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Giant Abdominal Masses from Obstructive Uropathy: Report of an Elusive Diagnosis

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Summary

Ogunbiyi OA, Oduwole O and Akingbehin NA. Giant Abdominal Masses from Obstructive Uropathy: Report of an Elusive Diagnosis. *Nigerian Journal of Paediatrics* **1981; 8: 98.** A 9-year old boy with abdominal masses from obstructive uropathy which eluded diagnosis over a 4-year period, is reported. Appropriate radiological investigations eventually established the diagnosis, but unfortunately after irreparable renal damage had occurred. The pitfalls in the diagnosis of the patient as well as the important role of radiology in establishing accurate diagnosis of 'difficult' abdominal masses are discussed.

Introduction

The discovery of an abdominal mass in a child should prompt an urgent search for the underlying pathological lesion. In Europe and North America, 50% of abdominal masses in children are due to non-surgical problems such as leukaemia, Hodgkin's disease and various storage disorders while the rest are of surgical importance. About two-thirds of the surgical tumours are of renal origin and 40–50% of these are obstructive uropathies, i.e. renal cysts and hydronephrosis.[1,2] In tropical Africa, the importance of obstructive uropathies as causes of abdominal masses is less appreciated because other causes of abdominal tumours such as Burkitt's lymphoma, tuberculosis and various parasitic tropical disorders are more commonly seen. Diagnostic delays of obstructive uropathies may therefore occur and result in irreversible renal damage. The present case report illustrates the occurrence of such a preventable catastrophy and emphasizes the important role of radiology in the prevention of diagnostic pitfalls in cases of abdominal tumours.

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Case Report

PF, a 9-year old schoolboy, was first seen at the University College Hospital (UCH), Ibadan, on 14/2/80 following a referral from the Adeoyo State Hospital, Ibadan. The presenting complaint was increasing abdominal mass of 5 years' duration. The mass was otherwise symptomless initially, but became painful and tender 2 years after the onset. He was, at this stage, taken to a general hospital in his home town, Benin City, where he was given some drugs but without improvement. He later sought and received medical care in 4 other general hospitals in Osogbo, Ife, Ogbomoso and Ibadan, but again without improvement. Recourse was then taken to traditional therapy for a while before he presented at

the Adeoyo State hospital, Ibadan, in late January, 1980, where provisional diagnoses of Burkitt's lymphoma and chronic myeloid leukaemia were made.

Clinical history taken on presentation at the UCH yielded no significant additional information. His appetite had remained good, and there had been no history of diarrhoea, vomiting or cough. His urinary stream was reportedly good, but haematuria had been observed.

General physical examination revealed a pale cachectic boy with grossly distended abdomen and distended abdominal veins (Fig. I). He was not jaundiced, and there was neither peripheral lymphadenopathy nor pedal oedema. The oral temperature was 37.5°C.

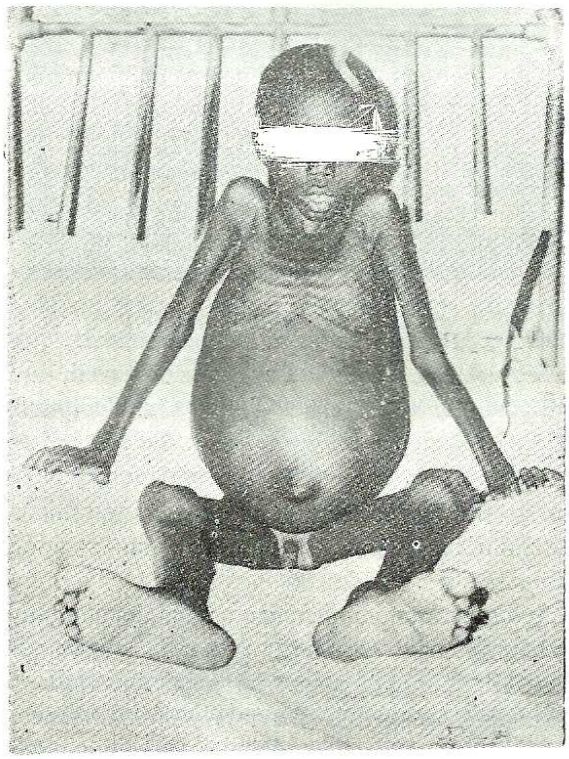

Fig. 1 *Photograph of the patient showing gross abdominal distension and cachexia.*

