasis

Introduction

Xanthogranulomatous Pyelonephritis (XGP) is a rare chronic inflammatory process leading to renal failure [1,2]. The most frequent symptomatology is flank pain associated with fever. In a high percentage of cases, the underlying condition is caused by an obstruction of urinary tract and its infection [2,3]. The presence of staghorn lithiasis has been described in 42-100% of cases, depending on the published series [3]. The most frequent bacteria associated with these stones is Proteus spp followed by Escherichia Coli [4,5]. In the literature, three stages are described depending on extension: focal, segmental and diffuse [5,6]. Its certain diagnose is anatomopathological with extensive infiltration of granulomatous tissue with lipid-filled macrophages [1]. Despite this, the diagnosis can be suspected by imaging test. In adults, Computerized Tomography (CT) is the gold standard [7]. Currently, Ultrasound (US) is proposed, together with Magnetic Resonance Imaging (MRI), as the preferred method for preoperative diagnosis of suspicion, especially in paediatric population, due to its lower irradiation dose compared to CT scan [8]. Additionally, gadolinium enhanced MRI has a better resolution that CT [8]. The initial management of XGP involves long-term antibiotic therapy, usually with no complete resolution of the condition. The comorbidity of the disease increases with the progressive renal failure, perirenal abscesses and the associated appearance of transitional cell carcinoma [3]. Nephrectomy persists as the treatment of choice, mainly in diffuse forms [1-4]. Both open and laparoscopic approaches are described in the literature, as well as partial and total nephrectomy [2-4]. The aim of this study is to describe our series of patients with XGP as well as the preoperative imaging techniques performed.

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Methods

Retrospective study of patients under age of 15 with suspected XGP by imaging who underwent nephrectomy from 2010 to 2020. Demographic data, clinical variables and imaging test, surgical techniques performed and results of pathology are collected. Quantitative variables were defined by mean and standard deviation or median and interquartile range. Qualitative variables were defined according to their percentage and absolute frequency.

Results

Five patients were analysed (Table 1). The median age at surgery was 3.3 (1.26 – 6.95) years. In all cases, urine culture was positive for non-resistant *Proteus* spp. All of them started with febrile urinary tract infection so as US and plain abdominal X-ray were performed, identifying lithiasis (localization: pelvic 80%, distal ureter 20%). Study was completed with CT in two patients. MRI was performed in three children (Figure 1). A renal scintigraphy (DMSA) was requested in all patients, with renal function <20 mL / Kg/h in 80% of the sample. Total nephrectomy was indicated in all patients. In four patients, open

Table 1: Clinical and demographical data of patients.

Patient	Gender	Age (years old)	Renal unit affected	Nº of lithiasis	Localization of lithiasis	CT	MRI	Surgical approach	Intraoperative time (min)	Complication
1	Male	3,37	Right	>2	pelvis	Yes	Yes	Lumbothomy	140	Blood transfusion
2	Male	5,68	Left	1	pelvis	Yes	No	Chevron	159	Omental eventration
3	Male	6,95	Left	>2	ureter	No	No	Lumbothomy	151	-
4	Male	3,35	Right	1	pelvis	No	Yes	Laparoscopy	264	-
5	Male	1,26	Left	>2	pelvis	No	Yes	Lumbothomy	119	-

approach was preferred, with conventional lumbotomy being the most frequently performed incision. A laparoscopic total nephrectomy was indicated in only one patient, completing the surgery with no need of conversion. In two patients there was a perioperative complication: one of them needed a blood transfusion due to bleeding, in the other patient we observed an omental evisceration on the surgical wound when the drainage was removed. It required reduction and closure of the defect. All surgical specimens were studied microscopically. The pathological diagnosis of XGP was confirmed in 4 specimens. In the remaining patient, a diagnosis of pseudotuberculous pyelonephritis was obtained.

