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Sir,

Horner's syndrome and sixth nerve palsy due to herpes zoster ophthalmicus arteritis

The association of sixth nerve palsy with ipsilateral third-order neuron painful Horner's syndrome has exquisite localizing value in the posterior cavernous sinus, also known as Parkinson's syndrome.[1] The syndrome has been observed mainly in aneurysms in and around the posterior cavernous sinus presenting acutely with variable pain in and around the orbit. We report for the first time MRA documented cerebral angitis with herpes zoster ophthalmicus (HZO) as a cause of ipsilateral painful Horner's syndrome with sixth nerve palsy.

Case report

A 24-year-old female patient presented to us with complaints of acute and rapidly progressive drooping of the right upper lid with moderate pain, double vision, and limitation of abduction in the right eye. This was preceded 6 weeks earlier by multiple painful vesiculobullous eruptions on the right side of the forehead and dorsum of the nose, with associated moderate-grade fever and dull ache in the right orbital region, which subsided without treatment. On presentation, the patient had hyperpigmented small deep scars in the distribution of the right frontal and nasociliary nerve, characteristic of HZO. There was no facial anhidrosis. The unaided visual acuity in both eyes was 20/20. There was a face turn to the right. Palpebral fissure width was 7.5 mm in the right eye and 9 mm in the left eye. Hertel's exophthalmometry was normal. There was a 2 mm ptosis in the right eye with good levator function. An anisocoria of 1.5 mm was present (right pupil: 2 mm, left pupil: 3.5 mm), which increased in the dark with no light near dissociation. The ocular motility is shown in Figure 1. Dry eye was noted in the right eye, proven by tear film breakup time and Schirmer's testing. Corneal sensations and blink reflex were normal. The rest of the neuro-ophthalmic examination, including colour vision, visual fields, fundoscopy, and other cranial nerves, was normal.

Investigations revealed normal blood counts except the ESR, which was 70 mm in the first hour. Other tests including BP, GTT, X-ray chest, VDRL, 'C' reactive protein, Rh factor, and ANA were all negative.

Gadolinium-enhanced MR imaging and MR angiography suggestive of HZO arteritis are shown in Figure 2. The patient was started on oral steroids

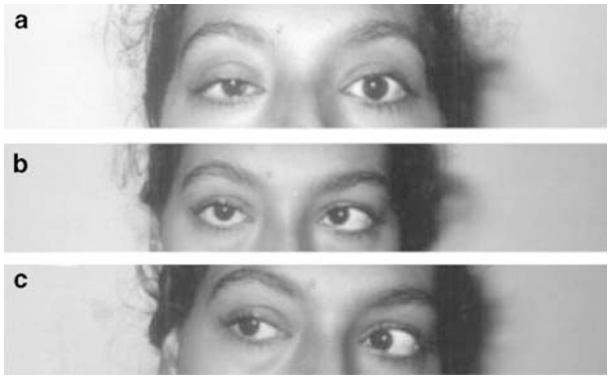

Figure 1 (a) Patient showing mild right upper lid ptosis with esotropia, at presentation. (b) Patient showing severe limitation of abduction of the right eye. The forced duction test, active forced generation test, and Tensilon test were negative. (c) Same patient sixth weeks later showing recovery of ptosis and 6th nerve palsy.

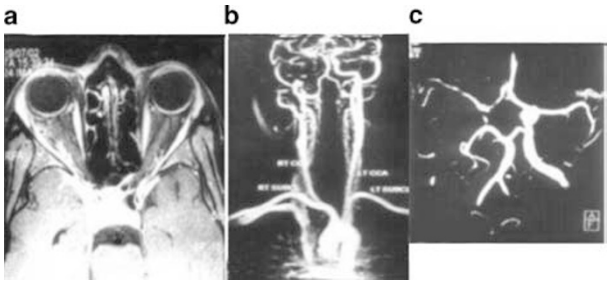

Figure 2 (a) Gadolinium enhanced MRI showing thickening and near total occlusion of distal right internal carotid artery. (b) MR angiography showing narrowing of the right internal carotid artery and external carotid artery. There was no evidence of dissection. (c) MR angiography showing narrowing of right anterior cerebral artery and middle cerebral artery.