Examination of the abdomen revealed gross ascites and multiple ballotable masses in the left flank, the left paraumbilical area, and in the right iliac fossa. The liver was enlarged to 10 cm below the costal margin, but the spleen was not enlarged.

Examination of the cardiovascular system revealed a pulse rate of 144/minute, and a blood pressure of 150/110. The jugular venous pressure was raised by 3 cm, but the heart was not clinically enlarged, and the heart sounds were normal. In the respiratory system, there was tachypnoea; the respiratory rate was 50/minute, and the lung fields were clear on auscultation. There was no significant abnormal finding in the nervous system. The provisional diagnoses listed by the admitting physician were abdominal tuberculosis, Burkitt's lymphoma, nephroblastoma, hydronephrosis and neuroblastoma.

Laboratory investigations included serum sodium, 113 mmol/L (113m Eq/L); potassium, 3.8 mmol/L (3.8 mEq/L); chloride 99 mmol/L (99mEq/L); bicarbonate, 9 mmol/L (9mEq/L) and urea, 30 mmol/L (180mg/100ml). The liver function tests were normal while the haematological indices were: PCV 20%; WBC 1.46×10^9/L (14,600 cm^3) with a neutrophil of 83% and a lymphocyte of 17%. Serum calcium, phosphorus, and alkaline phosphatase were within normal limits. The serum creatinine level was raised at 200 umol/L (2.3mg/100ml). Heaf test was negative and blood cultures remained sterile after 7 days' incubation. Urine analysis revealed microscopic haematuria, while an abdominal tap yielded a straw-coloured fluid which on microscopy, was found to contain numerous polymorphs and also aggregates of degenerated cells. Bacteriological cultures were negative for pyogenic organisms, as well as for acid-fast bacilli (AFB).

Radiological examination of the chest revealed elevated diaphragms, but there was no pleural or pulmonary abnormality and the heart was not enlarged. Radiographs of the abdomen revealed multiple soft tissue masses, but no calcifications. A skeletal survey revealed no evidence of bone disease. Renograms, including a high dose intravenous urography with tomography and delayed film were carried out but there was no excretion in both kidneys.

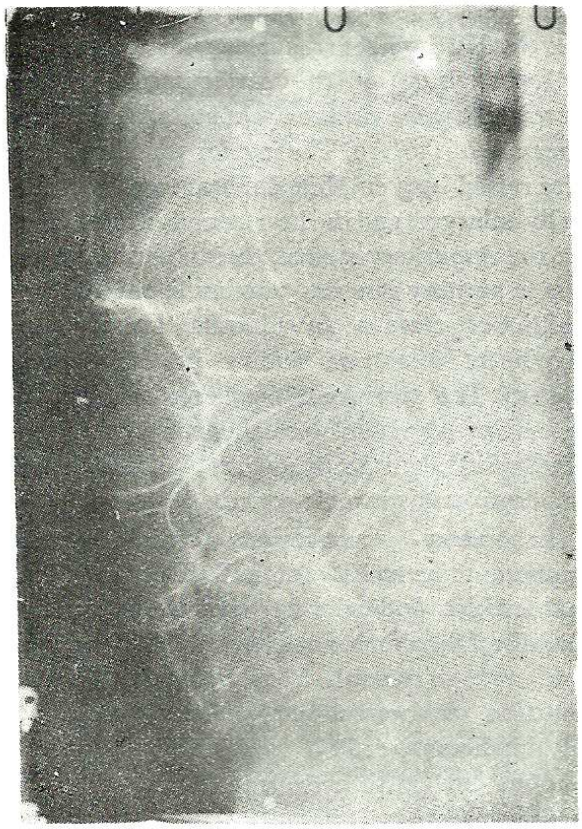

Fig. 2 Selective right renal arteriogram showing attenuated and stretched intra-renal arteries spread round "cysts" in a huge kidney. Note also, the very thin renal cortical tissue.

