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Inflammation and infection

A rare case of an enormous sacral meningocele causing ureteric obstruction

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ABSTRACT

Sacral Meningoceles, ductal ectasia and pseudomeningoceles are all rare spinal defects that occur due to errors in collagen biosynthesis in the setting of Marfan's Syndrome. Meningoceles, which are extradural collections of cerebrospinal fluid, can form large pelvic collections which can compress local structures. In rare cases, this can lead to extrinsic ureteric obstruction, which can result in acute renal failure and urosepsis. We present a case of a 35-year old female with Marfan's syndrome, with one of the largest sacral meningoceles reported in the literature, causing acute ureteric obstruction, requiring urgent surgical intervention.

Introduction

Marfan's Syndrome is an autosomal dominant connective tissue disorder, which results from a genetic mutation in fibrillin-1, responsible for the production of collagen.[1] The resulting weakness in the connective tissue, predisposes these patients to a number of conditions, including dural ectasia. Dural ectasia, present in over 90% of patients with Marfan's syndrome, refers to a widening of the thecal or dural sac as result of the defective elastic fibres.[2] In rare cases, the weakness of the dura results in a herniation of the cerebrospinal fluid out into retroperitoneum and pelvis, termed a meningocele.[1] Meningoceles can form both anteriorly and posteriorly to the spinal cord, and in their most severe form can present as large pelvic masses.[1] We report one of the largest sacral meningoceles seen in the literature, causing right sided ureteric obstruction and subsequent urosepsis in a 35-year-old female with a diagnosis of Marfan's syndrome.

Case presentation

The patient presented to a regional emergency department with a two-week history of worsening suprapubic pain, an abdominal mass and subjective fevers. She had previously undergone repair of sacral dural ectasia and a large meningocele extending from the S1 nerve root. This meningocele had reaccumulated on follow up, five years after her surgery, measuring 16.8 × 11.6 × 11 cm on magnetic resonance imaging (MRI) (Fig. 1.). The size of this meningocele is noted to be one of the largest reported in the literature.[2]

On clinical assessment she was eliciting signs of urosepsis – with a fever of 38.3° Celsius, a blood pressure of 95/64 mm Hg and a heart rate of 110 beats per minute. Her urine dipstick revealed presence of leucocytes and nitrites. Fluid resuscitation and broad-spectrum intravenous antibiotics were administered, and an indwelling urethral catheter inserted. She underwent a computed tomography (CT) scan, which revealed a persisting meningocele with evidence of external compression upon the right ureter with resultant right hydroureteronephrosis (Fig. 2a.) A decision was made to take the patient to theatre, where a rigid cystoscopy and bilateral retrograde pyelogram was performed, revealing evidence of extrinsic compression of the bladder and distal ureter with associated right hydronephrosis (Fig. 2b). A ureteric stent was then inserted. The patient made a full recovery. She was referred back to her neurosurgeon and booked for a ureteric stent exchange in six months' time.

Discussion

Due to errors in collagen biosynthesis, Marfan's syndrome predisposes patients to hypermobile joints, aortic root aneurysms and dural defects.[1] The description of meningoceles in the literature is limited to only a small number of cases.[3] Symptoms related to this condition depend on its size and location. Often meningoceles can remain

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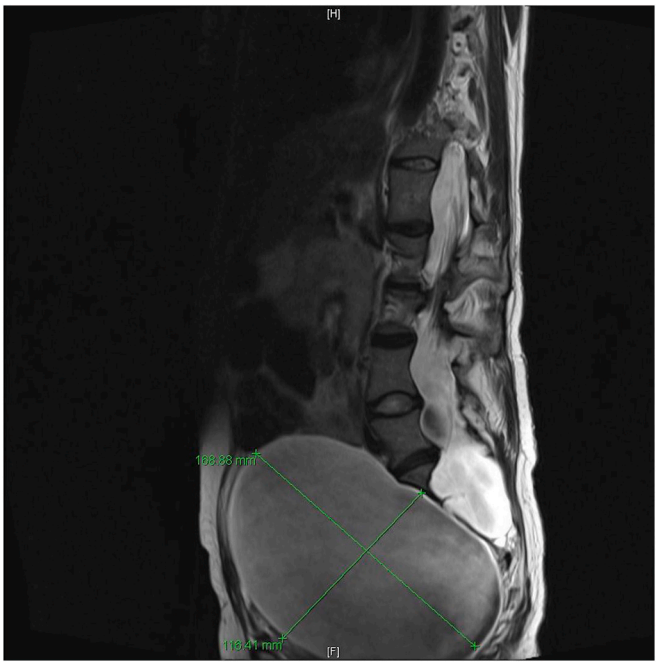

Fig. 1. Sagittal T2-weight fat supressed magnetic resonance imaging demonstrating a large meningocele extending from the right S1 nerve root, measuring 16.8 × 11.6 cm, causing significant displacement of pelvic organs.

