

A Topographical Anatomy of False-Localising Signs

One hundred years ago, Dr James Collier published a paper in *Brain* entitled *The false localising signs of intracranial tumour*. Based on his experience of 161 clinically and pathologically examined cases of intracranial tumour seen at the National Hospital, Queen Square, London, he observed false-localising signs in 20 (12.4%).¹ The term was coined to indicate clinically observed signs that violated the expected clinico-anatomical concordance on which clinical examination is predicated.²

Since 1904, many examples of false-localising signs have been described. They may occur in the clinical context of raised intracranial pressure (RICP) which is symptomatic of intracranial pathology (tumour, haematoma, abscess) or idiopathic (idiopathic intracranial hypertension: IIH), and with spinal cord lesions. Associated lesions may be intra- or extraparenchymal. The course of the associated disease may be acute (cerebral haematoma) or chronic (IIH, tumour).³

The pathogenesis of false-localising signs remains uncertain, but their importance from a clinical standpoint is not in doubt, since they may lead to inappropriate imaging and even interventions on the wrong side (although the risk of such errors of commission is less now that neuroimaging is widely available). This article gives a brief topographical overview of false-localising signs.

Motor system

Kernohan's notch syndrome: false-localising hemiparesis

A supratentorial lesion, such as acute subdural haematoma, may cause transtentorial herniation of the temporal lobe, with compression of the ipsilateral cerebral peduncle against the tentorial edge; since this is above the pyramidal decussation a contralateral hemiparesis results. Occasionally, however, the hemiparesis may be ipsilateral to the lesion, and hence false-localising; this occurs when the contralateral cerebral peduncle is compressed by the free edge of the tentorium. This is the Kernohan-Woltman notch phenomenon, or Kernohan's notch syndrome.⁴ There may be concurrent homolateral third nerve palsy, ipsilateral to the causative lesion.⁵

Cerebellar syndrome

Frontocerebellar pathway damage, for example as a result of infarction in the territory of the anterior cerebral artery, may result in incoordination of the contralateral limbs, mimicking cerebellar dysfunction. Suboccipital exploration to search for cerebellar tumours based on these clinical findings was known to occur before the advent of brain imaging.⁶

Brainstem compression: false-localising diaphragm paralysis

Hemidiaphragmatic paralysis with ipsilateral brainstem (medullary) compression by an aberrant vertebral artery has been described, in the absence of pathology localised to the C3-C5 segments of the spinal cord where phrenic motor neurones originate, hence a false-localising sign.⁷

Cranial nerves

Oculomotor nerve

Unilateral fixed dilated pupil (Hutchinson's pupil) may occur with an ipsilateral intracranial lesion such as an intracerebral haemorrhage, due to transtentorial herniation of the brain compressing the oculomotor nerve against the free edge of the tentorium. Because of the fascicular organisation of fibres within the oculomotor nerve, the externally placed pupillomotor fibres are most vulnerable. Very occasionally, fixed dilated pupil may

occur contralateral, and hence false-localising, to intracranial pathology.⁸ The exact mechanism for this clinical observation is not currently known.

Divisional third nerve palsy is usually associated with lesions at the superior orbital fissure or anterior cavernous sinus, where the superior division of the oculomotor nerve passes to the superior rectus and levator palpebrae, and the inferior division to the medial and inferior recti and inferior oblique muscles. Divisional third nerve palsies may sometimes occur with more proximal lesions, presumably as a consequence of the topographic arrangement of the fascicles within the nerve, for example with intrinsic brainstem disease (e.g. stroke)⁹ or with pathology in the subarachnoid space where the nerve rootlets emerge from the brainstem (e.g. malignant infiltration).¹⁰

Trochlear nerve

False localising fourth nerve palsies, causing diplopia on downward and inward gaze, have occasionally been described in the context of IIH.^{11,12}

Trigeminal nerve

Trigeminal nerve hypofunction (trigeminal sensory neuropathy) or hyperfunction (trigeminal neuralgia) may on occasion be false-localising, for example in association with IIH¹³ or with contralateral pathology, often a tumour.¹⁴ For example, trigeminal neuralgia has been associated with a contralateral chronic calcified subdural haematoma which caused rotational displacement of the pons, with resolution after removal of the haematoma.¹⁵

