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Cruciate bibrachial diplegia due to an acutely trapped fourth ventricle

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Abstract

A 20-year-old female presented to us with bibrachial diplegia and dysarthria. She had an earlier history of craniosynostosis, multiple cranial surgeries and recent meningitis followed by ventriculoperitoneal shunting. Her symptoms started with a cruciate paralysis followed by rapid descending quadriparesis. Imaging revealed a trapped fourth ventricle as the cause of her descending paralysis.

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Case presentation

A 20-year-old female presented with progressive bilateral upper limb weakness over 3 days. She was a known case of Crouzon syndrome with craniosynostosis (turricephaly with bicoronal/basal sutural stenosis) and mobile atlantoaxial dislocation. She had undergone posterior C1–C2 Brook's fusion 12 years earlier. One year earlier (2017), she underwent craniofacial reconstruction and an anterior cranial fossa floor repair for cerebrospinal fluid rhinorrhoea. She had developed bacterial meningitis 2 months earlier. Head CT showed communicating hydrocephalus and a left frontal porencephalic cyst (Figure 1). After resolution of meningitis, a medium pressure ventriculoperitoneal shunt was placed. She had started developing neck pain 10 days earlier.

On evaluation she was conscious, disoriented and dysarthric with a slow and spastic tongue. She had grade 0/5 power in the upper limbs (distal power was slightly more than proximal power) and grade 4/5 power in the legs. Sensory examination was normal. Deep tendon reflexes were absent in all four limbs. Plantar reflexes and abdominal reflexes were mute. Nerve conduction studies showed prolonged F-wave latencies from the median and ulnar nerves. Other parameters were normal. The possibilities considered were transverse myelitis or a descending variant of Guillain-Barré syndrome. Brain and spine MRI showed an enlarged fourth ventricle [trapped fourth ventricle (TFV)] that had tautly stretched the medullary–cervical segment like a bowstring at the craniovertebral junction. Compared to the CT scan images performed 2 months earlier, the fourth ventricle had dilated tremendously, and the sagittal anteroposterior diameter of the fourth ventricle had enlarged from 2.57 to 3.43 cm (an increase of over 130%) (Figure 1). This had resulted in medullary and upper cervical cord compression over the odontoid process of the axis. Secondarily it had produced a long segment congestive upper cervical cord oedema extending from the upper cervical cord to C7 segment (Figure 1). The diagnosis of a progressive craniovertebral junction compression syndrome, commencing as cruciate diplegia was made at this point. Surgical posterior fossa decompression with a view to depressurise the trapped ventricle and relieve the upper cervical cord compression was considered. In view of the prior Brook's fusion and the complexity of surgery, it was planned for the next day. However, over the next 12 hours, her weakness progressed rapidly. She developed descending weakness of both legs until she was finally quadriparetic (grade 0/5 Medical Research Council scale). She also developed type II respiratory failure and had to be mechanically ventilated. Over the next 24 hours, she lost all brainstem reflexes and expired.

Discussion

The corticospinal tract (CST) is the main tract that regulates fine motor movements. It originates from the cerebral cortex and passes through the internal capsule and brainstem until it reaches the lower medulla where it decussates. Conventionally, the upper limb CST fibres are thought to decussate just proximal to the foramen magnum, whereas the lower limb CST fibres decussate one cord segment below this in the superior cervical cord.[1] The decussated fibres form the lateral CST. This tract now descends ipsilaterally until the sacral cord, synapsing along the way with segmental anterior horn cells and controlling fine movements of distal extremities. Nearly 90% of projections from the motor cortex follow the lateral CST.

