False localising signs

A J Larner

Neurological signs have been described as “false localising” if they reflect dysfunction distant or remote from the expected anatomical locus of pathology, hence challenging the traditional clinicopathological correlation paradigm on which neurological examination is based. False localising signs occur in two major contexts: as a consequence of raised intracranial pressure, and with spinal cord lesions. Cranial nerve palsies (especially sixth nerve palsy), hemiparesis, sensory features (such as truncal sensory levels), and muscle atrophy, may all occur as false localising signs. Awareness that signs may be false localising has implications for diagnostic investigation.

The clinical examination of the nervous system is based on the premise of clinicopathological correlation, with a particular neurological sign indicating pathology at a specific locus or pathway within the nervous system. Occasionally, however, this semiology “breaks down”, in that neurological signs may reflect pathology distant from the expected anatomical locus.

The notion of “false localising signs” was first elucidated by James Collier in 1904: on the basis of clinical examination during life and subsequent postmortem studies, he noted false localising signs in 20 of 161 consecutive cases of intracranial tumour examined pathologically, most occurring in patients with supratentorial lesions. Despite this high frequency (12.4%), Collier felt false localising signs were being observed less frequently because of the earlier diagnosis of tumours, a theme reiterated by later authors, implying that such signs are a late feature in the natural history of tumours. Gassel noted false localising signs to be more common in patients with raised intracranial pressure.

As there have been few previous reviews of false localising signs, this article attempts a brief overview of these phenomena and their possible pathophysiology. It does not aim to provide an exhaustive review of publications on the subject, most of which are case reports or small case series rather than systematic studies. Rather, it selects illustrative examples, particularly where these elucidate possible pathophysiological mechanisms.

Structural brain imaging, particularly magnetic resonance imaging (MRI), affording as it does the opportunity to study pathological anatomy contemporaneous with clinical examination, has provided some new insights into the causes of these signs.

FALSE LOCALISING SIGNS ASSOCIATED WITH INTRACRANIAL LESIONS

Sixth nerve palsy

Sixth nerve palsy, either unilateral or bilateral, is the classic example of a false localising sign. It occurs in the context of raised intracranial pressure of whatever cause (supratentorial or infratentorial space occupying lesion, idiopathic intracranial hypertension, cerebral venous sinus thrombosis). It was the most common of the false localising signs observed by Collier (12/20) in cases of intracranial tumour. In a retrospective review of 101 cases of idiopathic intracranial hypertension (IIH), Round and Keane noted 14 cases (11 unilateral, three bilateral). IIH with sixth nerve palsy in the absence of papilloedema has also been reported.

The pathophysiological mechanism of sixth nerve palsy with increased intracranial pressure has traditionally been said to be stretching of the nerve in its long intracranial course, or compression against the petrous ligament or the ridge of the petrous temporal bone. Collier, however, was “unable to accept this explanation”, his view being that since the sixth nerve emerges straight forward from the brain stem, whereas other cranial nerves emerge obliquely or transversely, it is more liable to the mechanical effects of backward brain stem displacement by intracranial space occupying lesions.

Fifth and seventh nerve false localising signs

Dysfunction of the fifth and seventh cranial nerves has also been reported with raised intracranial pressure. This dysfunction may be hypoactive or hyperactive, manifesting with negative or positive Jacksonian symptoms, respectively; hence there may be trigeminal neuropathy or trigeminal neuralgia. Facial palsy of lower motor neurone type or hemifacial spasm. Fifth and seventh nerve involvement has been recorded in the context of posterior fossa neoplasms or more diffuse neoplastic disease, and with IIH. Reports of false localising seventh nerve palsies suggest they most often occur at the same time as, or after, the development of sixth nerve palsies. Brain stem distortion resulting in traction on cranial nerves has been suggested as the pathophysiological mechanism in tumour associated cases, but this seems a less compelling explanation for IIH cases.

Collier suggested that motor involvement preceded sensory features in fifth nerve palsy, but the clinical features reported in subsequent cases

Abbreviations: IIH, intracranial hypertension; MRI, magnetic resonance imaging.
suggest sensory findings are more prominent.\textsuperscript{7–10} Gassell found motor involvement in only two of eight patients with false localising fifth nerve involvement.\textsuperscript{9, 11} Arase and Kondo implicate adherence of arachnoid membrane to the nerve as a contributing factor and advocate its resection in order to straighten the nerve axis.\textsuperscript{12}

Hemifacial spasm as a false localising sign of posterior fossa tumour seems altogether rarer than trigeminal neuralgia, indeed exceptional.\textsuperscript{13, 14} As with trigeminal neuralgia, the question of whether the false localising sign follows neurovascular compression or mechanical distortion remains open.