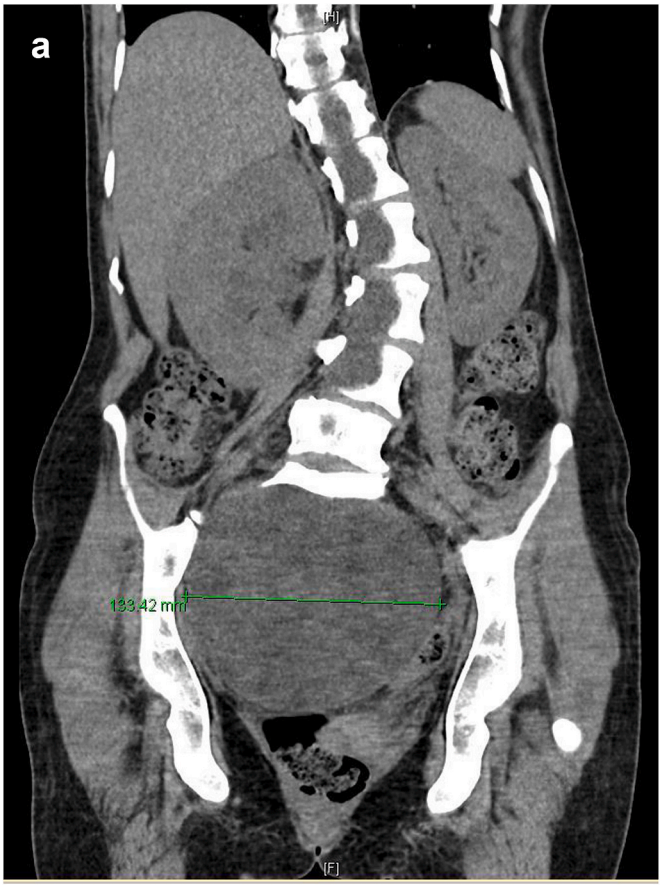

Fig. 2a. Coronal CT KUB view of new right hydroureteronephrosis caused by external compression of the pelvic meningocele. There had been no interval change in the size of the meningocele (measured). The scoliosis is a feature of the spinal dysraphism caused by Marfan's Syndrome.

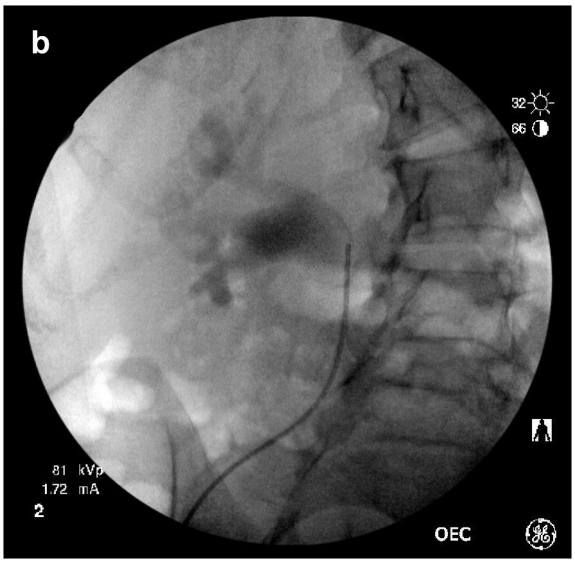

Fig. 2b. A Right Retrograde Pyelogram performed prior to insertion of ureteric insertion revealing hydroureteronephrosis and a very tortuous ureter, with evidence of extrinsic compression at the distal end.

asymptomatic. As the condition progresses, extrinsic compression of the bowel, rectum and urinary tract can result.[3] Radiculopathy from sacral nerve compression has also been reported.[3] Magnetic Resonance Imaging (MRI) is the recommend imaging modality to assess the meningocele sac.[3]

Definitive management by a spinal surgeon can be either observation or surgical decompression.[3] The surgical goal is to excise the sac and disconnect it from the spinal subarachnoid space.[4] This can be performed from either an anterior or posterior approach.[3] Lumboperitoneal shunting to divert CSF and the use of fibrin glue injection has also been used in some cases.[2,5] Needle guided aspiration should be avoided as it increases risks of meningitis.[1] Management of the ureteric obstruction should continue through ureteric stent exchange until the meningocele has been treated.

Conclusion

When assessing patients with a known diagnosis of Marfan's syndrome, rare spinal conditions such as dural ectasia, meningoceles and pseudomeningoceles must be considered. In the setting of urosepsis, appropriate resuscitation and timely administration of antibiotics should be instigated, followed by early involvement of both urological and spinal surgeons.

Consent

Consent was obtained from the patient for publication of this case report and accompanying images.

Declaration of competing interest

None to declare.

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Nil.

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