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References

- 1. Rados A. Conical cornea and mongolism. Arch Ophthalmol 1948;40:454–9.
- Cullen JF, Butler HG. Communications. Mongolism and keratoconus. Br J Ophthalmol 1963;47:321–9.
- 3. Cullen JF. Blindness in mongolism. Br J Ophthalmol 1963;47:331–3.
- Karseras AG, Ruben M. Aetiology of keratoconus. Br J Ophthalmol 1976;60:522–5.
- 5. Boger VP III, Petersen RA, Robb RM. Keratoconus and acute hydrops in mentally retarded patients with congenital rubella syndrome. Am J Ophthalmol 1981;91:231–3.
- 6. Slusher MM, Laibson PR, Mulberger RD. Acute keratoconus in Down's syndrome. Am J Ophthalmol 1968;66:1137–43.
- Beilin B, Kadari A, Shapira Y, Shulman D, Davidson JT. Anaesthetic considerations in facial reconstruction for Down's sydrome. J R Soc Med 1988;81:23–6.
- Miller R, Gray SD, Cotton RT, Myer CM, Netterville J. Subglottic stenosis and Down's syndrome. Am J Otolaryngol 1990;11:274–7.
- 9. Tuft SJ, Gregory WM, Buckley RJ. Acute corneal hydrops in keratoconus. Ophthalmology 1994;101: 1738–44.
- Doughman DJ. Corneal edema. In: Duane's clinical ophthalmology, vol 4. Philadelphia: JB Lippincott, 1991:11.
- Cameron JA, Al-Rajhi AA, Badr IA. Corneal ectasia in vernal keratoconjunctivitis. Ophthalmology 1989;96:1615–23.
- Kirkness CM, Adams GGW, Dilly PN, Lee JP. Botulinum toxin A-induced protective ptosis in corneal disease. Ophthalmology 1988;95:473–80.
- Freegard T, Mackie I, Rostron C. Therapeutic ptosis with botulinum toxin in epikeratoplasty. Br J Ophthalmol 1993;77:821–2.
- 14. Frantz JM, Insler MS, Hagenah M, et al. Penetrating keratoplasty for keratoconus in Down's syndrome. Am J Ophthalmol 1990;109:143–7.

Sir,

The Rules of Third Nerve Palsy in Children

The rules of third nerve palsy can be useful in the diagnosis of acquired third nerve palsy. However, it is not generally recognised that there are limitations when they are applied to children. This case report illustrates this fallacy and explores the reasons for their limitations.

Case 1

A 6-year-old boy presented with a complete, left third nerve palsy associated with frontal headache, periorbital pain, cough, vomiting and pyrexia. Tenderness was elicited over the left frontal and maxillary sinuses but there were no other orbital signs or meningism. Blood films showed neutrophilia but cultures were negative. CT scan showed mucosal thickening, consistent with sinusitis, in the left ethmoidal, maxillary and sphenoidal sinuses. No intracranial lesions were seen. The patient proceeded to have magnetic resonance angiography (MRA) but no intracranial aneurysms were identified. After a course of antibiotics, symptoms and signs of the sinusitis quickly resolved followed by a more gradual return to normal of third nerve function.

Discussion

According to the conventional rules of third nerve palsy, an aneurysm could certainly have been the cause of the child's third nerve palsy. For this reason, an MRA was felt to be necessary to exclude this possibility. Although in adults the rules of third nerve palsy can be useful as a clinical guide for indicating the likelihood of aneurysmal compression, in children they have a different significance and are of limited clinical value. The reasons for these limitations become apparent when the origins of these rules are explored.

Pain in third nerve palsy. Putative, trigeminal fibres 'hitch-hiking' in the oculomotor nerve are thought to give rise to the characteristic pain of ischaemic third nerve palsy.¹ Ischaemic pain is supposed to be dull and constant in character, in contrast to aneurysmal pain which tends to be sharp and throbbing in character.²

This rule distinguishes the subtle differences in the character of pain caused by ischaemic and aneurysmal lesions. Due to the subjective nature of these symptoms, this rule is of limited value even in adults.

