Rare disease

Xanthogranulomatous pyelonephritis in early infancy

Anand Upasani, JaiBharat Panwar, Sudhir Chandna, Dhiren Patel

Paediatric Surgery Department, Smt NHL Municipal Medical College and Sheth Vadilal Sarabhai General Hospital, Ahmedabad, India

Correspondence to Dr Sudhir Chandna, sudhirchandna@gmail.com

Summary

Xanthogranulomatous pyelonephritis is a severe, atypical form of chronic renal parenchymal infection, which mimics neoplasia and other inflammatory renal parenchymal diseases. Although xanthogranulomatous pyelonephritis has characteristic findings on sonography and CT scan, a clinical diagnosis is seldom possible. Correct diagnosis is made on exploration, by the presence of dense adhesions to the surrounding tissue and presence of pus in the kidney, which is confirmed histopathologically by the presence of lipid laden foamy macrophages, accompanied by both chronic and acute phase inflammatory cells.

BACKGROUND

About 50 cases of xanthogranulomatous pyelonephritis (XGP) in childhood have been reported. XGP is very rare in early infancy. Our case was 40 days old when seen by us, although the symptoms and the palpable lump were present since 1 month of age. On review of the English literature, this is probably the second youngest infant to be reported after Youngson reported a 21-day-old neonate with XGP in 1990.

As XGP is seen in the neonatal period as well as early infancy, there is a need of awareness for a good ante-natal follow-up to be able to understand the aetio-pathogenesis.

CASE PRESENTATION

A 40-day-old male child presented with excessive crying, fever and failure to thrive. The child was clinically pale and had a lump in the right flank which was ballotable and bimanually palpable. Systemic examination was otherwise normal.

INVESTIGATIONS

Blood investigations revealed microcytic hypochromic anaemia with a haemoglobin of 8 g/dl and leucocytosis. C reactive proteins were raised 102 mg/l and renal profile was normal. Ultrasound revealed a hypoechoic mass lesion replacing the right kidney while the left kidney was

Figure 1 CT scan of the abdomen with contrast showing heterogenous right renal mass with poor function on the right side.

Figure 2 Histopathology showing lipid laden foamy macrophages (FM) and diffuse inflammatory cell infiltrate (IF).

normal. The CT with contrast of the abdomen showed 6.74×5.60×4.5 cm sized heterogeneous mass lesion arising from the posterior surface of the right kidney (figure 1). The mass appeared to push renal parenchyma anteriorly. There was evidence of loss of fat planes at places. The renal function was significantly reduced on the right side. Left kidney appeared normal. A possibility of mesoblastic nephroma or Wilm's tumour of the right kidney was suggested.

The postoperative histopathology however, revealed lipid laden foamy macrophages (figure 2), accompanied by diffusely scattered both chronic and acute phase inflammatory cells. Normal renal parenchyma was seen at places along the perimeter. The final diagnosis was of XGP.

No antenatal investigations were available in this case.

DIFFERENTIAL DIAGNOSIS

XGP is a severe, atypical form of chronic renal parenchymal infection. Its manifestations mimic those of neoplasia (mesoblastic nephroma, Wilm's tumour) and other chronic inflammatory renal parenchymal diseases including tuberculosis. As a result it is often misdiagnosed clinically.

TREATMENT

The patient was taken up for laparotomy to carry out a transperitoneal right nephrectomy. The mass involved the right kidney and was densely adherent to the surrounding structures and the retroperitoneum. There were multiple areas of necrosis and haemorrhage. The pedicle of the kidney could be approached with great difficulty because of dense adhesions and while separating the kidney from the retroperitoneum, about 20 cc of yellowish pus like material was aspirated.

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery postoperatively and continues to do well 1 year postoperative.