Discussion

XGP is a rare chronic inflammatory disease that is associated with high morbidity in paediatric patients [1,5-12,13]. Lithiasis, usually staghorn type, prevents proper urinary drainage leading to a urine stasis favouring infection [1,5,8]. In the literature the most associated bacteria are *Proteus* spp and *E. Coli* [13,14]. In our series, all urine culture was positive to non-resistant *Proteus* spp. Although the certain diagnosis is pathological, the clinical situation and the results of imaging test lets clinicians to have a suspicion diagnosis [8] and it permits to know the evolutionary stage of the disease, differentiating it into three stages: nephric, perinephric and paranephric [9,10]. It has also been described the classification in focal or diffuse, which can be helpful in surgical decision, its approach and kind of resection [8]. In our centre we prefer the latter classification as it correlates really well with image findings. Initial test performed when XGP is suspected are US and abdominal x-ray, showing the presence of lithiasis in up to 83% of cases [8] and more than half being staghorn stones [8]. In our series, lithiasis were identified in all patients, being the responsible of urine stasis and infection. In the focal stage, US demonstrates a localized, hypoechoic mass, without observing an obstruction of the calyx due to lithiasis. On CT, a well-defined intrarenal mass is demonstrated with ring enhancement that is impossible to differentiate from a renal abscess or a neoplasm. On MRI, instead, an isointense lesion to the renal parenchyma is observed in T1, with a lower intensity in T2, suggesting that we are dealing with a fluid with extremely high protein content, allowing a cleared differentiation between XGP and tumour mass [8]. In the diffuse form, otherwise, US reveals an enlarged kidney, with a multiple caliceal dilations and parenchymal destruction, which can be accompanied by perirenal fluid and infiltration of fat with invasion and extension beyond the renal cell. CT has been the gold standard to identify this stage, especially in adult patients, with an diagnose accuracy up to 87%, as described in the series of Eastham et al. [11]. CT shows a thickening of the entire kidney and low attenuation round areas replacing the renal parenchyma, which are correlated with dilated calyces or areas filled with purulent material, appearing a characteristic sign called 'bear's claw' [14,15] (Figure 2). MRI also plays an important role in the diffuse form, mainly due to the use of contrast (gadolinium-DTPA) allowing an enhancement of the edges of the cavities and outlining the extent of the perirenal inflammation. This sign is important for surgical planning in order to decide the kind of approach used [8]. In our centre, the use of MRI together with US has recently been used in the diagnosis of XGP, as shown in the last two patients of the series. This allowed us an accurate diagnostic suspicion, supporting the surgical indication. We strongly believe that MRI may allow better diagnosis, with a better differentiating the focal form of the renal abscess or Wilms tumour, as well as it is more precisely delimiting the extension of the inflammation

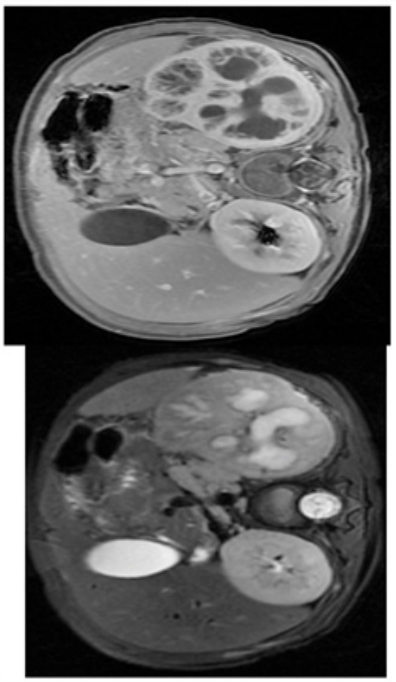

Figure 1: Left xanthogranulomatous pyelonephritis findings on MRI (Patient nº 4). It shows a diffuse increase in size of left kidney, with thinning of the parenchyma and multiple cystic images corresponding to dilated calyxes and focal areas of parenchymal destruction that are heterogeneous in T2 sequence and low signal in T1.

