

TEACHING CASE REPORT

A woman with progressive ataxia and hemiparesis on the right side: Where's the lesion?

The case: A 53-year-old right-handed woman reported 2 years of gait difficulties and progressive clumsiness and weakness of her right upper and lower extremities. She was referred to our neurology service for assessment and was found to have abnormal speech (spastic dysarthria) and horizontal nystagmus on leftward gaze, with fast component to the right. The left side of her body was normal, but she had right-side hypertonia, hyperreflexia and mild hemiparesis with extensor plantar response. She also had dorsiflexion and eversion of the right ankle. She exhibited past-pointing on the right upper limb and had a positive result on heel-to-shin testing of the right lower limb. Sensation was intact, and other results of the neurologic and general physical examinations were unremarkable. What is the differential diagnosis of this case of hemiparesis and incoordination on the right side?

Motor loss arising from the brain or the brain stem usually results from a lesion on the contralateral side.¹ The corticospinal axons descend from the motor cortex and pass through the internal capsule and crus and then through the brain stem with the corticobulbar tract. Most (80%–90%) of the corticospinal fibres decussate to the opposite side at the medullary pyramids, continuing their descent in the spinal cord as the lateral corticospinal tract. The remaining fibres descend uncrossed as the anterior corticospinal tract.¹ Coordination of movement is mediated by the cerebellum, which processes signals from the motor cortex to the ipsilateral spinal cord (and thence to the muscles) via the spinocerebellar tracts. Thus, ataxia is usually caused by a lesion in the cerebellar hemisphere or the spinocerebellar tracts on the same side.¹

Ipsilateral weakness may paradoxically

be caused by a contrecoup lesion or contusion but may also be attributed, if only rarely, to Kernohan's notch phenomenon.² In 1929, Kernohan and Woltman described hemiparesis in a patient with an ipsilateral supratentorial mass lesion,² which resulted in displacement of the brain stem against the incisura of the tentorium cerebelli. The groove in the peduncle caused by this displacement came to be called Kernohan's notch. The false-localizing ipsilateral hemiparesis, resulting from compression of the crus cerebri by the tentorium, likewise became known as the Kernohan–Woltman syndrome or Kernohan's notch phenomenon (Figure 1A). Although originally described in a patient with a primary brain tumour, the phenomenon may also occur with traumatic brain injury or with displacement of the cerebral peduncles.

The patient described here presented with ataxia and weakness on the

right side, and brain MRI revealed a 5 × 5.4 × 6 cm extra-axial mass in the right posterior fossa. The mass was isointense on T₁-weighted imaging (Figure 2A) and isointense to hypointense on T₂-weighted imaging (Figure 2C), with homogeneous contrast enhancement suggestive of meningioma (Figure 2B and Figure 2D). The mass was associated with significant mass effect on the adjacent right cerebellar hemisphere and pons, with compression of the fourth ventricle and dilatation of the lateral and third ventricles, consistent with noncommunicating hydrocephalus. The meningioma caused downward cerebellar tonsillar herniation and leftward and upward transtentorial herniation of the brain stem (Figure 2C and Figure 2D), which resulted in right hemiparesis because of compression of the left cerebral peduncle by the tentorium cerebelli (Figure 1B). A ventriculoperitoneal shunt was inserted, after which the meningioma was partially excised to preserve function of the adherent lower cranial nerves. The patient subsequently underwent gamma knife irradiation of the remnant tumour. Post-