This rule cannot be applied to children as ischaemic neuropathy is not known to occur in children.³ Aneurysms are also rare in childhood and other more common causes of third nerve palsy such as trauma, post-viral neuropathy, raised intracranial pressure, meningitis and neoplasm can often be associated with headaches. Therefore, in a child with a painful third nerve palsy, other causes of headache should be excluded before undertaking investigations for intracranial aneurysm.

Pupil involvement in third nerve palsy. The rule states that 'when an aneurysm compresses the oculomotor nerve, the iris sphincter will be impaired'.⁴

Pupillary fibres are distributed superficially on the dorsomedial aspect of the nerve trunk and derive their blood supply mainly from pial vessels on the nerve sheath. Although they are prone to compression from aneurysms, the pupillomotor fibres are spared in ischaemic third nerve palsy when occlusion of the vasa nervorum occurs causing microinfarction of the central somatomotor fibres.⁵

This rule is also based on the contrasting clinicopathological features between ischaemia and aneurysmal compression of the third nerve and for the same reasons is therefore also not strictly applicable to children. Furthermore, pupil involvement occurs almost universally in children with third nerve palsy from non-aneurysmal causes.³ In two large series, pupil involvement was found in 64% of adults and

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90% of children. Of those with pupil involvement, 41% of adult cases and only 7% of childhood cases had aneurysms.^{3,4} Pupil involvement is therefore a very unreliable indicator of aneurysm in children.

However, when there is no other clinical evidence to suggest an alternative cause of isolated third nerve palsy in children, an aneurysm has to be considered regardless of pupil involvement unless the third rule based on extraocular involvement can be satisfied.

Extraocular muscle involvement in third nerve palsy. This rule states that 'when pupil sparing occurs with complete extraocular involvement it is not due to an aneurysm, but if the extraocular muscles are only partially involved, an aneurysm can exist'.⁶ This is because fibres of the inferior division are located dorso-medially in the nerve and may be spared by aneurysms from the basilar apex or the cavernous sinus. However, when aneurysmal dilation progresses to cause complete extraocular involvement, the pupil sphincter is inevitably affected.

This rule should be applied in both adults and children and is helpful in preventing patients who have complete extraocular muscle involvement and absolute pupil sparing from unnecessary investigation. To our knowledge only one case has been reported which violates the above rules: a basilar aneurysm caused complete extraocular palsy with absolute pupil sparing in a 65-year-old.⁷

In conclusion, the rules of third nerve palsy are derived from observations on the adult population and are primarily based on the contrasting clinicopathological features between ischaemic and aneurysmal compression. Their limitations should be noted when assessing children with third nerve palsies so that appropriate investigations are instituted. Y. C. Yang D. E. Laws A. Chandna

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References

- 1. Bortolami R, D' Alessandro R, Manni E. The origin of pain in ischaemic diabetic third nerve palsy. Arch Neurol 1993;50:795.
- 2. Miller NR. Walsh and Hoyt's clinical neuro-ophthalmology, 4th ed, vol 2. Baltimore: Williams and Wilkins, 1988.
- 3. Miller NR. Solitary oculomotor nerve palsy in childhood. Am J Ophthalmol 1977;83:106–11.
- Rucker CW. The causes of paralysis of third, fourth and sixth nerves. Am J Ophthalmol 1966;61:1293–8.
- 5. Kerr FWL, Hollowell OW. Location of pupillomotor and accommodation fibres in the oculomotor nerve: experimental observations on paralytic mydriasis. J Neurol Neurosurg Psychiatry 1964;267:473–81.
- 6. Trobe JD. Third nerve palsy and the pupil. Arch Ophthalmol 1988;106:601–2.
- Lustbader JM, Miller NR. Painless, pupil sparing but otherwise complete oculomotor nerve paresis caused by basilar artery aneurysm. Arch Ophthalmol 1988;106:583.