**Third nerve palsy and Kernohan’s notch phenomenon**

Transientorial herniation of the medial temporal lobe (uncus), as a consequence of shift of intracranial contents secondary to raised intracranial pressure, may be associated with third nerve palsy. Perhaps because of the intra-axial fascicular arrangement of fibres within the third nerve, the peripherally located pupilloconstrictor fibres are most vulnerable, such that unilateral pupillary dilatation contralateral to mass lesions is axiomatic in neurosurgical practice, yet the consequent fixed dilated pupil is ipsilateral to a lesion as well as ipsilateral third nerve palsy results. However, other cases have been presented in which trigeminal neuralgia did not resolve after tumour removal alone.\textsuperscript{15} Matsuura and Kondo implicate adherence of arachnoid membrane to the nerve as a contributing factor and advocate its resection in order to straighten the nerve axis.\textsuperscript{12}

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**FALSE LOCALISING SIGNS ASSOCIATED WITH SPINAL CORD LESIONS**

**Foramen magnum and upper cervical cord lesions**

Lesions at the level of the foramen magnum may produce, in addition to suboccipital and neck pain and upper motor neurone (long tract) signs, false localising signs: paraesthesia in the hands and lower motor neurone signs in the upper limbs.\textsuperscript{15} The wasting (“remote atrophy”), weakness, and areflexia may suggest the involvement of cervical cord segments below the level of the foramen magnum, hence are false localising. Similarly, a syndrome of “numb and clumsy hands” has been described with midline cervical disc protrusions at the C3/C4 level; concurrent with numbness of fingertips and palms, there may be a tightening sensation at mid-thoracic level.\textsuperscript{24} Cervical spinal cord lesions (cervical spondylosis, herniated disc) at or above the level of C4 in which finger and hand dysaesthesia with hand muscle atrophy preceded limb spasticity or gait disturbance have also been reported.\textsuperscript{25}

There is currently no compelling pathophysiological explanation for these false localising signs, but three principal mechanisms have been invoked: arterial, venous, and mechanical.\textsuperscript{26} The arterial hypothesis postulates that tumour in some way compromises descending anterior spinal artery blood supply to the lower cervical cord with resultant focal ischaemia. However, this would also seem to require insufficiency of
radicular spinal arteries entering intervertebral foramina to join anterior spinal artery. Moreover, there is some evidence that blood flows rostrally rather than caudally in the anterior spinal artery, the low pressure venous system would seem inherently more vulnerable to the effects of compression than the arterial system, with resultant tissue stasis and hypoxia perhaps causing the clinically observed neurological dysfunction. Mechanical stresses within the spinal cord, consequent perhaps on the conjunction of extrinsic compression with the anchoring of the spinal cord by the denticulate ligaments, have also been suggested to account for remote signs. The paucity of pathological material has made it difficult to obtain direct evidence in support of any of these hypotheses. In all cases, the differential decussation of corticospinal tract fibres to fore and hind limbs and the lamination of sensory tracts may be prerequisites for the clinical features.28

**Lower cervical cord and upper thoracic cord lesions**

Because of the anatomical decussation of spinothalamic tract fibres two or three segments above the level at which they enter the spinal cord from their respective dermatomes, it is not unusual for sensory levels two or three segments distant from the level of cord pathology to be observed clinically. However, more distant sensory and motor signs may be observed with cord pathology, whose explanation is altogether more challenging. Compressive cervical myelopathy may produce a false localising thoracic sensory level,29 sometimes called a mid thoracic girdle sensation, in addition to lower limb weakness and hyperreflexia. Similarly, lumbar spinal disease may be simulated by more rostral pathology, for example, urinary retention, leg weakness, and lumbar sensory findings may be the presenting features of high thoracic cord compression, with clinicoradiological discrepancy of as much as 11 segments.30

As with foramen magnum and upper cervical cord lesions, opinion is divided as to the pathophysiological explanation for these observations. Comparing axial MR images of the cervical cord in 10 patients with compressive cervical myelopathy, five with girdle signs and five without, Ochiai et al noted that the midline ventral portion of the cervical cord was more markedly compressed in those with the false localising sensory level. This observation led them to suggest that ischaemia in the watershed zone of the anterior spinal artery, affecting the medial portion of the lamination of the spinothalamic tract, was the causative mechanism for the girdle sensation, rather than direct compression of spinothalamic fibres.31 Jamieson et al, however, favoured venous obstruction with ensuing intramedullary stasis and hypoxia as the explanation for remote phenomena.32 Of course, different cases may have different pathogenesis, and/or more than one mechanism may be operating in any one case.

