CASE REPORT

Diagnostic pitfall of progressive isolated abducens nerve palsy: a report of two cases

Noel CY Chan[1,2], FRCSEd(Ophth); Venice SW Li[3], MBChB; Andy CO Cheng[2,4], FRCOphHK; Tom CY Cheung[5], FRCR; Sherman SM Lo[6], FRCR; Carmen KM Chan[2,3], FRCSEd(Ophth)

[1]*Department of Ophthalmology & Visual Sciences, Prince of Wales Hospital & Alice Ho Miu Ling Nethersole Hospital, Hong Kong*
[2]*Department of Ophthalmology & Visual Sciences, The Chinese University of Hong Kong, Hong Kong*
[3]*Hong Kong Eye Hospital, Hong Kong*
[4]*Department of Ophthalmology, Hong Kong Sanitorium & Hospital, Hong Kong*
[5]*Diagnostic Radiology and Imaging Department, Prince of Wales Hospital, Hong Kong*
[6]*Scanning Department, St Teresa's Hospital, Hong Kong*

Correspondence and reprint requests:
Carmen KM Chan, Hong Kong Eye Hospital, 147K Argyle Street, Kowloon, Hong Kong. Email: kmcc2001@hotmail.com.

Abstract

We describe two cases of progressive abducens nerve palsy secondary to a compressive lesion in the Dorello canal. Blood tests, lumbar puncture, nasopharyngeal examination, computed tomography, magnetic resonance imaging, and angiography were performed, but the etiology could not be identified. After consultation with a neuroradiologist, fine-cut magnetic resonance imaging along the course of the abducens nerve with gadolinium and constructive interference in steady state sequences was performed, and the diagnosis was made. In cases of small intracranial pathology, the use of thinner slices or specific sequences is suggested to better visualize the course of cranial nerves.

Key words: Abducens nerve; Cranial nerve palsy; Magnetic resonance imaging

Case presentation

Patient 1

In 2010, a 49-year-old woman with hypertension presented with a 2-week history of acute onset binocular horizontal diplopia. Clinical examination revealed isolated right eye abduction defect. Blood tests showed hypercholesterolemia only. Results of computer tomography of the brain and orbits and magnetic resonance imaging (MRI) of the brain were non-contributory. A preliminary diagnosis of right abducens nerve (CN6) palsy of microvascular origin was made. The patient was then started on aspirin and simvastatin, but the CN6 palsy progressed in the following months. Tests for thyroid eye disease and myasthenia gravis, lumber puncture, ear, nose and throat examination, repeated MRI with gadolinium, and angiography of the brain and neck (reviewed with neuro-radiologists and neurologists) showed no causative lesion. After consultation with a neuroradiologist, fine-cut MRI along the course of CN6 with gadolinium and constructive interference in steady state (CISS) sequences was performed 8 months after symptom onset. A 0.9×0.9×0.9 cm enhancing extra-axial lesion was identified at the right retro-clival region, situating at the Dorello canal in the vicinity of CN6, suggestive of a nerve sheath tumor or meningioma (**Figure 1**). She underwent craniotomy with tumor excision and then stereotactic radiosurgery for residual tumor and strabismus surgery for symptomatic relief. The pathology of the lesion was meningioma. At the 6-year follow-up, MRI of the brain showed a residual tumor of 0.6×0.9×0.9 cm at the right petroclival region, with no significant interval change.

Patient 2

In 2019, a 20-year-old woman presented with a 1-year history of painless progressive binocular horizontal diplopia. Clinical examination revealed isolated left eye abduction defect. Results of computed tomography of the

