



Case Report

Interscapular pain with Chiari Type I malformation attributed to atypical spinal accessory neuralgia

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ABSTRACT

Background: The spinal accessory nerve (XI) is traditionally considered a motor nerve. However, as some studies have documented the presence of nociceptive fibers in XI, vascular XI neural compression may lead to an atypical neuralgia.

Case Description: A 27-year-old male presented with a Chiari Type I malformation contributing to interscapular pain. Following a posterior fossa and microvascular decompression of XI, the patient improved, thus confirming the underlying diagnosis of a XI atypical neuralgia.

Conclusion: Unilateral, posterior-interscapular deep pain may be due to an atypical spinal accessory nerve (XI) neuralgia rather than a Chiari Type I malformation or syrinx. Posterior fossa decompression, subpial tonsillar resection, and XI nerve microvascular decompression resolved this patient's complaints. In the future, CTA or MRA vascular studies along with a balanced steady-state gradient echo MRI sequence would be better to document the presence of XI nerve neurovascular compromise.

Keywords: Chiari malformation Type I, Spinal Accessory nerve, Spinal accessory neuralgia

INTRODUCTION

The spinal accessory nerve (CNXI) is traditionally considered a motor nerve. However, the extent of its' sensory components has been underestimated.^[8] Here, we report a patient with a Chiari Type I malformation, whose unilateral posterior interscapular pain was ultimately attributed to an atypical XI nerve neuralgic pain.

CASE REPORT

A 27-year-old male presented with a left-sided chest, shoulder, suprascapular/interscapular pain, and burning paresthesia in the left upper extremity of 3 months' duration. On examination, the only neurological deficit was 50% hypoesthesia involving the left C5 to D2 distributions (i.e., pain and temperature). The preoperative cervical MRI T2-weighted study showed a Type I Chiari malformation and syrinx (i.e., cerebellar tonsillar herniation up to the C1 level, diffusely dilated central canal from C5 to D3, and nonenhancing hyperintensity, present in the upper cervical cord [Figure 1]).

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Figure 1: MRI cervical spine T2-weighted sagittal image.

Surgery

The patient underwent a posterior fossa decompression consisting of a suboccipital craniectomy, C1 laminectomy, and “Y” shape dural incision. Once the dura was opened, the tonsils were found to be herniated into the spinal canal up to the C1 level; arachnoidal adhesions between the tonsils and cord were removed. Further, at the level of the foramen magnum, the left XI cranial nerve was severely compressed between the left-sided herniated tonsil and posterior inferior cerebellar artery (PICA) [Figure 2]. After subpial resection of the left tonsil [Figure 3], a muscle patch was interposed between the XI cranial nerve and PICA. This was followed by an augmentation duraplasty using a fascia graft and routine closure.

Postoperative recovery

Postoperatively, the left interscapular pain reduced significantly with immediate effect; however, the left upper extremity burning paresthesia took 2 months to subside, while the sensory hypoesthesia persisted even after 18 months.

DISCUSSION

Chiari I malformation is characterized by herniation of the cerebellar tonsils or PICA loops below the foramen magnum.

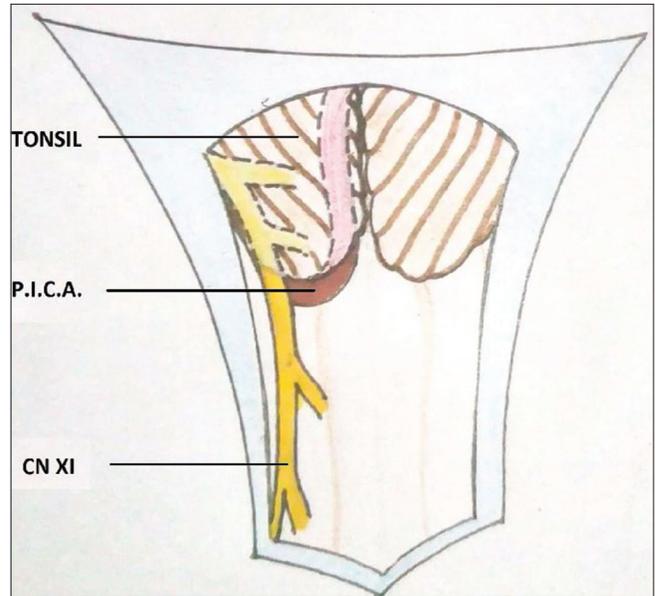


Figure 2: Diagrammatic representation of the first view after dural opening during surgery. PICA: Posterior inferior cerebellar artery, CNXI: Spinal accessory nerve.