**OTHER “FALSE LOCALISING” SIGNS**

The “false localising” terminology has also been used in other contexts, for example to describe intraneural ophthalmoplegia, usually indicative of medial longitudinal fascicular dysfunction, in patients with myasthenia gravis; this “pseudo-intraneural ophthalmoplegia” has also been observed in dermatomyositis. Similarly, myasthenic nystagmus may suggest a brain stem lesion. Other “pseudo-signs” are described which might be encompassed by the “false localising” rubric,1 such as pseudo-(choiro-)atresia and pseudoastereognosis, which may occur with high cervical cord lesions rather than intracranial pathology. Signs traditionally thought to be of cortical origin, such as aphasia and inattention, may sometimes occur with exclusively subcortical pathology; conversely, exclusively cortical lesions may sometimes result in dysarthria. Sensory stimuli (tactile, visual) may be detected contralateral to the stiimuolated side (alloaesthesia, allochiria). “Pseudoasympotymymelina” has been used to describe a selective loss of pain and temperature sensation with relative preservation of vibration and position sense seen in amyloid polyneuropathy and Tangier disease, a small fibre sensory neuropathy in the absence of any spinal cord pathology, and hence false localising.

**DISCUSSION**

False localising neurological signs have presented significant challenges to clinical neurologists: in the era before neuroimaging, operations were sometimes performed on, and treatments administered to, the wrong side based on these signs.2 The preponderant association with extrinsic mass lesions, such as intracranial tumours (especially meningioma), subdural haematoma, and intervertebral disc prolapse, has long been noted, although intrinsic lesions may certainly be responsible on occasion.2 3 Some of these pathologies exert their effects acutely, whereas for others (for example, meningiomas) it is their slow growth which is implicated. The possibility of multifactorial pathophysiology therefore seems likely.

After almost one hundred years use,2 the term “false localising sign” is thoroughly ingrained in clinical teaching, although it is important to be aware that such signs are in no way “false”. The practical implication of the name is that such signs cannot be relied on when assessing clinical localisation, which challenges traditional clinicanoatomical correlation. “Diaschisis” is a technical term for dysfunction distant from pathology, particularly familiar in the field of functional imaging.2 However, it is associated with the notion of “transneural depression”, which may not be relevant to the pathophysiology of these signs, besides being something of a mouthful for clinical practice. Perhaps “distant signs” or “remote signs” would fit the bill, though lacking any emotive resonance. Most important, since false localising signs may be indicative of serious, even life threatening, pathology within neural pathways, awareness of them and the situations in which they occur, will facilitate appropriate and timely investigation and management.

**ACKNOWLEDGEMENTS**

Thanks are extended to Drs Alasdair Coles and Peter Enevoldson for commenting on earlier versions of the manuscript.

Conflict of interest: None.

**REFERENCES**

Intracranial hypertension secondary to giant arachnoid granulations

We present a case of intracranial hypertension in a non-obese male in whom the only finding resulting from additional tests was the presence of giant arachnoid granulations in lateral sinuses. A 51 year old man presented episodes of transient obscurations of vision in the left eye lasting a few seconds. On general physical examination, the patient was not obese (170 cm height, 70 kg weight) and no relevant findings were detected. During neurologic examination a complete left papilledema was detected. In the right optic disc a nasal effacement with peripapillary hemorrhages was observed. Campimetry was normal in the right eye; in the left eye an enlargement of the blind spot was observed. Visual acuity was normal. A lumbar puncture in lateral decubitus showed an opening pressure of 27 cm of water. Laboratory studies, cranial magnetic resonance imaging and left transverse sinus hypoplasia.

For correspondence: A Arjona, C/Violeta 70, E-04720 Aguadulce, Almería, Spain; arjona@medynet.com

References

Intracranial hypertension secondary to giant arachnoid granulations

A Arjona, F Delgado and E Fernandez-Romero

*J Neurol Neurosurg Psychiatry* 2003 74: 418
doi: 10.1136/jnnp.74.4.418

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