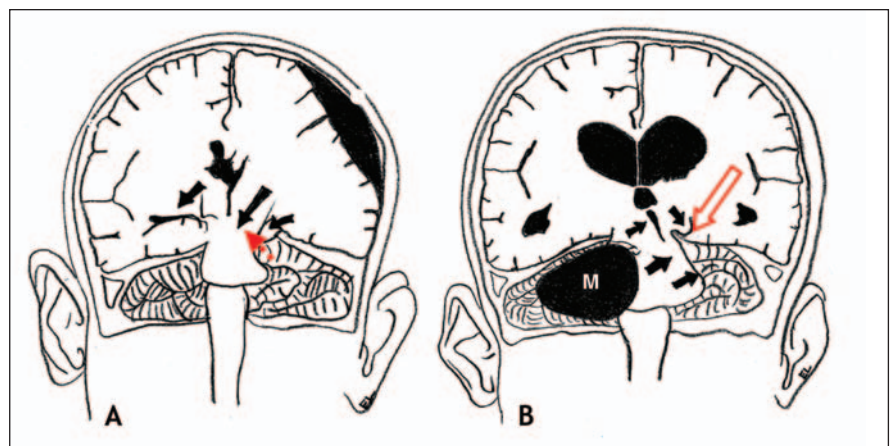


Figure 1: (A) Schematic coronal section of the brain showing large (supratentorial) right subdural hematoma causing ipsilateral transtentorial herniation, which has resulted in compression of the contralateral cerebral peduncle (broken, red arrow). This led to ipsilateral (right-sided) weakness. (B) Schematic coronal section of the brain showing (infratentorial) right meningioma (M) causing upward and leftward displacement of the midbrain and cerebral peduncles, which has resulted in compression of the left cerebral peduncle and corticospinal tracts against the tentorium cerebelli (open arrow), which contributed to the ipsilateral (right-side) weakness. In both figures, the solid arrows indicate the direction of shift due to the mass effect, either from the subdural hematoma or from the tumour.

operative recovery was uneventful, and the patient recovered normal function.

The patient's ipsilateral weakness resulted from an infratentorial, rather than a supratentorial, mass. The large cerebellar meningioma pushed upward and to the left, causing the midbrain and left cerebral peduncle to be compressed by the tentorium cerebelli. Kanis and associates³ described a similar phenomenon in a patient with an inferior cerebellar infarction; that patient experienced ipsilateral hemiparesis because of cerebellar edema, which caused displacement of the medulla with impaction of the pyramids upon the clivus.

The combination of right-side weakness and ataxia in the patient reported here mimicked ataxic hemiparesis, which is usually associated with a lacu-

nar stroke in the pons or internal capsule. Weakness in ataxic hemiparesis is caused by disruption of the corticospinal tracts, and the ataxia is attributed either to involvement of the frontopontocerebellar connections or to impaired position sense.⁴ A lesion localized to the cerebellar hemisphere should cause ipsilateral ataxia, but not weakness, unless there is concomitant disruption of the corticospinal tracts. In this patient, the displacement of the brain stem and cerebral peduncle to the left caused weakness on the right side by compressing the contralateral corticospinal fibres against the tentorium cerebelli, whereas the cerebellar meningioma on the right side caused ipsilateral ataxia. Other conditions, such as multiple sclerosis, chronic subdural hematoma and neuro-

cysticercosis, can mimic ataxic hemiparesis by involving the contralateral corticospinal tract and the ipsilateral cerebellum or spinocerebellar tracts.

Meningiomas are the second most common brain tumour in adults.⁵ Most are benign, but up to 20% exhibit clinically aggressive features. The annual incidence of meningiomas is estimated at between 0.8 and 4.9 per 100 000, but the true incidence is probably higher, as many benign meningiomas are asymptomatic.⁵ Intracranial meningiomas are usually surgically excised, although radiosurgery and radiation therapy may be considered for patients whose lesions are deemed inoperable, who refuse surgery, who have histologic findings suggestive of an aggressive lesion or who undergo inadequate resection. The patient described here, who underwent surgical resection and then radiosurgery (because of incomplete resection of the tumour), had an uneventful recovery.

Jing-Hui Yik

Yong Loo Lin School of Medicine
National University of Singapore
Raymond C.S. Seet MBBS MMed
(IntMed)

Yong Loo Lin School of Medicine
National University of Singapore
Division of Neurology
National University Hospital

Chong-Han Pek

Yong Loo Lin School of Medicine
National University of Singapore
Erle C.H. Lim MBBS MMed (Int Med)
Yong Loo Lin School of Medicine
National University of Singapore
Division of Neurology
National University Hospital
Singapore

This article has been peer reviewed.

Competing interests: None declared.