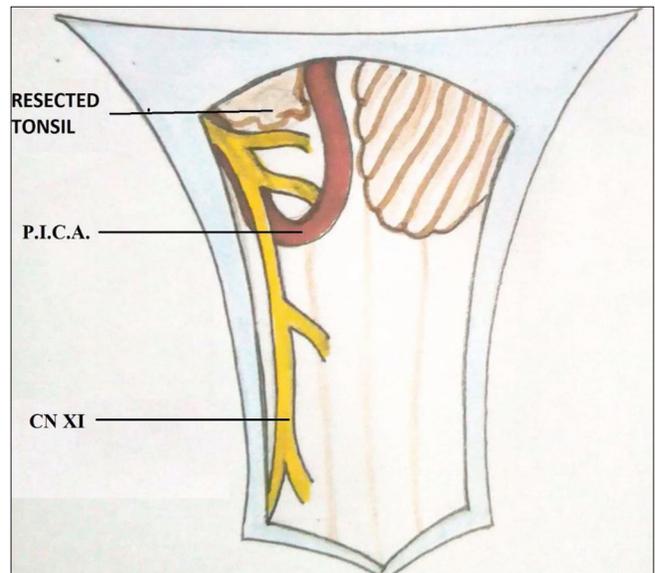


Figure 3: Diagrammatic representation of view after subpial resection of the left tonsil showing CNXI running just above the PICA, PICA: Posterior inferior cerebellar artery, CNXI: Spinal accessory nerve.

Neurological complaints may include; spastic paraparesis, amyotrophy, dissociated sensory loss, cerebellar signs, and lower cranial nerve palsies.^[2,4]

Spinal accessory nerve (XI) anatomy

The spinal accessory nerve (CNXI) includes two parts; a smaller cranial and a major spinal component. The cranial

division emerges from the medulla oblongata. The fibers of the spinal portion arise from special visceral efferent motor cells in the spinal accessory nucleus from C2 to C6. The spinal part leaves the anterior surface of the spinal cord between the dorsal and ventral sulci; after fusion of rootlets, the nerve travels cranially [Figure 4].

CNXI nociceptive fibers

Although the literature shows that CNXI is predominantly a motor nerve and conveys proprioceptive information only, there is pathological evidence that CNXI also contains nociceptive sensory components.^[6] Bremner showed that CNXI contained a high percentage of small (<2 μm) unmyelinated fibers, which are compatible with dorsal root C fibers and a small percentage of 2–3 μm myelinated fibers which represent A δ fibers. Both of these fibers are known to transmit pain sensation.^[1] Wetmore demonstrated clusters of sensory neurons and microganglion with nociceptive sensory functions involving the muscles supplied by the spinal accessory nerve in rats.^[5] Williams *et al.* further demonstrated pain as the most common (67%) presenting symptom in patients with spinal accessory nerve injury.^[7]

Anatomy of the PICA

The tonsillomedullary segment of PICA passes posterior to lower cranial nerves. At the craniovertebral junction, PICA frequently has a tortuous course with complex loops, and the relationship between the tonsillomedullary segment of PICA, tonsils, and the foramen magnum vary; this creates a potential site for an anatomical anomaly (i.e., ACM Type 1 can cause vascular compression syndrome).^[3]

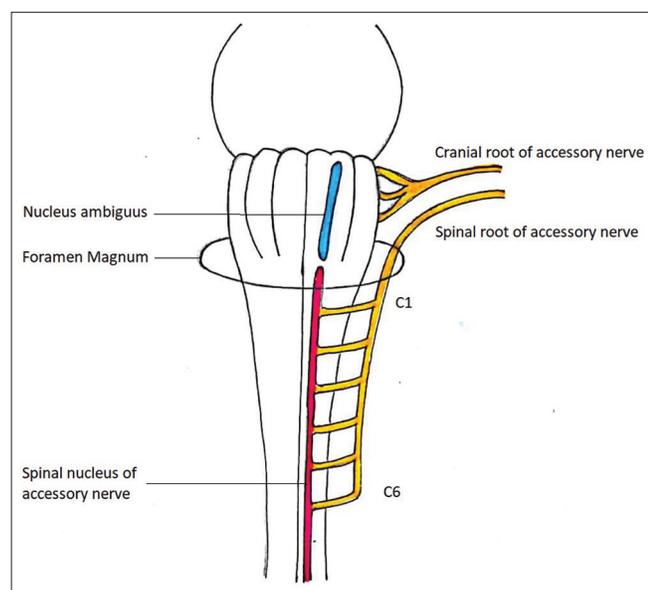


Figure 4: Origin and formation of spinal accessory nerve.

Syringomyelia

Conventionally, nuchal and scapular pain in ACM patients is attributed to associated syringomyelia. Still, showing the presence of nociceptive fibers in CNXI, we have raised the question of whether atypical XI cranial nerve neuralgic pain is actually responsible for these symptoms.

CONCLUSION

Posterior fossa decompression along with subpial resection of the left-sided cerebellar tonsil and microvascular decompression of the left cranial nerve XI resolved this patient's interscapular pain/dysesthesias/atypical XI nerve neuralgia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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