Approximately 10–15% of CST fibres do not decussate at the medulla but descend ipsilaterally as the ventral (anterior) CST. Instead, these fibres crossover lower down at the spinal

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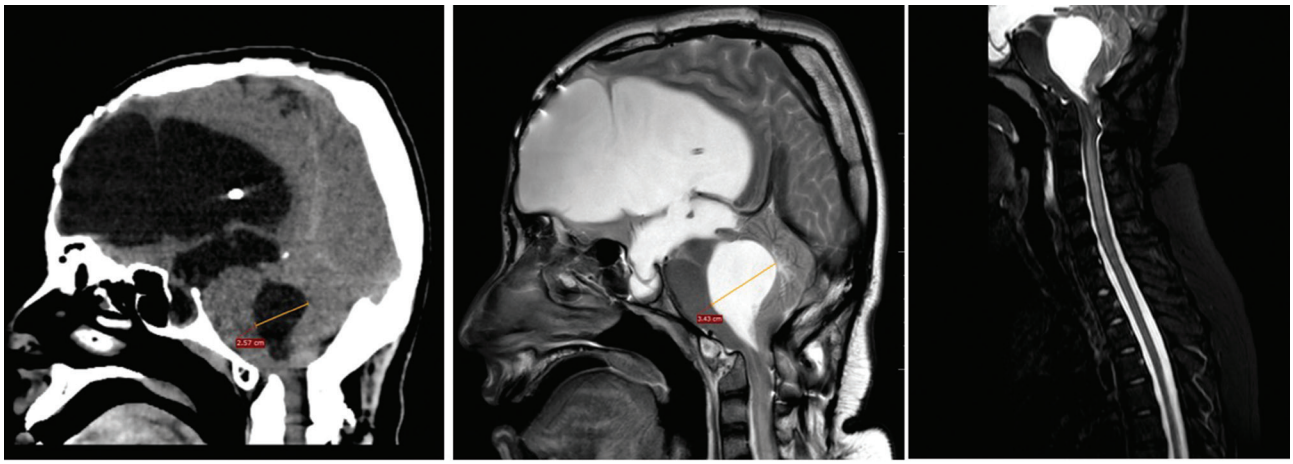
Figure 1 (a) CT sagittal image shows an enlarged trapped fourth ventricle with an anteroposterior diameter of 2.57 cm. (b) Sagittal brain MRI shows a larger trapped fourth ventricle with cervicomedullary compression at the craniovertebral junction (anteroposterior diameter, 3.43 cm). (c) Sagittal spine MRI shows a long segment cervical cord hyperintensity and oedema



cord segments that innervate the proximal muscles of the limbs, sometimes with bilateral projections (Figure 2).

In 1901 Wallenberg described 'hemiplegia cruciata' (with ipsilateral arm and contralateral leg weakness).[2] He postulated that a lateral lesion at the level of the decussation of the pyramid in the lower medulla impinged on the crossed arm and uncrossed leg fibres of the CST resulting in ipsilateral arm and contralateral leg weakness (Figure 3).

In 1970, Bell distinguished a distinct 'cruciate paralysis' [Bell's cruciate paralysis (BCP)] in three patients. BCP was characterised by bibrachial diplegia (both upper limbs) without lower limb weakness.[3] Bell also attributed BCP to differential CST decussation with selective damage of only the decussating arm fibres at the caudal medulla.

In the same paper, interestingly, Bell also described a largely forgotten fourth patient with paraplegia (both legs, sparing the arms) due to injury to the pyramidal decussation at the C1 cord level, which is probably better referred to as an upper cervical cord paraplegia (Figure 3). Nevertheless he did issue a caveat that 'a tiny variation in the position of the pyramidal decussation relative to the tip of the odontoid process or anterior rim of the foramen magnum could result in either cruciate paralysis, paraplegia or quadriplegia'.

Figure 2 Graphic representation of the medullary–cervical junction and the arrangement of various fibres of the corticospinal tract

