Cluster headache-like attack as an opening symptom of a unilateral infarction of the cervical cord: Persistent anaesthesia and dysesthesia to cold stimuli

Vincent de la Sayette, Stéphane Schaeffer, Oguzhan Coskun, François Leproux, Gilles Defer

Abstract
A 54 year old man experienced excruciating left retro-orbital pain with lacrimation and redness of the eye representative of a cluster headache attack. This was followed by left hemiparesis with plegia of the lower limb and left Horner’s syndrome. Five days later the hemiparesis recovered while the patient developed hypoaesthesia to cold stimuli that evoked painful burning dysesthesia on the right side below the C4 level. MRI disclosed a discrete infarct in the left lateral aspect of the cord at C2 level concomitant to a left vertebral artery thrombosis. This limited infarct and the clinical symptoms suggest a hypoperfusion in the peripheral arterial system of the left hemicord, supplied both by the anterior and posterior spinal arteries. Cluster headache-like attack and persistent dysesthesia to cold stimuli are discussed respectively in view of the central sympathetic involvement and partial thalamic system dysfunction.

Keywords: hemicord infarction; cluster headache attack; dysesthesia; allodynia

The cervical cord is supplied by two arterial systems, central and peripheral, which overlap but are discrete. The first is dependent entirely on the single anterior spinal artery (ASA). The second, without clear cut boundaries, receives supplies from the ASA and both posterior spinal arteries. Because the ASA is medial and dominant, unilateral cord infarctions are very rare. They may occur in the perfusion territory supplied by the ASA as a result of the obstruction of either a duplicated ASA or the obstruction of one of the sulcal arteries which arise from the ASA and turn alternatively left or right, to supply one side of the central cord. Peripheral hemicord infarction may result from ischaemia in the territory of the ASA or posterior spinal artery. We report on a patient with a unilateral infarction limited to the left lateral column of the cervical cord in relation to a vertebral artery thrombosis. The clinical feature was peculiar in that the cluster headache-like attack presented as the opening symptom and, thereafter, the occurrence of anaesthesia and dysesthesia to cold stimuli (allodynia) that affected the right side below C4 became apparent.

Case report
A 54 year old man with a history of mild hypertension (treated with 50 mg Iosartan/day) and common migraine without known autonomic feature presented on 13 October 1997 with an abrupt boring and throbbing pain in the left retro-orbital region spreading to the jaw, and the supraorbital, and temporal regions. There were photophobia, abundant lacrimation, and redness of the left eye. The excruciating pain lasted 30 minutes, then improved and disappeared about 20 hours later. Half an hour after onset of the headache, the patient experienced paraesthesia of the left lower limb which progressed, within 3 hours, to weakness of the left foot. The next days, a left hemiparesis occurred. The patient was admitted to our department on 15 October 1997.

His arterial pressure was initially 160/100 mm Hg and general examination showed no abnormalities. Neurological examination disclosed a left Horner’s syndrome and a flaccid left sided hemiparesis affecting the lower limb severely and partially the proximal part of the upper limb. The hand was slightly weak and the face was spared. Bilateral Babinski’s signs were present. Light touch, vibration, and sense of position were normal. Pinprick and cold and warm thermal stimuli were perfectly discriminated on both sides although felt appreciably more strongly on the left. There was equal facial sensation to light touch, pain, and temperature, including symmetric corneal response, despite the patient having reported persistent soreness and left hypersensitivity of the face and head to touch since the initial attack of retro-orbital pain. Five days after onset, the hemiparesis cleared almost completely while the patient experienced unpleasant and painful right sided provoked dysesthe-
At that time, examination of the patient’s sensibility disclosed normal and symmetric sensations to light touch, vibration, sense of position, and pin prick. Temperature perception and discrimination were normal above C4. Below C4, on the right side, the thermal perception for warm stimuli (>28°C) was unchanged (quickly and strongly felt on the left side) and contrasted with more severe hypoaesthesia (upper limb) and anaesthesia (trunk and lower limb) to cold stimuli (<28°C). Moreover, cold stimuli (<28°C) evoked severe dysesthesia described as burning, aching sensations radiating around the place of stimulation. Any stimulus other than cold could elicit them.

Normal investigations included routine laboratory examinations, CT of the head, transoesophageal echocardiography, and Holter monitoring. Brain MRI was unremarkable except for a left punctiform low void vertebral artery. MRI of the cervical spine, 11 days after the onset, disclosed a spontaneous abnormally increased signal on T2 and proton density weighted images in the lateral aspect of the left cord at C2 level (figure). Vertebral angiography showed a thrombosis of the left vertebral artery. A diagnosis of infarction in the left upper cervical hemicord was made. The patient was treated with heparin and subsequently fluindione. Four months later his motor strength fully recovered. A left Babinski’s sign and Horner’s syndrome persisted. The patient continued to complain of left soreness and hypersensitivity of the face and head and, above all, of a bitter, burning dysaesthesia to cold stimuli, refractory to carbamazepine, clonazepam, and amitriptyline.

