Case Report

Cord injury after spinal anaesthesia in a patient with previously undiagnosed Klippel–Feil syndrome

E. Stevens,¹ B. Williams,² N. Kock,³ M. Kitching⁴ and M. P. Simpson⁴

1 Core Trainee, 2 Specialty Trainee, 4 Consultant, Department of Anaesthetics, 3 Consultant, Department of Neurology, East and North Hertfordshire NHS Trust, Stevenage, United Kingdom

Summary

This report presents an obstetric patient with no significant past medical history who underwent spinal anaesthesia for a category-three caesarean section. On examination, she had asymmetrical scapular alignment and a mild scoliosis of the lumbar spine with no functional limitation. Postoperatively the patient developed neuropathic pain symptoms in the right leg which failed to resolve with conventional analgesia. Cervical and lumbar spine magnetic resonance imaging was performed resulting in a diagnosis of a low-lying tethered spinal cord terminating at the level of L5 and congenital fusion of the C7/T1 vertebrae. A tethered spinal cord is a rare condition, which in this case had been completely asymptomatic. However, we suggest that the patient displayed musculoskeletal signs not previously widely reported, which could have indicated the presence of a potential underlying neural tube defect. Based on the imaging findings and the presence of Sprengel's deformity, the patient was diagnosed with Klippel–Feil syndrome.

Correspondence to: E. Stevens

Email: emmastevens@gmx.com

Accepted: 3 December 2018

Introduction

Spinal anaesthetics are widely performed procedures and are the most common form of anaesthesia for caesarean sections. Permanent neurological complications resulting from spinal anaesthesia are rare. The Third National Audit Project (NAP3) of the Royal College of Anaesthetists reports the risk of permanent harm from central neuraxial blockade as being 1 in 23,500 to 1 in 50,500. This is further defined within the obstetric population as being between 1 in 80,000 and 1 in 320,000 [1]. The risk of temporary nerve damage, lasting less than 6 months, is more common and difficult to quantify.

The mechanism of central neuraxial injury can be direct or indirect. Direct causes of neurological injury include mechanical trauma from the needle insertion, intraneural injection, local anaesthetic toxicity and ischaemic incidents secondary to vascular injury [2]. Indirect causes include aetiologies resulting in nerve/cord compression such as haematomas or abscesses. Cases of direct spinal cord injection of local anaesthetic mixture in the presence of low tethered cords, have been reported [3].

A tethered spinal cord is most commonly diagnosed in childhood but can be asymptomatic and the first presentation or diagnosis might rarely occur in the adult population. As such, it should be considered as part of a broad differential diagnosis when patients develop neurological symptoms following spinal anaesthesia.

Report

A 40-year-old primiparous woman was listed for a category-three lower segment caesarean section due to failure to progress despite hormonal induction of labour and augmentation with oxytocin. During labour an epidural was sited following anaesthetic review which ascertained no significant past medical history or contraindications to central neuraxial blockade. On

insertion of the epidural it was noted that the patient had asymmetrical scapular alignment and a mild scoliosis of the lumbar spine. The insertion was otherwise straightforward and effective analgesia established with a patient-controlled epidural analgesia regimen using bupivacaine 0.1% with fentanyl $2 \mu\text{g}\cdot\text{ml}^{-1}$.

Following a decision to proceed to caesarean section, the oxytocin infusion and patient-controlled epidural analgesia regimen were discontinued. However, due to an intercurrent urgent surgical delivery, there was a delay of more than 4 h before arrival to the operating theatre. On assessment, the patient had no sensory or motor block and the decision was made to remove the epidural catheter and proceed with a single-shot spinal anaesthetic.

Spinal anaesthesia was performed at the L4/5 interspace (assessed by palpation of Tuffier's line) using a 25-G Sprotte spinal needle. Following insertion of the needle, the patient reported a shooting pain in the right leg. The needle was withdrawn slightly, after which the pain settled completely, clear free-flowing cerebrospinal fluid was observed and 0.5 ml easily aspirated. There was no pain or paraesthesia on injection of 0.5% hyperbaric bupivacaine 2.5 ml with diamorphine 300 μg . Block onset was mildly delayed, taking 20 min to achieve loss of cold sensation to T4, after which caesarean section was performed without complication.

