

Ophthalmoplegic migraine and paediatric oculomotor schwannoma



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Title : Ophthalmoplegic Migraine and Paediatric Oculomotor Schwannoma:
Cause or Co-Incidence?

Structured Abstract:

Objective: To report a case of Ophthalmoplegic migraine with Pediatric Oculomotor Schwannoma which is very rare.

Methods: A 16 year old boy admitted as inpatient at our tertiary referral centre and University hospital in South India, with history of recurrent headaches and oculomotor palsy of 14 years duration was evaluated in detail to rule out posterior fossa, orbital fissure, parasellar lesions, granulomatous disorders and aneurysms.

Results: Initial CT Brain revealed a nodular non-enhancing lesion in the interpeduncular cistern, MR Imaging along with CISS 3D sequence done two years later after CT, revealed a small enhancing nodular lesion at the level of midbrain in the interpeduncular cistern at nerve exit level suggestive of schwannoma of third nerve.

Practice: Patient was treated with analgesics, nimodipine and valproate with which there was a partial response. During his subsequent 2 year follow-up, his frequency and severity of attacks had reduced.

Conclusion: Oculomotor nerve schwannomas are extremely rare. Only 12 children under the age of 18, without neurofibromatosis have been

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sufficiently documented. The coexistence of OM and Oculomotor schwannoma suggests that it is not a coincidence. Awareness and suspicion is required to identify cranial cephalalgia /OM and it warrants thorough investigation to rule out intrinsic lesions mimicking OM.

Implications.

Controversies exist till date regarding etiology, pathophysiology, imaging findings and management guidelines of OM. The rare association reported in our report gives insight into better understanding of the pathophysiology and clinico-radiological correlations in OM.

Keywords: Ophthalmoplegic migraine, Oculomotor Schwannoma

Introduction

Ophthalmoplegic migraine is very rare with annual incidence being 0.7 per million. It most often occurs in infancy or childhood. There are recurrent attacks of headache in association with ophthalmoplegia due to paresis of cranial nerve III, IV, or VI. The episodes of ophthalmoplegia may persist for several hours to several weeks, months, or permanently. Most often it is self-limited condition. Ophthalmoplegic migraine is also recognized as a cranial neuralgia according to 2004 edition of the International Classification of Headache Disorders. ^[1] Paediatric Oculomotor Schwannoma is extremely rare and it can mimic OM. We describe a young boy with recurrent ophthalmoplegic migraine and oculomotor schwannoma in MR imaging.

Clinical Observation

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A 16 year old boy presented with history of left hemicranial headache of 14 year duration. The headache was throbbing, severe associated with drooping of left eye, blurring of left eye vision, photo-phonophobia, nausea and vomiting. The frequency of attacks was around 15 per month each lasting for 3-48 hours. During the attack, he was found to have left sided ptosis, poorly responsive normal sized pupil and mild left elevation and adduction restriction. (Figure 1). There was persistent residual vision loss in left eye with acuity of 6/60. Fundus was normal. There were no other deficits.

Investigations were carried out to rule out posterior fossa, orbital fissure and parasellar lesions. Initial CT Brain revealed a nodular non-enhancing lesion in the interpeduncular cistern, MR Imaging along with CISS 3D sequence done two years later (Figure 3a, 3b) revealed a small enhancing nodular lesion at the level of midbrain in the interpeduncular cistern at nerve exit level suggestive of schwannoma of third nerve. MR Angiography was normal. (Figure 3c). Patient was treated with analgesics, nimodipine and valproate with which there was a partial response. Steroids were not administered. During his subsequent 2 year follow-up, his frequency and severity of attacks had reduced.

Paediatric Oculomotor Schwannoma is present as painless oculomotor deficit or may be asymptomatic and detected incidentally. Its presence with OM poses a question whether it was a mere coincidence or the cause of OM.

Discussion:

Ophthalmoplegic migraine is a rare distinct neurologic syndrome

characterized by recurrent headache and ophthalmoplegia. The third cranial
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nerve is most commonly affected. Most patients recover completely within days to weeks, but a minority are left with persistent neurologic deficits. ^[1] . As per the International Classification of Headache, ophthalmoplegic migraine is defined as at least 2 attacks of “ migraine-like” headache followed within 4 days by paresis of the third, fourth, and/or sixth cranial nerves, including ophthalmoparesis, ptosis, or mydriasis . ^[2] Gap between the onset of headache and the cranial nerve palsy has varied between 2 days and 10 weeks. ^[3] The exact aetiology of this condition remains unknown. Oculomotor nerve compression, ischemia, swelling of the posterior cerebral artery, pituitary swelling, vascular anomaly, benign viral infection, demyelinating neuropathy, activation of trigemino-vascular system are the various pathogenesis implicated.

Diseases such as vascular malformation, granulomatous infections, pituitary apoplexy, sarcoidosis and chronic inflammatory, demyelinating polyneuropathies may have similar clinical presentation like OM. So contrast enhanced MRI and magnetic resonance angiography should be the investigations of first choice for the diagnosis of OM, followed by a careful clinical examination and spinal tap. Sometimes, conventional angiogram may be necessary to exclude an aneurysm. ^[4].

