CASE REPORT

Dural ectasia associated with Mounier-Kuhn syndrome: a novel association in the context of spinal stenosis

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SUMMARY

We present the case of a patient undergoing lumbar spine decompression for stenosis with a history of Mounier-Kuhn syndrome. The patient presented with axial lumbar spine pain over 6 months with progressive radicular pain to the left L3 dermatome. MRI confirmed spinal stenosis at L3/4 level with associated dural ectasia. The patient had an uneventful spinal decompression with resolution of radicular symptoms and axial spine pain. Dural ectasia poses a significant risk when operating on the lumbosacral spine. Larger cerebrospinal fluid volumes and a capacious dural canal can result in anaesthetic and orthopaedic complications such as inadequate spinal anaesthesia, complicated epidural analgesia, intraoperative dural tears and difficult pedicle screw insertion due to narrow pedicles. This is the first case in the literature detailing the association between dural ectasia and Mounier-Kuhn syndrome. We recommend adequate spinal imaging in patients with Mounier-Kuhn syndrome to exclude dural ectasia prior to undergoing lumbosacral spinal procedures.

BACKGROUND

This case is of interest as it highlights a novel association between Mounier-Kuhn syndrome and dural ectasia. Given the increased anaesthetic and orthopaedic risks associated with dural ectasia, it is important to acknowledge that patients with Mounier-Kuhn syndrome should have adequate spinal MRI to detect this condition prior to their spinal procedure.

CASE PRESENTATION

This 69-year-old woman presented to the spinal service with a 6-month history of progressive axial lumbar spine pain. The pain intermittently radiated to the left L3 dermatome. Mobilisation exacerbated these symptoms and was severe enough to restrict the patients' walking distance.

This woman was diagnosed with Mounier-Kuhn syndrome 13 years prior to this presentation by a respiratory physician. Symptoms due to this syndrome included recurrent respiratory tract infections and dyspnoea. There was no other relevant background history in this case.

Examination confirmed axial spine tenderness and the radicular nature of the pain by demonstrating

Lasègue's sign. The remaining neurological and vascular examinations were normal.

INVESTIGATIONS

MRI was performed to characterise the lumbar spine pathology. T2 sagittal MRI slices confirmed multilevel lumbar disc herniations with thecal indentation. The most significant level of spinal stenosis was at the L3/4 level (figure 1). At this level, disc protrusion also involved the left neural exit foramen which accounts for the clinical finding of a left L3 radiculopathy.

On review of the images, an incidental diagnosis of dural ectasia was made. Based on the diagnostic criteria described by Ahn *et al*, one major and two

Figure 1 T2 sagittal MRI spine showing L3/4 disc with sacral dural ectasia.

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minor criteria were met as follows: *major criterion*—dural sac wider below L5 compared with the sac above L4. *Minor criteria*—dural sac/vertebral body ratios larger than normal values for each lumbar level, and the nerve root sleeves at L5 were greater than 6.5 mm bilaterally (figure 2).¹

CT of the trachea was also performed preoperatively to confirm the safest position of the endotracheal (ET) tube during general anaesthesia. Mounier-Kuhn syndrome is characterised by tracheobronchomegaly, and so high-resolution imaging with detailed preoperative planning was essential for safe intubation in this case.

DIFFERENTIAL DIAGNOSIS

Dural ectasia may be associated with Marfan syndrome, Ehlers-Danlos syndrome, neurofibromatosis type 1 and ankylosing spondylitis. The diagnosis of dural ectasia in the setting of a patient with Mounier-Kuhn syndrome presenting with lumbar spine stenosis was made on both clinical examination and radiographic evaluation.

TREATMENT

The patient underwent a lumbar spine decompression at L3/4 level consisting of bilateral partial laminectomies, ligamentum flavum hypertrophy excision and discectomy. Intraoperative findings included a tense dural sac with a large volume of cerebrospinal fluid (CSF). This made the decompression more difficult than usual with a higher risk of dural tear.

The patient then underwent a physiotherapy regime postoperatively on the ward and at home on discharge.

OUTCOME AND FOLLOW-UP

The lumbar spine was successfully decompressed without damaging the enlarged dural sac.

The patient was clinically reviewed 2 months postoperatively. She was then mobilising with very little pain, and her axial lumbar spine pain and radicular symptoms at rest were fully resolved.

DISCUSSION

This case illustrates a novel and important association between Mounier-Kuhn syndrome and dural ectasia.

Figure 2 T2 axial MRI spine showing widened exit neural foramina at L5 vertebra.

Mounier-Kuhn syndrome is a rare congenital syndrome affecting the respiratory tract. Loss of elastin fibres and thin smooth muscle in the trachea and bronchi results in tracheobronchomegaly.² This can lead to recurrent respiratory tract infections as our patient had experienced. It may also pose great difficulty with ET intubation. Definitive airways during general anaesthesia rely on an airtight seal between the inflated ET cuff and the trachea. If the cuff is inflated in an area of trachea dilation, the airway will be ineffective leading to ventilation complications.

Dural ectasia has been described in detail by Ahn *et al.*¹ This patient was found to meet one major and two minor diagnostic criteria for the diagnosis of dural ectasia. A diagnosis of dural ectasia has both anaesthetic and orthopaedic implications. Anaesthetic considerations include a higher risk of failed spinal anaesthesia.³ Raised CSF volumes and ineffective anaesthetic circulation in the CSF have been postulated as the cause of this.

Orthopaedic complications of dural ectasia are due to the thecal sac which is more prominent on dissection and therefore more likely to be perforated leading to a dural tear.⁴ Infection rates, blood loss and fixation failure are all increased in dural ectasia. The pedicles are smaller and therefore more difficult to instrument with pedicle screws.

The association between these two entities has not been made before. In recent literature, a novel classification system for Mounier-Kuhn syndrome describes extrapulmonary manifestations as ‘generalised elastolysis’ which is now referred to as Mounier-Kuhn syndrome type 3.⁵ Considering this histological hallmark of elastin atrophy in Mounier-Kuhn syndrome, we propose that this soft tissue involvement may have a causative role in the pathogenesis of dural ectasia in this patient.

We acknowledge the importance of obtaining MRI spine images prior to any spinal procedure on a patient with Mounier-Kuhn syndrome. It is important for the patient’s outcome to rule out the presence of dural ectasia, thereby pre-empting the anaesthetic and orthopaedic complications that go with this condition.

Contributors GAS acquired the data, composed the manuscript and performed the literature review. OG assisted with the literature review, edited the figures included in the paper and assisted with editing the manuscript. AD oversaw the research, provided guidance regarding the scientific angle to approach this paper from, edited the final manuscript and assisted with the literature review.

Competing interests None declared.

Patient consent Obtained.

Patient’s perspective

- I am very happy with the operation I had. My pain is much improved now, and I’m walking better than I was before this all happened.

Learning points

- Mounier-Kuhn syndrome may be associated with dural ectasia.
- Spinal imaging should include preoperative MRI to identify potential pathology in these patients undergoing spinal intervention.
- Anaesthetic difficulties may arise for both general and spinal anaesthesia in patients with Mounier-Kuhn syndrome and dural ectasia. These difficulties must be planned for in the preoperative assessment and workup.

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