Because of the urographic findings and the presence of microscopic haematuria, the working diagnosis was at this stage revised to Wilm's tumour, and the patient was accordingly started on actinomycin D and vincristine. He was also started on anti-hypertensive agents for his hypertension. The cytotoxic drugs however produced no detectable reduction in the tumour size after one week.

Further radiological investigations[3,4] were considered desirable at this stage. A renal angiogram and flush aortogram were then carried out and these showed grossly enlarged kidneys and many cysts. The renal arteries were of diminished calibre, while the intra-renal vessels were pruned, attenuated and spread round the cysts. The renal cortex was very thin and bordered on non-existence. The appearance was suggestive of polycystic kidneys or bilateral giant hydronephrosis (Fig. 2). A subsequent micturating cystourethrogram revealed a large thick-walled bladder with sacculations and multiple diverticula. No vesico-ureteric reflux was however observed, but the posterior urethra was dilated and elongated (Fig. 3). These appearances were consistent with a diagnosis of obstructive uropathy due to posterior urethral valve. The urinary stream was fairly good during micturition, but there was considerable bladder residue.

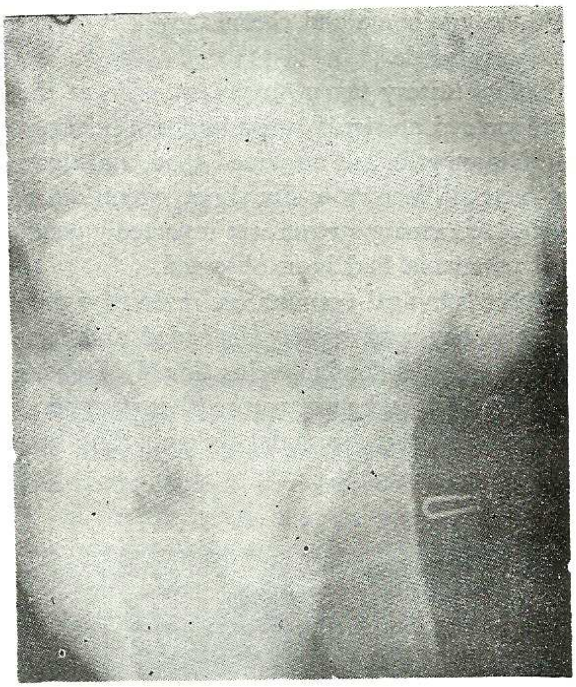

Fig. 3 Micturating cystourethrogram showing elongated and dilated posterior urethra.

At laparatomy, both kidneys were found to be grossly distended with large quantities of infected urine which, apparently, was withdrawn during the abdominal tap. The normal renal architecture was undefinable. The urinary bladder and urethra were also distended and contained 2.2 litres of urine.

Two days post-operatively, an antegrade pyelogram was carried out through the nephrostomy tube and this revealed elongated, dilated and tortuous ureters (Fig. 4). The patient's

