CONGENITAL ANTERIOR URETHRAL DIVERTICULUM

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SYNOPSIS

We report the first documented case in Singapore of a congenital saccular anterior urethral diverticulum causing bladder outlet obstruction in a two year old boy. Anterior urethral diverticulum is a rare cause of bladder outlet obstruction and usually presents with a poor urinary stream and dribbling. A palpable mass, at the ventral surface of the mid penile urethra or the peno-scrotal junction, which empties on pressure should alert the clinician to the diagnosis. The diverticulum has unique radiologic features that are readily demonstrable by urethrography. A micturating cystourethrogram is the investigation of choice. Keywords: Congenital saccular anterior urethral diverticulum, Penile, peno-scrotal mass, Micturating cystourethrogram

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CASE REPORT

A healthy two year old boy, the first child of a young couple, presented with a history of dribbling urine since birth. The parents noticed that he had never had a good urinary stream. There was no history of urinary tract infection or colicky pain. On examination, except for a distended bladder, no abnormality was found. Both testes were descended, urine microscopy was normal. An ultrasound examination was done as a preliminary radiological study. This showed a distended bladder with thickened walls. The posterior urethra could not be imaged. The kidneys were normal. A posterior urethral valve was suspected and a micturating cystourethrogram (MCU) was done. There was no difficulty in catheterizing the child. The MCU revealed a trabeculated bladder which had a capacity of about 400c.c. (Fig. 1) and a dilated posterior urethra (Fig. 2). There was no vesico-ureteric reflux. On micturition, the classical arcuate filling defect of the proximal lip of an anterior urethral diverticulum was shown (Fig. 2). The diverticulum was at the ventral surface of the mid penile urethra and measured about 3.5 cm in length. Distal to the diverticulum, there was an abrupt change in calibre of the stream. This is marked by the distal lip (Fig 2, see also Fig 6a). An ascending urethrogram was then done to demonstrate the penile urethra better (Fig. 3). A 24 hour delayed film showed the div-

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Department of Paediatrics Tan Tock Seng Hospital H K Cheng, M. Med. (Paeds), F.R.A.C.P. Head and Consultant Paediatrician erticulum to advantage (Fig. 4). Some residual contrast was also seen in the bladder (not shown). A repeat physical examination revealed a cystic mass in the ventral surface of the penile urethra which was missed initially (Fig. 5). The mass was not attached to the overlying skin and was mobile laterally. No attempt was made to empty it. A careful urethroscopy revealed the midline diverticulum in the ventral surface of the penile urethra. Diathermy of the obstructing lip was attempted with partial success and an open diverticulectomy and urethroplasty had to be done. This relieved the obstruction. The patient has remained symptom free.

DISCUSSION

Congenital saccular anterior urethral diverticulum is rare. Enriquez et a(1) in 1978 in a review of the literature discovered only 36 cases to which he added a further six. It may present at any age; from the moment of birth(6) to as old as 21 years(4). Most present in infancy with urinary obstruction or retention. When they present after infancy, they do so with recurrent urinary tract infections or dribbling of urine(2). Although frequently confused with the anterior urethral valve, the urethral diverticulum has unique radiologic features. Indeed, many previously diagnosed cases of anterior urethral valve were probably misdiagnosed cases of anterior diverticulum(3). Once recognised as a distinct entity, the diagnosis is simple. A careful history will reveal that the child never had a good urinary stream since birth and a tell tale sign is a cystic mass at the mid penile urethra or the peno-scrotal junction. If it is uninfected and without complications, the mass is unattached to the overlying skin, nontender and mobile laterally. On compression, urine will be seen dribbling out of the external meatus and the mass is seen to empty. The presence of the mass also serves to exclude the anterior urethral valve with which this condition is often confused. A palpable bladder may also be present and if there is vesicoureteric reflux, there may also be unilateral or bilateral hydronephrosis.

The radiologic findings are classical. Smith(2) emphasized the importance of good quality conventional radiographs as well as a high flow rate of urine during the MCU. The radiographs must demonstrate the proximal lip to distinguish it from the anterior urethral valve (2,3,4). The high flow rate will ensure proper filling of the diverticulum so that it will obstruct and demonstrate the distal lip and marked change in cal-

ibre of the penile urethra. In other words, the filled diverticulum functions like a valve. An ascending urethrogram was also done in our case. This showed the distal lip better (Fig 4). This is in contradiction to the belief of Smith(2) and Williams(4). According to Smith(2), if an ascending urethrogram is performed alone, the distal obstructing fold may be pressed against the urethral wall and thus may not be shown and the diagnosis missed. The antegradely filled anterior urethra may also attain the same calibre as the diverticulum and the proximal lip, which is essential for diagnosis, may not be valvular to the ascending fluid(4). Unless the radiographs are of a good quality and the index of suspicion high, the diagnosis may not be apparent. The diverticulum has a wide mouth and is situated in the midline, in the ventral surface of the anterior urethra. Its usual site is the mid penile urethra although a more proximal position at the peno-scrotal junction has also been described.

Urethral diverticula may be congenital or acquired. Congenital diverticula can be further subclassified into the saccular, the narrow necked diverticulum and the diffuse or megalourethra. Acquired diverticula are usually post-traumatic. Only the congenital saccular diverticulum will be discussed below.

The aetiology of congenital saccular diverticulum is not known(3) although many hypotheses have been put forward. These include a lack of or faulty development of the corpora spongiosa(5), defective fusion of the ventral urethral folds over the urethral grooves(5), partial duplication, urinary obstruction in foetal life and ectopic epidermal pockets communicating with the ventral wall of the urethra(4).

The primary differential diagnostic considerations include anterior urethral valve, dilated Cowper's gland ducts and post traumatic diverticulum. The presence of a penile or peno-scrotal mass clinically and the proximal lip radiologically, which is seen as an arcuate filling defect, should readily distinguish the diverticulum from the valve. In addition, the proximal lip of the diverticulum forms an acute angle with the rest of the urethra (Fig. 6a) whilst that of the anterior valve forms an obtuse angle(3) (Fig. 6b). In dilated Cowper's gland ducts, a tubular channel is seen in the ventral surface of the bulbous urethra which it parallels. Its termination is in the urogenital diaphragm. In acquired traumatic diverticulum, the undersurface is usually rough as it is formed by granulation tissue. It is also slightly more proximal in the bulbous urethra, a site less common for the congenital saccular diverticulum. It must be stressed that absence of the mucosa lining does not differentiate one form of diverticulum from the other(5). The urethrogram will not show the proximal and the distal lips of the diverticulum but rather, a narrowing proximally and distally.

In summary, a history of poor urinary stream and dribbling, recurrent urinary tract infections and a palpable penile or peno-scrotal mass on physical examination strongly suggest the diagnosis of a congenital saccular anterior urethral diverticulum. A micturating cystourethrogram will confirm the diagnosis. Early surgical intervention will prevent the onset of uraemia.



Fig. 1. Filling phase of the MCU showing a trabeculated bladder.



Fig. 2. MCU demonstrating the anterior urethral diverticulum. (The proximal and distal lips have been retouched for clarity)



Fig. 3. Better demonstration of the distal lip by the ascending urethrogram.



Fig. 4. A 24 hour delayed radiograph showing residual contrast in the diverticulum (arrow).



Fig. 5. Clinical demonstration of the cystic mass (arrow) in the ventral surface of the mid penile urethra.



Fig. 6a. Diagram of the anterior urethral diverticulum.



Fig. 6b. Diagram of the anterior urethral valve.

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