Comment
The combination of neurological signs, left flaccid hemiparesis sparing the face with plegia of the lower limb, Horner’s syndrome, and slight spinothalamic hypoaesthesia on the contralateral side, suggests a lesion located in the periphery of the left lateral column of the cervical cord. A T2 weighted cervical MRI disclosed a focal high intensity signal within the left lateral aspect of the cord, at C2 level, consistent with a limited infarct of the hemicord. Angiography documented further a left vertebral artery thrombosis that could correspond, although without certainty, to a dissection. Our
patient shared the main clinical features found in the ASA hemicord syndrome—namely, Horner’s syndrome and hemiparesis that progress over many hours on the side ipsilateral to the infarction while a spinothalamic hypoesthesia develops on the contralateral side. None of these patients in the above mentioned studies had either posterior column loss or facial hypoesthesia, both of which are relevant to posterior spinal artery syndrome. Even though our patient is similar in many aspects to others, two particular points have to be emphasised: (1) the initial excruciating retroorbital pain; (2) the limited anaesthesia and persistent dysaesthesia to cold stimuli which developed soon after the onset.

All the reported cases of cervical hemicord infarction began with severe pain that involved predominantly the neck and often radiated to the shoulder or the upper limb. None of them presented with headache. On the contrary our patient never had cervicobrachial pain but developed extremely severe unilateral headache as an opening symptom. It could account for pain from arterial dissection but, in this outlook, rather from a carotid than a vertebral artery dissection. In return, the excruciating boring and throbbing pain in the left retroorbital region, associated with autonomic features, lacrimation, and conjunctival injection, satisfied principally the criteria of cluster headache attack. Nevertheless, as our patient experienced only a single episode of retroorbital pain which contrasted with the striking rhythmicity of typical cluster headache attack, the generic terminology of trigeminal-autonomic cephalalgia (TAC) seems more appropriate. This kind of intense short lasting headache with autonomic features including cluster headache attack, chronic paroxysmal hemicrania, episodic paroxysmal hemicrania, and SUNCT syndrome (short lasting unilateral headache attack). It might be postulated that dysaesthesia may result from selective lesions of the spinothalamic system as touch and pinprick were normally felt contrasting with hypoesthesia to thermal stimuli and above all anaesthesia (trunk and lower limb) to cold stimuli. Thus considering dysaesthesia in our patient in terms of imbalance within the spinothalamic system itself seems an attractive hypothesis. Interestingly dysaesthesia occurred while the patient recovered from hemiparesis and hypoesthesia to pinprick and developed anaesthesia to cold stimuli. This response (dysaesthesia) suggests a functional rearrangement so that the cold stimuli could raise an inappropriate activation of pain message instead of cold thermic sensation. In this way, Triggs and Beric considered such an alternative explanation as did Nathan and Smith, who suggested that dysaesthesia may result from selective lesions of the spinothalamic tract sparing the spinothalamic connections.

Considering the anatomical organisation of the two spinothalamic pathways (anterior and posterior tracts) and the topographical arrangement of the fibres that they convey (crude tactile, pain, and thermal sensations), clinical symptoms suggest, in our patient, lesioning in the most dorsal and superficial part of the spinothalamic tract. This accords well with the initial hemiparesis with plegia of the lower limb which, in turn, also indicate (considering the somatotopic arrangement of the fibres within the corticospinal tract) a superficial lesion of the lateral cord. MRI confirmed the small size of the lesion, which was
confined to the lateral fasciculus and limited in height. The deep extension could correspond to oedema rather than to ischaemia. This suggests a hypoperfusion in a "watershed area" between both anterior and posterior spinal peripheral arterial systems rather than ischaemia in the perfusion territory of a sulcal artery or a duplicated ASA. This hypothesis agrees with the common origin of the anterior and posterior spinal arteries which both arise from the vertebral artery.


Cluster headache-like attack as an opening symptom of a unilateral infarction of the cervical cord: Persistent anaesthesia and dysaesthesia to cold stimuli

Vincent de la Sayette, Stéphane Schaeffer, Oguzhan Coskun, François Leproux and Gilles Defer

*J Neurol Neurosurg Psychiatry* 1999 66: 397-400
doi: 10.1136/jnnp.66.3.397

Updated information and services can be found at:
[http://jnnp.bmj.com/content/66/3/397](http://jnnp.bmj.com/content/66/3/397)

These include:

**References**

This article cites 15 articles, 6 of which you can access for free at:
[http://jnnp.bmj.com/content/66/3/397#BIBL](http://jnnp.bmj.com/content/66/3/397#BIBL)

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Topic Collections**

Articles on similar topics can be found in the following collections

- Headache (including migraine) (459)
- Pain (neurology) (763)
- Drugs: CNS (not psychiatric) (1945)
- Ophthalmology (842)

**Notes**

To request permissions go to:
[http://group.bmj.com/group/rights-licensing/permissions](http://group.bmj.com/group/rights-licensing/permissions)

To order reprints go to:
[http://journals.bmj.com/cgi/reprintform](http://journals.bmj.com/cgi/reprintform)

To subscribe to BMJ go to:
[http://group.bmj.com/subscribe/](http://group.bmj.com/subscribe/)