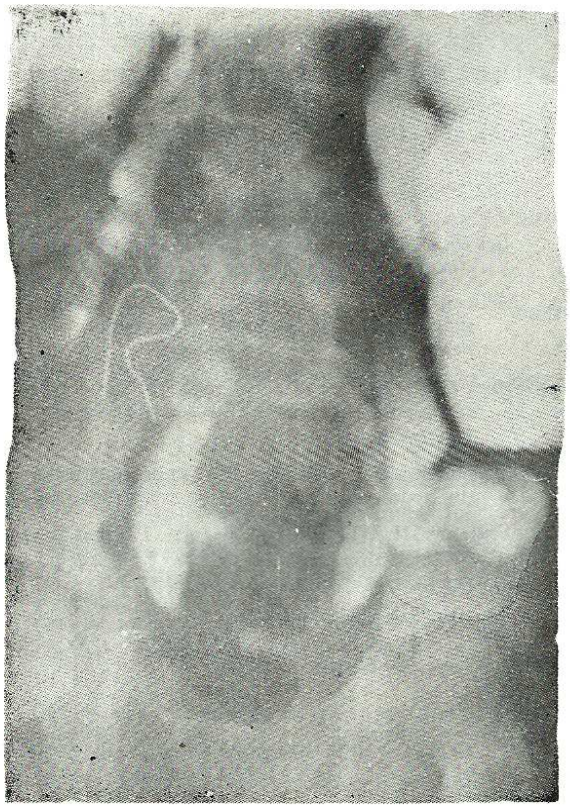

Fig. 4 Antegrade pyelogram showing elongated, dilated and tortuous ureters.

general post-operative status remains satisfactory, but he continues to require maintenance anti-hypertensive medication.

Discussion

The long delay in the diagnosis of posterior urethral valve in this patient may be attributed to a combination of several factors. Perhaps the most important was the absence of symptoms normally associated with urethral valves. A poor urinary stream is said to be the hallmark of urethral valves, but this was conspicuously absent even during observations in hospital. Vesical symptoms such as frequency, enuresis and burning sensation on micturition are other common complaints. These are thought to be due to the associated urinary tract infections, but such symptoms were again not reported by our patient.

Williams *et al*[5] and Hendren[6] have drawn attention to the broad clinical spectrum of urethral valves, and not infrequent occurrence of misleading presentations similar to those exhibited by this patient. Some affected children may have remarkably good urinary stream, and, in these, it seems that massive hypertrophy of the bladder may be able to compensate for the urethral obstruction, but usually at the expense of very considerable upper renal tract damage. In addition, such children may also fail to exhibit vesical symptoms despite the presence of associated urinary tract infection. Misleading presentations like these may, therefore, not readily suggest the correct diagnosis except to physicians with a high index of suspicion.

Another important factor for the long diagnostic delay in this patient is the very limited laboratory and radiological facilities available in most of our general hospitals. Although radiological facilities may be available in some of these hospitals, they are generally limited in scope and have to be used very selectively because of financial constraints. Viewed against this background, the fact that appropriate radiological examinations were not done until the patient came to UCH becomes less surprising. Whatever the constraints however, the importance of adequate radiological studies in the diagnosis of "difficult abdominal tumours" cannot be over-emphasized, and an intravenous urogram is a particularly useful investigation in this regard. If properly conducted, a urogram will reveal whether the urinary tract is the source of a tumour, and even if a mass arises outside the urinary tract, the kidneys and ureters serve as very useful landmarks since their displacement by any mass will give clues about the organ causing the displacement. Further relevant studies may then be carried out as necessary.

The ultimate prognosis in our patient is poor in view of the advanced renal damage that has already taken place. Renal transplantation may offer some hope to patients with his degree of renal damage, but this is as yet not available in Nigeria, nor can it be considered a justifiable alternative to early diagnosis and corrective surgery in cases of urethral valves. Some system must, therefore, be evolved for preventing this type of tragedy in all future cases of abdominal masses due to congenital urethral valves. It is relevant to know in this regard that posterior urethral valves are not rare in our environment since an average of 6 cases present in UCH annually. The long-standing dictum that "the sun should never set on an abdominal tumour in a child" may not yet be feasible in our setting, but every effort should be made to achieve early diagnosis in all cases. Medical practitioners in our peripheral hospitals should, therefore, be aware of their limitations in this regard and refer promptly, "difficult diagnoses" of abdominal tumours to large referral hospitals where facilities for comprehensive investigations are available.

Acknowledgements

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