Mark et al. 1998., ^[5] found focal thickening of the nerve in non-contrast studies, and further thickening was present on the contrast-enhanced images in the area of the exit zone of the nerve in the inter peduncular cistern. Carlow studied the magnetic resonance scans in six patients diagnosed with OM and did a retrospective literature survey in 17 patients

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with OM, all of whom showed thickened ipsilateral oculomotor nerves at the midbrain exit in noncontrast T 1 -weighted images. Contrast T 1 -weighted magnetic resonance scans showed enhancement of the ipsilateral oculomotor nerves. ^[6] Many cases show improvement in the enhancement of cranial nerve III with resolution of the symptoms, but the timing and degree of resolution has not been consistent in reports. Contrast enhancement on MRI is not a sine qua non for the diagnosis of OM.

Gelfand AA et al., 2011 ^[1] systematically reviewed all cases of OM in literature between 1995 to 2010. There were a total of 80 cases . The median age at the time of the first ophthalmoplegic migraine attack was 8 years (3-16 years) . The third cranial nerve was involved in the vast majority of cases (83%), sixth cranial nerve was involved in 20% and the fourth nerve in 2% of cases. The interval between headache onset and ophthalmoparesis ranged from immediate to up to 14 days. The ophthalmoplegia tended to last longer (2 to 3 weeks to 2 to 3 months) . In 54%, persistent deficits were observed. Of 52 patients who had a contrast brain MRI during an acute attack, 75% had contrast enhancement of the third nerve and 76% had nerve thickening. There was a benefit from corticosteroid treatment in 54%.

Schwannomas are benign peripheral nerve sheath tumours with great propensity to arise from vestibular nerves. Oculomotor nerve schwannomas are extremely rare. There are only 40 cases reported in the literature. Only 12 children under the age of 18, without neurofibromatosis have been sufficiently documented. ^[7]

In 1982, Leunda et al. [8] reported a case in an 11-year-old boy whose tumour was resected en bloc and ranked as the largest oculomotor nerve schwannoma documented at that time, with a 55-mm diameter. Since then, another 11 histologically proven paediatric cases have been described successively in the literature. The average diameters of the paediatric tumours size is 19.5 mm. Oculomotor nerve paresis was the most common neurological sign and a variable degree of oculomotor nerve dysfunction, including ptosis, diplopia, or dilated pupil, was present in all but one. Duration of preoperative symptoms and signs ranged from 2 weeks to 12 years. Authors opine that lesion size did not correlate with the degree of oculomotor nerve deficit. Ipsilateral ophthalmoplegic migraine was found in two cases of cisternal microlesions involving the initial prepontine segment of oculomotor nerve. [7]. Total removal of schwannoma usually resulted in severe postoperative parent nerve paresis. Surgical treatment was indicated only for large tumours that presented in association with consciousness disturbance, other cranial nerve signs, or hemiparesis due to mass effect, or in cases where the lesion showed malignant features with rapid enlargement.

Murakami et al., 2005 [9] described a case of an 11-year-old girl with oculomotor nerve schwannoma who had been suffering from symptoms mimicking OM. Her attacks became more frequent and were not controlled by medication. After surgery, the frequency of OM attacks reduced. This was the first report to describe a pathologically confirmed case of oculomotor nerve schwannoma mimicking OM. Riahi A et al., 2014 [10] described a 12 year old girl with recurrent painful ophthalmoplegia, who on examination <https://assignbuster.com/ophthalmoplegic-migraine-paediatric-oculomotor-schwannoma/>

was found to have left oculomotor palsy. Her third MRI focusing on third nerve revealed schwannoma of the oculomotor nerve in left cisternal portion. Kawasaki et al., 1999 ^[11] reported a case of the coexistence of OM and ipsilateral 3rd nerve schwannoma . The authors had an acute and a follow-up MRI during a recovery phase and noticed no difference. The coexistence of two rare conditions suggests that a mere coincidence is unlikely. It has been proposed that repeated inflammation could lead to a demyelination/remyelination process with Schwann cell proliferation and onion bulb formation.

There are no published treatment trials for ophthalmoplegic migraine. Oral steroids may be of possible benefit in treating acute exacerbations based on available case series. Prompt steroid therapy at the time of attack might minimize permanent sequelae of OM, including residual weakness of the third cranial nerve and pupillary dysfunction. Bharucha et al., 2007 ^[4] state that various treatments have been described, including steroids acutely and flunarizine, acetazolamide, propranolol, cyproheptadine, or verapamil prophylactically. OM is a heterogeneous disorder that needs adequate scrutiny, periodic magnetic resonance monitoring, and clinical correlation before we can conclude and draw management guidelines.

Conclusion: Awareness and suspicion is required to identify cranial cephalalgia /OM as it is a rare entity and diagnosis even by headache experts is made by exclusion. It warrants thorough investigation to rule out intrinsic lesions mimicking OM. The rare association reported in our report gives insight into pathophysiology and clinico-radiological correlations in OM.

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