In the recovery area, following the resolution of spinal anaesthesia, the patient reported worsening shooting pain symptoms and allodynia in the right leg with no sensory or motor neurological deficit. She was initially managed with intravenous morphine for pain control. With no subsequent improvement in symptoms a lumbar spine magnetic resonance imaging (MRI) scan was performed demonstrating low tethering of the cord and conus termination at L5. The filum terminale was visibly thickened and several Tarlov cysts noted within the sacral canal (Fig. 1). At L2, a central linear area of fluid attenuation was noted which could be related to the spinal anaesthetic or a pre-existing syrinx.

Opinions were sought from neurosurgical and neurology teams. The neurosurgical team advised that, as the patient had been previously asymptomatic, despite radiological evidence of a tethered cord neurosurgical intervention was not indicated at this time. Neurology review highlighted an association between a tethered spinal cord and Chiari malformations, advising further MRI imaging of the cervical spine. Although this imaging appeared negative for a Chiari malformation, it did reveal congenital fusion of C7 and T1 vertebral elements, consistent with undiagnosed Klippel–Feil syndrome.

Figure 1 Magnetic resonance imaging of the lumbar spine showing a low-lying tethered spinal cord terminating at the L5 level (white arrow) and central linear area of fluid attenuation at the L2 level (grey arrow).

At follow-up, 3 weeks after discharge, the patient's symptoms had improved. She was later reviewed in a neurology outpatient clinic and her symptoms had almost completely resolved, with only occasional neuropathic pain managed with paracetamol. A small area of numbness on the right ankle persisted, but this was not functionally limiting.

Discussion

Interesting aspects of this case include the identification of a physical symptom that may indicate an increased likelihood of a patient having an underlying neural tube defect. Our case also demonstrates a patient presenting with atypical signs of a neurological injury compared with other similar case reports. In addition, alternative anaesthetic management options that could have been utilised are worth considering for future cases.

The term 'tethered spinal cord' was first used in 1976 by Hoffman et al. to describe an elongated spinal cord and a thick filum terminale [4]. When this is combined with a stretch-induced functional disorder it is known as tethered cord syndrome. The neurological symptoms associated with tethered cord syndrome present most commonly in the paediatric population where it has been demonstrated that surgical intervention may prevent neurosurgical deterioration. A tethered spinal cord may also be asymptomatic until adulthood or may never result in a functional or neurological deficit. However, as awareness of this syndrome has risen as well as increased access to radiological imaging, more adults are being diagnosed. Neurosurgical management (operative vs. non-operative) in adults remains controversial and it is felt that larger studies are needed to evaluate the risks and benefits of both treatment options [5].

It is currently not logistically possible to undertake cross-sectional spinal imaging in all patients before central neuraxial blockade or invasive procedures, such as lumbar punctures, to detect the presence of neural tube defects. Some conditions, such as spina bifida occulta, have well-recognised physical manifestations which may give clinicians a high index of suspicion of neural tube pathology.

In this case, on initial assessment, there was neither immediate evidence nor suspicion of any underlying neurological abnormalities. However, asymmetry of the scapulae was noted, and research suggests that this was likely to be a congenital high scapula (Sprengel's deformity) – a rare congenital deformity of the pectoral girdle [6]. Sprengel's deformity is found in 10–45% of patients with Klippel–Feil syndrome.

The Klippel–Feil syndrome is characterised by congenitally fused cervical vertebrae, as demonstrated on the cervical MRI of this patient. Diagnosis is difficult, as the classic triad of short neck, low posterior hairline and decreased neck range of motion is absent in most patients with the condition and is often dependent on the level of the fusion [7]. The Klippel–Feil syndrome has been reported to be associated with neural tube defects such as spina bifida. Our literature search identified only one case report of a similar nature: a 14-year-old patient who presented with bilateral Sprengel's deformity and neurological symptoms. Imaging revealed spina bifida occulta and a tethered spinal cord [8].

Given its strong association with Klippel–Feil syndrome, it is also worth considering the importance of a thorough airway assessment and recognition of a potentially difficult airway in any patients presenting with Sprengel's deformity due to the possibility of a reduced range of movement at the neck, particularly if the cervical spine fusion is at a higher level than the patient described in this report.