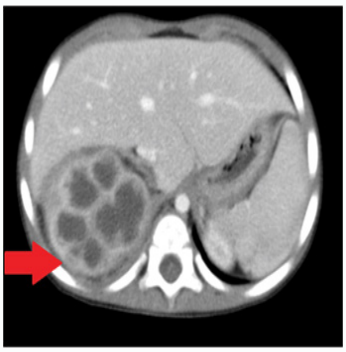

Figure 2: Right xanthogranulomatous pyelonephritis findings on CT scan (Patient nº 1). Bear paw sign (arrow).

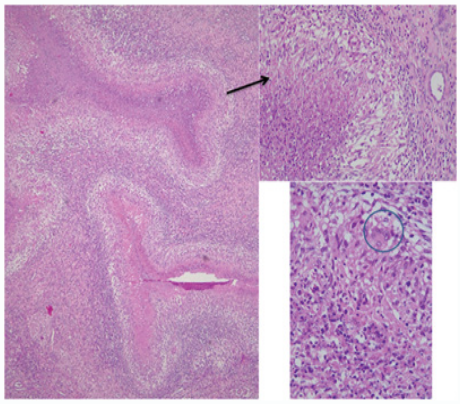

Figure 3: Pseudotuberculous pyelonephritis. Left: Chronic granulomatous uretero-pyelonephritis with necrotizing granulomas and foci of caseification and suppuration. Right (arrow showing a higher magnification): detailed wall of necrotizing granulomas. There are no xanthoidal cells. Circle: Multinucleated giant cell of Langhans.

in the diffuse form, with the added benefit of no radiation, especially in a growing population as childhood [8]. In only one patient the renal scintigraphy did not show a total functional cancellation of the affected kidney. The nephrectomy in this patient was assessed together with the entire urological team. The parents were offered this option after several admissions for pyelonephritis with long period of intravenous antibiotics, the quantification of a decrease in glomerular filtration compared to previous controls and the suspicion of XGP by imaging. Microscope showed a chronic inflammatory process of the renal parenchyma. In four samples, a mixture of acute and chronic inflammation with giant cell infiltrates and lipid-filled macrophages were observed, consistent with the cases described in the literature [8,16-18]. In one patient, microscopic analysis showed an image compatible with pseudotuberculous pyelonephritis. This entity is a pathological diagnosis in which a granulomatous inflammation and mixed cellularity, similar to that found in tuberculous pyelonephritis, are observed. *Mycobacterium tuberculosis* were not identified with Ziehl-Neelsen stain [16,17] (Figure 3). Clinically, it is indistinguishable from tuberculous renal involvement. In this patient, a Mantoux test was performed, with a negative result. In literature, nephrectomy is the preferred choice [2,4,8,11,12,19]. It may be partial or total depending on the presentation of the XGP [2,4]. In our series, we preferred the total resection due to the lack of renal function and the lower possibility of complications. There is controversy regarding the kind of surgical approach [2]. Laparoscopy has been described in these patients [4,17] but due to the large renal and perirenal inflammatory component, the dissection can be quite tedious, increasing both the surgical time and the risk of intraoperative complications [19]. In fact, open surgery is often very complex [19]. An individualised pre surgical study is required in order to identify the ideal candidates to undergo the minimally invasive approach. In our series, we performed one laparoscopic nephrectomy due to the less marked perirenal extension showed in the MRI in that patient, with low probability of finding a complicated dissection during surgery. Despite the fact that comorbidities such as perirenal abscess or nephrocolonic fistula have been described in the literature, in our series the perioperative complications were grade II-III in the Clavien-Dindo classification, with low morbidity [20,21].

Conclusion

Xanthogranulomatous pyelonephritis is a rare renal entity whose diagnose can be suspected by imaging tests. Abdominal US remains the first study in these patients. Recently, MRI has shown great precision in the diagnosis of suspicion. Possible in the future this test will displace CT as the technique of choice. Total nephrectomy remains the treatment of choice in these cases. The laparoscopic approach has been described for highly selected cases.

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