A case of Collet-Sicard syndrome associated with traumatic atlas fractures and congenital basilar invagination

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SHORT REPORT

An 18 year old man with congenital basilar invagination developed multiple lower cranial nerve (CN) palsies including CN IX to XII after a traffic accident. Computed tomography of his skull base revealed a two part atlas Jefferson fracture. Normally, lower cranial nerves (CN IX–XII) pass through a space between the transverse process of the atlas medially and the styloid process laterally. Zielinski et al identified that there was only 8–10 mm of space between CN IX to XII after a traffic accident. Computed tomography can be used to investigate the presence of an atlas fracture and can be performed to confirm that the space is greater than 8–10 mm in order to rule out a basilar invagination.

DISCUSSION

Multiple CN palsies are often a diagnostic challenge because the nerves can be affected at any site along their course. CN IX–XI exit the cranial vault through the jugular foramen, whereas CN XII leaves from the hypoglossal canal close to the occipital condyle. Near their points of exit, CN IX–XII lie between the transverse process of the atlas medially and the stylid process of the skull laterally. Zielinski et al identified the mechanism by which atlas fractures could produce cranial nerve symptoms. Through their study of cadavers, they found that there was only 8–10 mm of space between the transverse process of the atlas and the styloid process. CN IX–XII pass through this narrow space (fig 2A). If the atlas were abnormally displaced laterally after a Jefferson fracture, this space would be reduced to nearly nothing and lower CN palsies would be likely to develop.

Fractures of the atlas are rare and comprise only 2% of all spinal injuries. The mechanisms of injury include motor vehicle accidents, falls, pedestrian impacts, hang-gliding, skateboarding, diving injuries, waterskiing accidents, and an equestrian accident. Atlas fractures usually occur as a result of axial compression loads, which result in the transmission of forces through the occiput to the cervical spine via the lateral masses of C1. The forceful separation of the lateral masses of C1 also causes the occipital condyle to be forced further away from the atlas. This increased distance between the atlas and the occiput is thought to be the cause of the CN palsies. If there was no fracture of the atlas, the CN IX–XII would not pass through the space between the transverse process of the atlas medially and the styloid process laterally.

Abbreviations: CN, cranial nerves; MRI, magnetic resonance imaging
atlas fracture. MRI of the cervical spine may facilitate the assessment of the surrounding soft tissue and the integrity of the transverse ligament.17 Tears of the transverse ligament appear as a loss of anatomical continuity of the ligament containing regions of high signal intensity compared with the homogeneous low signal intensity of the normal ligament on gradient echo MRI.18 Treatment should be aimed at stabilisation of the symptoms and prevention of further neurological damage.

Basilar invagination is a developmental defect implying protrusion of the vertebral column into the skull base. Basilar invagination might be associated with Chiari malformations, syringobulbia, and syringomyelia. The prevalence of neurodysgenesis is between 25% and 30% in basilar invagination. The neurological deficits are from direct compression of the neural tissue by bone and soft tissue at the craniovertebral junction, or a compromise of the vertebral anterior spinal and perforating arteries to the cervicomedullary junction. Minor trauma may set off a pattern of symptoms and signs that progress at a speedy pace.19 In basilar invagination, the odontoid process is above Chamberlain’s line, which joins the hard palate to the opisthion on radiological images. A minor degree of basilar invagination may produce no neurological symptoms. However, combined with an atlas fracture, the lower CN are vulnerable to be stretched and compressed because of both upward indentation of the skull base and lateral displacement of the atlas lateral mass (fig 2B).

When a subject has Collet-Sicard syndrome after a traumatic injury, we should take atlas fractures associated with basilar invagination into account. Other cervical spine injuries (for example, Hangman fracture or cervical spine subluxation/dislocation) have also been reported to result in CN palsies. This report highlighted the importance of neurological examinations and the anatomical relation of the posterior fossa, skull base, and upper cervical spine.
REFERENCES

NEURONLINE

We move: www.wemove.org

This snappily named site claims to be the internet’s most comprehensive resource for movement disorder information and the hub of movement disorder activities on the web. It is certainly a very well presented and useful site. There are a number of free services on offer. There is a movement disorder research news email update service (E-MOVE), links to other, mainly American, sites and support groups, both a chat room and forum (for which you have to register), and a link to movement disorder virtual university (MDVU). This is a closely related site that has an even snappier appearance than the original We move site but actually contains much of the same information under the topic headings. On the We move site these are listed on 21 buttons down the left edge. There is an interesting mix of the expected diseases, such as Parkinson’s; general, non-disease specific problems, such as tremor (which then covers paediatric tremor); and other slightly unexpected topics such as spasticity, Huntington’s disease, and Rett syndrome. The information on each topic is well organised and reasonably current. The site is funded by a long list of pharmaceutical companies and the treatment section of each subject is peppered with their branded products in addition to the generic names. The American parentage of the site is also reflected in the suggested use of drugs unavailable in the UK, such as tolcapone. Strangely, considering its title and subject, what the site lacks is movement in the form of video clips. There is an animated clip of how botulinum toxin works but this took long enough, across a broadband connection, for me to make a cup of tea before it started. Overall though, it is a great place to start on the web for movement disorders information.

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