Abducens nerve

Sixth nerve palsies are the most common false-localising sign of raised intracranial pressure. In one series of 101 cases of IIH, 14 cases were noted, 11 unilateral and 3 bilateral.¹⁶ Stretching of the nerve in its long intracranial course or compression against the petrous ligament or ridge of the petrous temporal bone have been suggested as the mechanism for false-localising sixth nerve palsy.³

Facial nerve

Lower motor neurone type facial weakness has been described in the context of IIH,¹⁷ sometimes occurring bilaterally to cause facial diplegia,¹⁸ usually with concurrent sixth nerve palsy or palsies. Hemifacial spasm has rarely been described with contralateral posterior fossa lesions.¹⁴

Vestibulocochlear nerve

Hearing loss has on occasion been reported as a complication of IIH,¹⁹ although the commonest otological complication of IIH is tinnitus.¹⁶



Andrew Larner is the editor of our Book Review Section. He is a Consultant Neurologist at the Walton Centre for Neurology and Neurosurgery in Liverpool, with a particular interest in dementia and cognitive disorders. He is also an Honorary Apothecaries' Lecturer in the History of Medicine at the University of Liverpool.

Correspondence to:

AJ Larner
Consultant Neurologist,
Walton Centre for Neurology
and Neurosurgery,
Lower Lane,
Fazakerley,
Liverpool,
L9 7LJ,
United Kingdom.
Fax: 0151 529 5513,
Email: a.larner@
thewaltoncentre.nhs.uk

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Multiple and lower cranial nerve involvement

Concurrent false-localising involvement of multiple cranial nerves has been noted on occasion, for example trigeminal, abducens and facial nerves with a contralateral acoustic neuroma,²⁰ and trigeminal, glossopharyngeal and vagus nerves with a contralateral laterally-placed posterior fossa meningioma.²¹

Spinal cord and roots

False localising signs within the spinal cord are well attested to. Broadly, these may be said to occur with lesions around the foramen magnum, or lower cervical/upper thoracic spinal cord.

Foramen magnum/upper cervical cord

Paraesthesia in the hands with intrinsic hand muscle wasting and distal upper limb areflexia, with or without long tract signs, suggestive of a lower cervical myelopathy may occur with lesions at the foramen magnum or upper cervical cord ("remote atrophy").²²

Lower cervical/upper thoracic cord

Compressive lower cervical or upper thoracic myelopathy may produce spastic paraplegia with a mid-thoracic sensory level (or "girdle sensation").^{23,24} For example, in one case a spastic paraplegia with a sensory level at T10 was associated with cervical compression from a herniated disc at C5/C6.²⁵

Radiculopathy

False-localising radiculopathy may occur in the context of IIH and cerebral venous sinus thrombosis, manifesting as acral paraesthesias, backache and radicular pain, and less often with motor deficits,²⁶ which on occasion may be suf-

ficiently extensive to mimic Guillain-Barré syndrome (flaccid-areflexic quadriplegia).²⁷ The postulated mechanism for such radiculopathy is mechanical root compression due to elevated CSF pressure.

Higher cognitive function

Hemineglect is much commoner with right rather than left parietal lobe lesions. An example of false-localising neglect has been encountered: in a patient with a posterior fossa meningioma causing left pontine compression, long tract signs and hydrocephalus, ipsilesional neglect was found, despite normal structural imaging of the cerebral hemispheres. The neglect resolved promptly after shunting and did not recur despite progressive brainstem compression (PC Nachev & IH Jenkins, personal communication).

Comment

As false-localising signs most often occur in the context of RICP, this seems likely to be the most important factor in the pathogenesis of these signs. Suggested mechanisms include mechanical distortion of cranial nerves with intracranial pathology and venous and/or arterial ischaemia with spinal cord pathology.³ It is worth remembering that RICP itself may be a false localising sign when associated with spinal tumours, even in the thoracolumbar region, perhaps related to elevated CSF protein concentration.²⁸

Of the various false-localising signs described, sixth nerve palsies are the most commonly observed. However, the possibility of false localisation should be borne in mind when any of the above-mentioned signs occur without obvious clinical-anatomical or clinical-radiological correlate.²

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