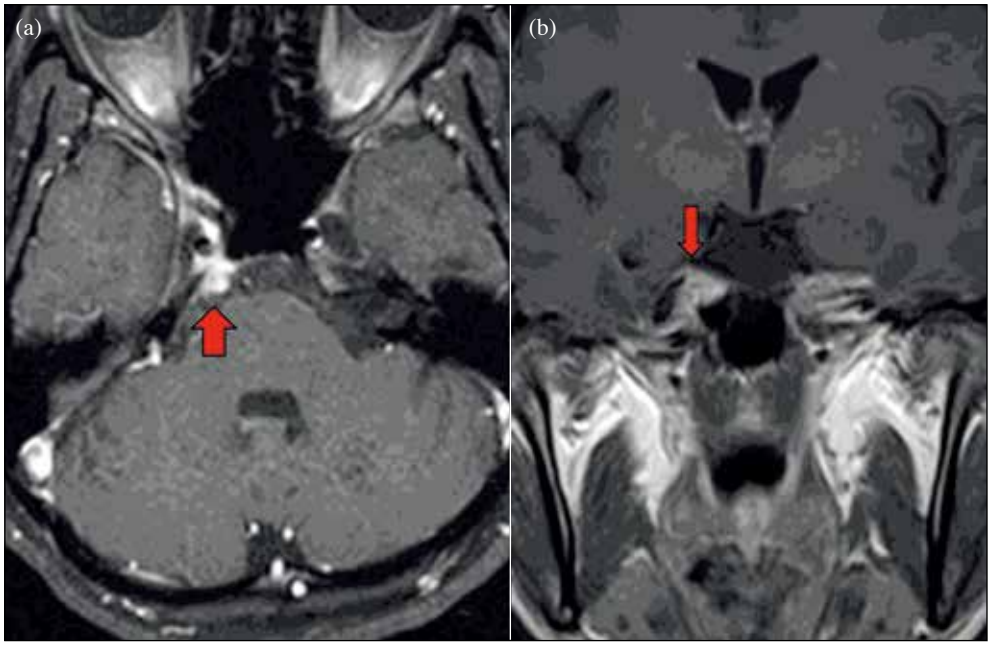

Figure 1. Patient 1: (a) Axial and (b) coronal T1-weighted high-resolution fat-suppressed post-gadolinium contrast magnetic resonance images along the course of abducens nerve revealing a 0.9×0.9×0.9cm contrast-enhancing extra-axial mass (arrows) at the right retroclival region along the course of Dorello canal.

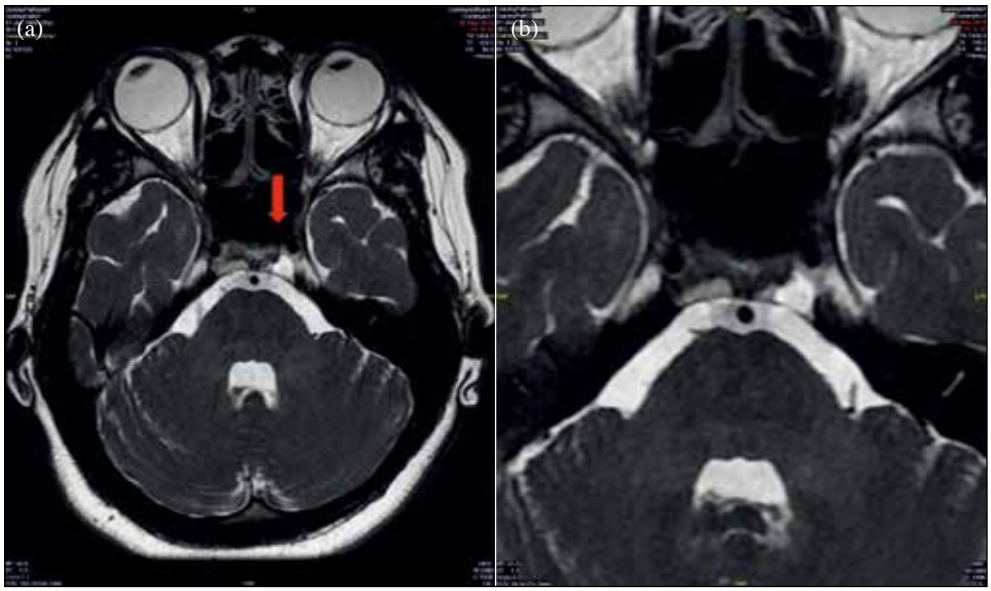

Figure 2. Patient 2: (a) Axial T2-weighted 3D turbo spin-echo sequence magnetic resonance images along the course of abducens nerve revealing a 0.5×0.6 cm non-enhanced mass (arrow) at the left Dorello canal. (b) Post-contrast image does not show the lesion well; the lesion's cystic nature with T1 hypointense signal is similar to the adjacent normal bone marrow.

brain and MRI of the brain, orbits, and internal acoustic meatus with contrast (reviewed by neuro-radiologist) were unremarkable. Further extensive investigations were non-contributory. However, the left CN6 palsy was progressive. After consultation with the neuro-radiologist, a fine-cut MRI along the course of CN6 was performed 4 months after presentation. A focal T2-hyperintense, T1-hypointense signal was identified in the dorsal aspect of clivus involving left Dorello canal, which was only visible in the fine cut of 3D volumetric acquisition using turbo spin-echo with submillimeter section thickness (**Figure 2**). There was no mass effect or contrast enhancement. The signal was not suppressed on fat suppression sequence. Corresponding bony lucent changes in the clivus were retrospectively noted in the initial computed tomographic scan. The lesion involved the left Dorello canal and hence may be accountable for the symptoms. The lesion (0.55×0.6 cm) had a well-demarcated border with no extraosseous soft tissue or aggressive bone destruction, which was likely to be a benign notochordal cell tumor. Differential diagnoses include arachnoid cyst, red marrow deposition, and ecchordosis physaliphora. The patient was referred to the neurosurgical unit and opted for

observation. At the 2-year follow-up, she was coping with a combination of Fresnel prism and head posturing.