REFERENCES

1. Young PA, Young PH. *Basic clinical neuroanatomy*. Baltimore: Williams and Wilkins; 1997.
2. Kernohan JW, Woltman HW. Incisura of the crus due to contralateral brain tumor. *Arch Neurol Psychiatry* 1929;21:274-87.
3. Kanis KB, Ropper AH, Adelman LS. Homolateral hemiparesis as an early sign of cerebellar mass effect. *Neurology* 1994;44:2194-7.
4. Gorman MJ, Dafer R, Levine SR. Ataxic hemiparesis: critical appraisal of a lacunar syndrome. *Stroke* 1998;29:2549-55.
5. McMullen KP, Stieber VW. Meningioma: current treatment options and future directions. *Curr Treat Options Oncol* 2004;5:499-509.

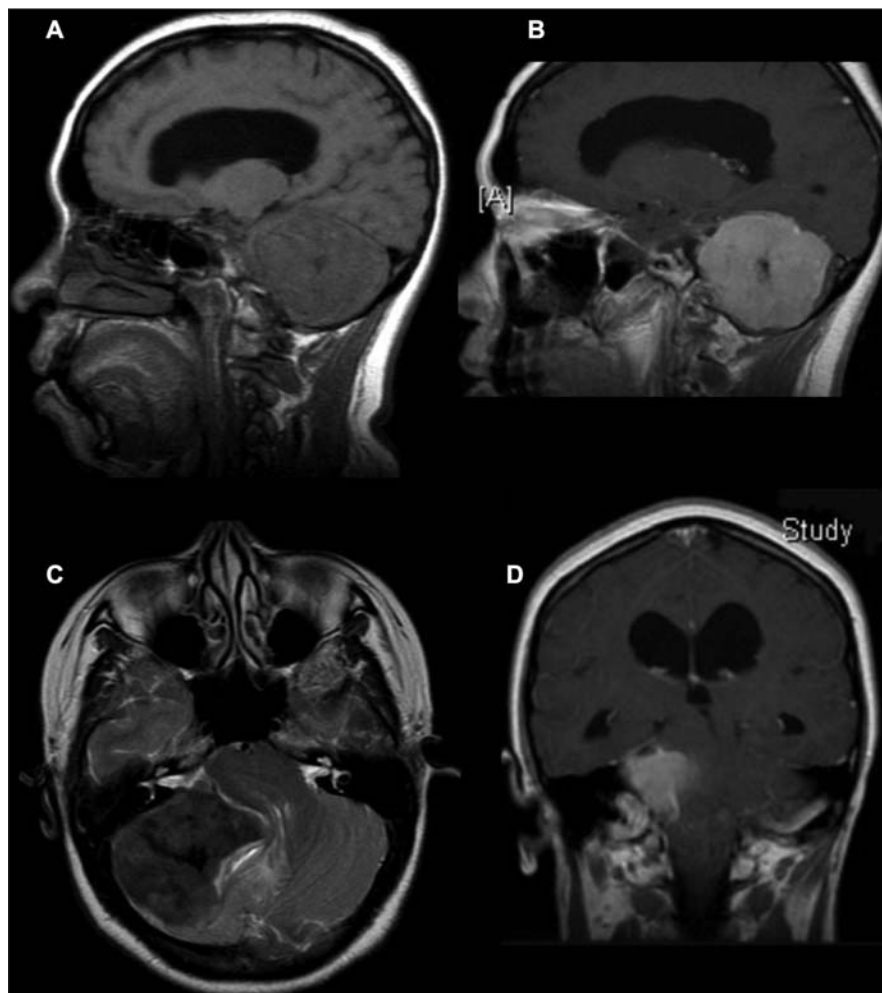


Figure 2: Sagittal T_1 -weighted MRI, without (A) and with (B) gadolinium contrast agent. Transverse T_2 -weighted (C) and coronal T_1 -weighted (D) images, both with gadolinium contrast agent, show $5 \times 5.4 \times 6$ cm meningioma. The infratentorial mass is evident in Figure D, pushing the brain stem upward against the tentorium cerebelli at Kernohan's notch.