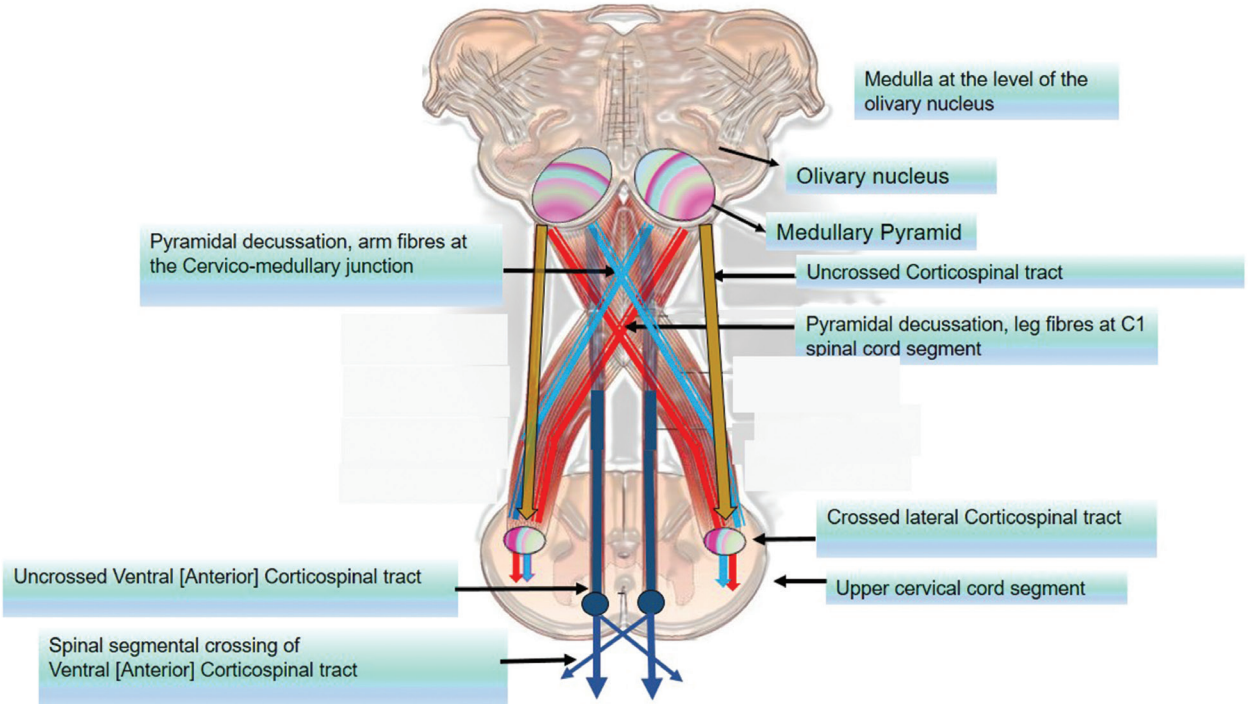
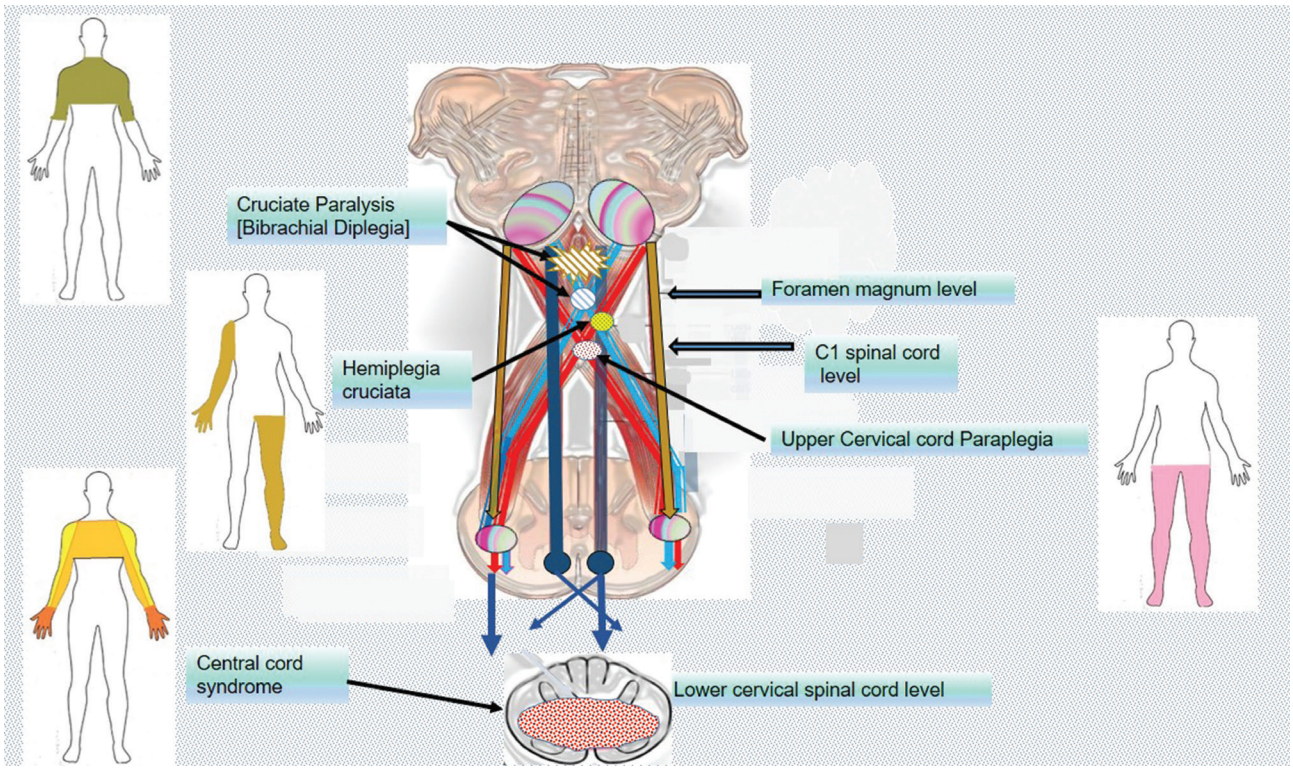


