Clinical Reports

Gram-negative meningitis and chronic constipation: an unusual presentation of caudal regression syndrome

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Summary: Congenital malformations may present as meningitis caused by enteric organisms, but this is extremely rare and occurs almost exclusively in the paediatric population. We report an unusual case of a young man with chronic constipation presenting with spontaneous Gram-negative meningitis due to an underlying congenital spinal malformation known as the caudal regression syndrome.

Introduction

Gram-negative meningitis due to enteric organisms is a rare condition, usually associated with a predisposing cause such as craniovertebral trauma, neurosurgical procedures, septicaemia, alcoholism or debilitating illness. Congenital malformations are rarely considered in adult patients. We present the case of a 30 year old man with spontaneous meningitis due to enteric Gram-negative bacilli. Subsequent investigations led to the diagnosis of a congenital spinal malformation known as the caudal regression syndrome. To our knowledge, this is the first report of meningitis complicating caudal regression syndrome in a previously asymptomatic adult.

Case report

A 30 year old man presented with a 2 hour history of severe occipital headache, photophobia, nausea and rigors. The headache commenced suddenly while straining at stool. He had chronic constipation secondary to congenital anal stenosis, treated by dilatation soon after birth. He passed stool only every 7–10 days. There was no impairment of urological or sexual functioning. On examination he was febrile (38.9°C) and distressed with marked neck stiffness. Pes cavus was present. There were no cutaneous abnormalities or deformities in the lumbosacral region to suggest spina bifida occulta. There were no localizing neurological signs.

Examination of the heart, lungs and abdomen was normal. A computed tomographic scan of the brain was normal. Lumbar puncture revealed turbid cerebrospinal fluid. The opening pressure

was 32 cm H₂O (normal <20). The glucose was <0.1 mmol/l and the protein was 7.90 g/l. Microscopy revealed 393 leucocytes/μl (99% polymorphs) and Gram-negative bacilli. Cultures grew *Escherichia coli* and Group B Streptococcus.

The patient was treated with intravenous cefotaxime, amoxycillin and gentamicin, but remained unwell with fever, headache and neck stiffness. A further cerebrospinal fluid examination showed persisting neutrophil pleocytosis consistent with ongoing infection. The bacteriological findings suggested the presence of a neurenteric fistula and the pes cavus suggested an underlying spinal malformation. Plain films of the lumbosacral spine showed a hypoplastic malformed sacrum which deviated to the left (Figure 1). A magnetic resonance imaging scan showed tethering of the spinal cord and dysgenesis of the sacrum below S2. The dural sac extended below the sacral remnant and was contiguous with a cystic structure having the features of an anterior meningocele (Figure 2). The meningocele was closely applied to the posterior wall of the rectum. The rectum was dilated and the deep supporting structures of the anal canal were abnormal, with associated stenosis of the canal. A direct communication between the rectum and the meningocele (neurenteric fistula) was not demonstrated.

Because of failure to improve on appropriate antibiotics, surgery to explore the meningocele was undertaken. The rectum and colon were found to be dilated. The rectum was mobilized and separated from the meningocele and omentum was interposed. A temporary loop colostomy was fashioned. Subsequently, the patient improved with resolution of his symptoms and fever. Antibiotics were continued for a further 3 weeks and the colostomy was later closed.

Figure 1 Plain X-ray of the lumbosacral spine demonstrating hypoplastic malformed sacrum.

Figure 2 Sagittal T₁-weighted magnetic resonance imaging scan of pelvis showing tethering of spinal cord, sacral dysgenesis, dilated rectum and anterior meningocele.

Discussion

Our patient developed meningitis while straining at stool, presumably consequent to the passage of enteric organisms through the bowel wall into the subarachnoid space by way of the meningocele. Such passage would be facilitated by trauma to the rectal mucosa from hard faeces and by high intraluminal pressure during straining. There was no evidence of a neurenteric fistula on the magnetic resonance study or at operation. If a fistula were present, one would have expected recurrent episodes of meningitis from childhood rather than a single episode at the age of 30.

The clinical and radiological features of this case are best explained by the diagnosis of caudal regression syndrome. This term encompasses a spectrum of malformations resulting from a defect of the caudal axial mesoderm during embryogenesis, leading to partial or absent development of the lumbosacral spine and malformations of the anus, rectum and urogenital tract.¹⁻³

In the very mildest case there may be absence of the coccyx only. More severe cases exhibit partial dysgenesis, hypoplasia or unilateral agenesis of the sacrum, as seen in this patient. The sacrum and lower lumbar vertebrae may be absent and these deformities are usually associated with major neurological deficits. Other conditions common to the syndrome include spina bifida, myelomeningocele, tethered cord, anal stenosis or atresia, kidney and genital malformations. Extreme cases of the syndrome exhibit sirenomelia (mermaid deformity) and renal agenesis (Potter syndrome). Among all newborns, anorectal malformations are relatively common congenital defects and up to one third of affected infants have an associated lumbosacral malformation. It has therefore been proposed that at least some of these may represent the minimal expression of the caudal regression syndrome.¹

In our patient, the sacrococcygeal malformation, tethered cord, anterior meningocele and congenital anal stenosis indicated a variant of the caudal regression syndrome. Chronic constipation and straining led to the unusual complication of Gram-negative meningitis. We would suggest that clinicians consider asymptomatic variants of the caudal regression syndrome when previously healthy adults present with Gram-negative meningitis.

References

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