In previously reported cases of complications following spinal anaesthesia in patients with undiagnosed tethered cords, the presenting symptoms were most commonly paraesthesia or weakness in the lower limbs [3]. This patient had no demonstrable loss of sensory or motor function and presented solely with pain in the right leg, which appears to be unusual for this type of injury. We, therefore, suggest that acute persistent pain post central neuraxial blockade warrants the same urgent imaging as more common presentations of a neurological deficit.

With regard to the anaesthetic management, there is no consensus on the approach to a patient with a previously effective labour epidural that has been discontinued for a period of time resulting in no discernible block, who then requires early surgical delivery. In this case, the decision was made to remove the epidural catheter and proceed under spinal anaesthesia as this was felt to be the safer and more reliable option compared with the potential risks of using what was, at that point, considered to be a non-established epidural. These risks included adverse effects due to catheter migration or an inadequate block either pre- or intra-operatively, potentially resulting in conversion to general anaesthetic. It is, however, appreciated that an alternative management plan would be an attempt at an epidural top-up, with further troubleshooting in the event of a block that was unsuitable to proceed or providing inadequate anaesthesia during the course of the delivery.

The central area of fluid attenuation at L2, possibly related to the spinal anaesthetic, highlights the unreliability of using landmark palpation to identify vertebral levels as the intended level had been L4/5 [9]. This unreliability might be exaggerated in

patients with congenital spinal anomalies. A single-shot spinal anaesthetic performed in this patient at L4/5 would have still endangered the cord as it was tethered to L5.

In summary, we suggest that what initially appeared to be a benign musculoskeletal anomaly may indicate the presence an underlying neural tube deformity and should not be overlooked. Performance of central neuraxial blockade in a patient with Sprengel's deformity should include a thorough neurological history and examination and, if time allows, consideration of obtaining appropriate imaging before proceeding. In emergency situations where this is not possible, a risk-benefit analysis must take place and the patient should be carefully consented for the potentially increased risk of neurological injury. We would also suggest that early imaging is important in patients presenting with unexpected pain rather than more classical signs of neurological involvement such as a sensory or motor deficit.

Acknowledgements

Published with the written consent of the patient. No external funding or competing interests declared.

References

1. Cook TM, Counsell D, Wildsmith JAW. Major complications of central neuraxial block: report on the third National Audit Project of the Royal College of Anaesthetists. *British Journal of Anaesthesia* 2009; **102**: 179–90.
2. Kent C, Bollag L. Neurological adverse events following regional anesthesia administration. *Local and Regional Anesthesia* 2010; **3**: 115–23.
3. Liu J-J, Guan Z, Gao Z, Xiang L, Zhou F, Huang SL. Complications after spinal anesthesia in adult tethered cord syndrome. *Medicine* 2016; **95**: 29–33.
4. Hoffman HJ, Hendrick EB, Humphreys RP. The tethered spinal cord: its protean manifestations. *Diagnosis and Surgical Correction Article Child's Brain* 1976; **2**: 145–55.
5. Düz B, Gocermer S, Secer HI, Basal S, Gönül E. Tethered cord syndrome in adulthood the. *Journal of Spinal Cord Medicine* 2008; **31**: 272–8.
6. Guillaume R, Nectoux E, Bigot J, et al. Congenital high scapula (Sprengel's Deformity): four cases. *Diagnostic and Interventional Imaging* 2012; **93**: 878–83.
7. Gruber J, Saleh A, Bakhsh W, Rubery PT, Mesfin A. The prevalence of Klippel-Feil syndrome: a computed tomography-based analysis of 2,917 patients. *Spine Deformity* 2018; **6**: 448–53.
8. Mittal N, Majumdar R, Chauhan S, Acharjya M. Sprengel's deformity: association with musculoskeletal dysfunctions and tethered cord syndrome. *BMJ Case Reports* 2013; **bcr-2013**: 009182.
9. Broadbent CR, Maxwell WB, Ferrie R, Wilson DJ, Gawne-Cain M, Russell R. Ability of anaesthetists to identify a marked lumbar interspace. *Anaesthesia* 2000; **55**: 1122–6.