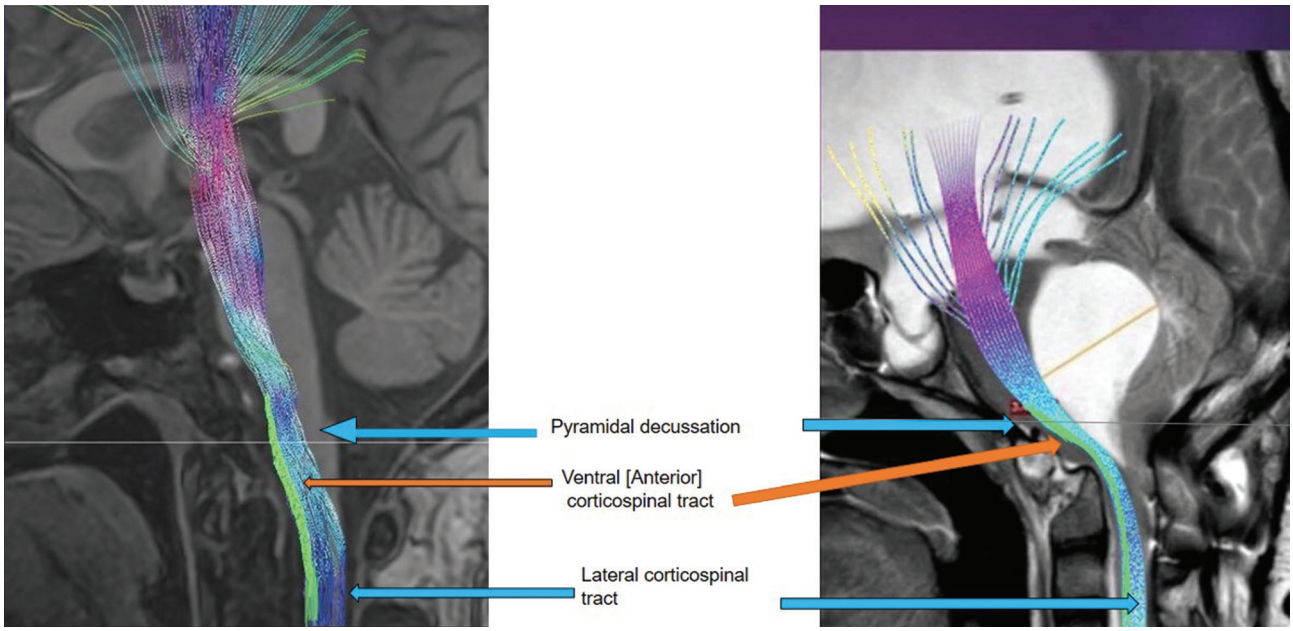
Figure 3 Four different brainstem/spinal cord syndromes occurring at the medullary–cervical segment and their various anatomical localisations. The yellow hatched superior lesion is the contemporaneous explanation for cruciate diplegia, whereas the blue hatched oval lesion is Bell's purported site of lesioning