DISCUSSION

XGP is an atypical form of severe chronic renal parenchymal infection.¹ It accounts for 1/6000 surgically proven cases of chronic pyelonephritis.² About 50 cases of XGP in childhood have been reported.³ The earliest age of presentation reported so far ranges from 21 days in English literature⁴ and 48 days in Japanese literature.⁵ XGP is very rare in early infancy. Our case was 40 days old when seen by us, although the symptoms and the palpable lump were present since 1 month of age.

XGP is characterised by destruction of parenchyma, and accumulation of lipid laden macrophages which surround abscess cavities or present as discrete yellow nodules.⁶ The exact aetiology of XGP is not known, but there is a general acceptance that a combination of long-term renal obstruction and infection is responsible for the process.² Most of these children present with symptoms of chronic infection, like recurrent fever, weight loss, pallor and failure to thrive etc.³ Although bacterial culture from urine or renal abscess reveal *Proteus* as the most common offending organism,^{1 3} but in this case cultures were sterile. Both diffuse and focal forms of the disease have been reported, with the diffuse form being more common.^{1 7} Characteristic sonography and CT appearances of XGP have been reported. However, these features are not specific and mimic neoplasia, so an accurate diagnosis is seldom made preoperatively.^{1 2 6} This patient also was misdiagnosed presumptively as a renal neoplasm and the correct diagnosis was established postoperatively.

The fact that neonates and infants are presenting with a full blown picture of XGP as early as 21 and 40 days of birth raise a suspicion of a congenitally obstructed system with the possibility of a transplacental vertically transmitted infection. This highlights the need of a good antenatal follow-up including documentation of maternal infections and foetal sonography in such cases.

Learning points

- ▶ XGP is very rare in early infancy; one should keep it in mind when a unilateral kidney lump is observed in this age group.
- ▶ Although preoperative differentiation of XGP from neoplasia and inflammatory renal conditions including tuberculosis is almost impossible, nephrectomy remains the treatment of choice.
- ▶ On exploration presence of dense adhesions to the surrounding tissues and aspiration of pus should alert the surgeon to the possibility of XGP, so that a less heroic approach may be taken while performing the nephrectomy and thus reduce postoperative morbidity.
- ▶ Good antenatal follow-up will add to our understanding of the aetio-pathogenesis of XGP in early infancy.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Quinn FMJ**, Dick AC, Corbally MT, *et al*. Xanthogranulomatous pyelonephritis in childhood. *Arch Dis Child* 1999;**81**:483–6.
2. **Thattakkat K**, Morgan H. A child with abdominal mass and severe anaemia. *BMJ Case Rep* 2009;2009.
3. **Watson AR**, Marsden HB, Lendon M, *et al*. Renal pseudotumours caused by xanthogranulomatous pyelonephritis. *Arch Dis Child* 1982;**57**:635–7.
4. **Youngson GG**, Gray ES. Neonatal xanthogranulomatous pyelonephritis. *Br J Urol* 1990;**65**:541–2.
5. **Yazaki T**, Ishikawa S, Ogawa Y, *et al*. Xanthogranulomatous pyelonephritis in childhood: case report and review of English and Japanese literature. *J Urol* 1982;**127**:80–3.
6. **Sharma A**, Rattan KN, Arora BB, *et al*. Xanthogranulomatous pyelonephritis in children. *Indian J Urol* 2000;**16**:153–4.
7. **Subramanyam BR**, Megibow AJ, Raghavendra BN, *et al*. Diffuse xanthogranulomatous pyelonephritis: analysis by computed tomography and sonography. *Urol Radiol* 1982;**4**:5–9.

This pdf has been created automatically from the final edited text and images.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit

<http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Upasani A, Panwar JB, Chandna S, Patel D. Xanthogranulomatous pyelonephritis in early infancy. *BMJ Case Reports* 2012; 10.1136/bcr.12.2011.5305, Published XXX

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow

Keep up to date with all published cases by signing up for an alert (all we need is your email address) <http://casereports.bmj.com/cgi/alerts/etoc>