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

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CASE 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 styloid process and the atlas transverse process. Atlas burst fractures rarely cause neurological deficits because of a greater transverse and sagittal diameter of the spinal canal at the atlas, and a tendency of the lateral masses to slide away from the cord after injury. However, when associated with a rare condition—congenital basilar invagination—atlases fractures can compromise the space and make CN IX–XII more vulnerable to compression injury. This report discusses the correlation between the anatomical lesions and clinical features of this patient.

Collet-Sicard syndrome refers to unilateral lesions of cranial nerves (CN) IX, X, XI, and XII.1, 2 This rare syndrome has been attributed to tumours of the skull base, coiling and dissections of the internal carotid artery, multiple myeloma, vasculitis, carotid fibromuscular dysplasia, shotgun injuries, idiopathic cranial polynuropathy, atlas fractures, and occipital condyle fractures.2, 3 There is only one reported case of atlas Jefferson fracture causing Collet-Sicard syndrome in the English language literature of the Medline database from 1966 to 2003.1, 3 Jefferson fractures are bursting injuries of the atlas, which rarely cause neurological deficits. This has been suggested to be a result of a greater transverse and sagittal diameter of the spinal canal at the first cervical spine, in addition to the mechanism of the lateral masses sliding away from the cord. Any neurological sequelae could be attributed to spinal cord damage or vertebral artery compromise rather than cranial nerve injury. The classic presentations of atlas fractures include neck pain, cervical muscle spasm, reduced range of neck motion, and head tilt. Compression of the greater occipital nerve causes suboccipital paresthesia or pain.3 We review the literature and will briefly discuss the probable mechanisms contributing to Collet-Sicard syndrome.

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 stylloid process of the skull laterally. Zielinski et al identified the mechanism by which atlas fractures could produce cranial nerve symptoms.9 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. CT demonstrates basilar invagination and rupture of the transverse ligament, without other anomalies such as Arnold-Chiari malformations, syringomyelia, syringobulbia, or hydrocephalus (fig 1B, C). He underwent external immobilisation with neck collar and reduction by traction. The residual sequela was slurred speech after an 18 month duration of follow up.

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 stylloid process of the skull laterally. Zielinski et al identified the mechanism by which atlas fractures could produce cranial nerve symptoms.9 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 stylloid process. CT demonstrates basilar invagination and rupture of the transverse ligament, without other anomalies such as Arnold-Chiari malformations, syringomyelia, syringobulbia, or hydrocephalus (fig 1B, C). He underwent external immobilisation with neck collar and reduction by traction. The residual sequela was slurred speech after an 18 month duration of follow up.

Fractures of the atlas are rare and comprise only 2% of all spinal injuries.10 The mechanisms of injury include motor vehicle accidents, falls, pedestrian impacts, hang-gliding, skateboarding, diving injuries, waterskiing accidents, and an equestrian accident.11–13 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 places the transverse ligaments at risk for rupture or avulsion, which could result in atlantoaxial instability.14 Plain radiography and computed tomography can be used to investigate the presence of an

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. 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. 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. 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.

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Competing interests: none declared

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Received 21 July 2003
In revised form 16 September 2003
Accepted 1 October 2003
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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,
amongst American, sites and support
groups, both a chat room and forum
(for which you have to register), and a
link to movement disorder virtual uni-
versity (MDVU). This is a closely related
site that has an even snappier appear-
ance 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 infor-
mation 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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doi: 10.1136/jnnp.2003.012963