Discussion

The CN6 is the most frequently affected ocular motor cranial nerve[1] and is susceptible to injury along its course from the pons to the orbits. The prevalence and etiologies of isolated CN6 palsy vary across different age groups. In a Japanese cohort, most cases were attributed by microvascular origin (35.9%) and neoplasm (22.2%), but the most frequent cause was head trauma and congenital in patients of age <20 years.[1] In patients aged 20 to 49 years, 44% of cases were attributed to an intracranial organic lesion such as aneurysm, neoplasm, or arteriovenous malformation.[1] In those aged ≥50 years, microvasculopathy accounts for most cases.[1,2] Despite advances in modern imaging techniques, causes remained undetermined in 22% to 30% of cases.[2]

We demonstrated a diagnostic pitfall in patients with isolated CN6 palsy. Close monitoring without neuroimaging is advocated in patients aged ≥50 with isolated CN6 palsy and cardiovascular risk factors,[3] as most will improve spontaneously by 6 months.[1] However, controversies remain regarding prompt versus delayed neuroimaging, as pre-existing vasculopathic risk factors do not preclude intracranial neoplasm. Adequate follow-up is crucial for isolated CN6 palsy of presumed microvascular etiology. However, prompt neuroimaging is recommended for patients of young age or without identifiable vasculopathic risk factors. MRI is recommended if the nerve palsy progresses or should additional neurological sign emerge.

We highlighted the importance of choosing the correct scanning sequence and communication with neuroradiologists. MRI is preferred; CT is complementary in cases with suspicion of a bone or skull base lesion or with contraindication for MRI. Intravenous administration of gadolinium is essential in detecting small enhancing lesions of nerves, whereas leptomeningeal/dural/carvenous sinus enhancement may indicate an inflammatory or neoplastic lesion.[2] Apart from fat saturation technique, a heavily T2-weighted sequence, usually a 3D turbo or fast spin-echo sequence (SPACE or CUBE) with submillimeter section thickness or a balanced steady-state free-precession sequence (such as constructive interference in steady state sequence or fast imaging employing steady-state acquisition) should also be performed.[4,5] Although there is no standardized imaging technique for the cisternal part of CN6, the use of pre-contrast constructive interference in steady state sequence / fast imaging employing steady-state acquisition and post-contrast 3D turbo or fast spin-echo to evaluate this part of the abducens nerve that traverses the Dorello canal is recommended.[5]

Though rare, neoplasm of the Dorello canal may present as isolated progressive CN6 palsy. In cases with no abnormality found, we suggest consultation with neuroradiologists before concluding the cases as idiopathic. When suspicion of intracranial pathology remains high, repeat imaging with thinner slices or specific sequences is suggested to better visualize the course of cranial nerves. Appropriate neuroimaging prevents unnecessary investigations, which are potentially invasive or costly.

Contributors

All authors designed the study, acquired the data, analyzed the data, drafted the manuscript, and critically revised the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

Funding/support

This study received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data Availability

All data generated or analyzed during the present study are available from the corresponding author on reasonable request.

References

1. *Akagi T, Miyamoto K, Kashii S, Yoshimura N. Cause and prognosis of neurologically isolated third, fourth, or sixth cranial nerve dysfunction in cases of oculomotor palsy. Jpn J Ophthalmol 2008;52:32-5.* Crossref
2. *Elder C, Hainline C, Galetta SL, Balcer LJ, Rucker JC. Isolated abducens nerve palsy: update on evaluation and diagnosis. Curr Neurol Neurosci Rep 2016;16:69.* Crossref
3. *Patel S, Mutyala S, Leske D, Hodge DO, Holmes JM. Incidence, associations, and evaluation of sixth nerve palsy using a population-based method. Ophthalmology 2004;111:369-75.* Crossref
4. *Razek AA, Huang BY. Lesions of the petrous apex: classification and findings at CT and MR imaging. Radiographics 2012;32:151-73.* Crossref
5. *Kontzialis M, Choudhri AF, Patel VR, et al. High-resolution 3D magnetic resonance imaging of the sixth cranial nerve: anatomic and pathologic considerations by segment. J Neuroophthalmol 2015;35:412-25.* Crossref