Central cord syndrome (CCS) can mimic BCP. However, CCS produces disproportionate lower motor neuron weakness of the upper limbs, sensory impairment in a cape-like distribution and variable upper motor neuron weakness in the lower limbs. Whereas, transient respiratory insufficiency or lower cranial nerve palsy are more common with BCP.[3] Moreover, CCS occurs at a lower level in the cervical cord and involves the medial fibres of the lateral CST along with the anterior horn cells and the central grey matter of the cord. CCS often occurs with cervical hyperextension injuries and cervical canal stenosis.[4] These closely related brainstem/spinal cord syndromes are detailed in Figure 3.

BCP can mimic a descending variant of Guillain-Barré syndrome. Our patient did have electrophysiological findings compatible with an early demyelinating neuropathy (absent F-waves in the upper

Figure 4 Sagittal MRI tractography of a normal brain (left panel) and superimposed tractography of our patient (right panel) showing the predilection of the ventral corticospinal tract to be compressed



limbs). However, the prolongation of upper limb F-waves likely reflected an acute upper cervical cord injury or a spinal shock.[5]

Differential decussation of the CST was an apodictic concept until Pappas et al. refuted this in primates.[6] Consequently other theories were invoked to explain this phenomenon. Compression of the CST, which is phylogenetically more important for upper limb function than for leg function, would preferentially involve the arms more than the legs. Another possibility is that the ventral CST lies anterior to the lateral CST and compression against the bony odontoid would result in early injury to this tract. The ventral CST preferentially innervates the proximal upper limb muscles rather than the lower limb. Hence BCP would be an early manifestation before progressive compression led to total quadriparesis, as in our patient (Figure 4).

Furthermore, there are collateral projections of CST fibres to the reticular nuclei, other brainstem, dorsal column nuclei and the central grey matter of the cervical cord. Reticulospinal fibres in particular may play a part in controlling proximal limb movements.[7] Damage to these collateral fibres from the CST might also contribute to BCP.[8]

The vast majority of BCP occurs after trauma (80%), although foramen magnum lesions, such as tumours, aneurysms or other lesions, are also implicated.[9]

Our patient had a TFV with periventricular interstitial oedema and cervical cord oedema. Compared to the prior CT 2 months earlier, the TFV enlarged by approximately 138%. In conjunction with the herniating cerebellar tonsil posteriorly, the TFV would have caused increasing compression of the CST in a rostrocaudal direction starting near the odontoid and implicating the ventral CST before affecting the lateral CST. TFV refers to an obstructed fourth ventricle when cerebral spinal fluid shunting of the lateral ventricles fails to adequately relieve the obstruction.[10] TFV is rare and presents with cranial nerve palsies or alteration of sensorium. It can also result in a descending transtentorial herniation. Management of a TFV is complex and includes shunting, endoscopic, ventriculoscopic or open surgical approaches. Nonsurgical management is used only in clinically and radiologically stable patients. Our case presented with BCP as a very unusual manifestation of an expanding TFV. Thus an acute TFV causing a brainstem/spinal cord syndrome should be considered in the presence of bibrachial cruciate diplegia as it is amenable to surgical correction.

Informed consent

Written informed consent for the paper to be published (including images, case history and data) was obtained from the patient for publication of this paper, including accompanying images.

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