(Prednisone 80 mg/day in tapering doses 10 mg/week) and oral Acyclovir (800 mg, 5 times/day for 3 weeks). There was a gradual improvement over the next 3 weeks in the ocular motility and ptosis (Figure 1c). There was almost complete recovery of the sixth nerve with minimal abduction deficit, but Horner's did not recover completely.

Comment

HZO involves the first division of the trigeminal nerve. Ocular involvement occurs in about 50% of the cases. Ocular complications in HZO could have many mechanisms like presumed viral spread, nerve damage, ischaemic vasculitis, and an inflammatory granulomatous reaction.

Varicella zoster necrotizing angitis of cerebral vessels, usually occurring in older age and immunocompromised situations, is presumed to be a viral-induced vasculitis spreading along the Gasserian ganglion. It occurs weeks to months subsequent to the eruptions.[2] It may present with transient ischaemic attacks, cerebral infarction or strokes in evolution, internuclear ophthalmoplegia, and temporal arteritis. The most common presentation is contralateral hemiplegia. Vasculitis is characterized by vascular wall necrosis, inflammatory exudates and development of giant cells in medium- and small-sized vessels.[3] Pathogenesis of vasculitis remains obscure, but has been found to be associated with immune complexes, mechanical factors, and infection by varicella zoster virus. The combination of periorbital pain and postganglionic Horner's syndrome indicates a lesion along the internal carotid artery.

Ophthalmoparesis occurs far more frequently within a week or two after the onset of the rash in about 30% of cases of zoster. Isolated or multiple cranial nerves may be involved. Horner's syndrome has rarely been reported with HZO. Smith *et al*,[4] reported a case of HZO that progressed to sixth nerve palsy and then to Horner's. However, no neuroimaging was performed and direct damage by the virus was implicated as the cause. Tola-Arribas *et al*[5] reported a case of Horner's with simultaneous involvement of the third and sixth nerves. The neuroimaging was negative and the causative mechanisms of the lesion were discussed. With thoracic zoster,[6,7] Horner's has been described in five reports; although it has not been localized, it could be second-order neuron involvement. We report for the first time MRI-documented cerebral angitis involving internal carotid artery, external carotid artery, middle cerebral artery, and anterior cerebral artery of the ipsilateral side due to HZO as a cause of third-order neuron Horner's and sixth nerve palsy in a young woman.

The unusual features in this case include relatively young age, inflammatory involvement due to vasculitis, and associated dry eye. Cerebral angitis can have ischaemic sequelae; however, in this case the underlying pathology seems to be inflammatory presenting as Parkinson's syndrome. Dry eye could be either due to ocular involvement due to HZO or due to damage to oculosympathetic fibres to the lacrimal, and accessory lacrimal gland. The role of sympathetic supply in the tear secretion and dry eye remains speculative and poorly understood.

Since the advent of neuroimaging especially MRI and MRA, isolated painful Horner's syndrome has been described with many pericarotid syndromes like reactive arteritis,[8] dissection,[9] fibromuscular dysplasia,[10] and occlusion of extra-cranial internal carotid.[11] However, herpes zoster vasculitis does not find mention in any report.

Herpetic cerebral angitis should always be considered in the differential diagnosis of pericarotid syndrome and Parkinson's syndrome and requisite neuroimaging performed to assess the extent of involvement, as cerebral angitis/vasculitis has devastating consequences for the eye and can even be fatal. The pathogenetic mechanisms for vasculitis, clinical presentation, treatment, and prognosis are poorly understood.

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Sir,

Uveitis–glaucoma–hyphema syndrome and systemic anticoagulation

Uveitis–Glaucoma–Hyphaema (UGH) syndrome is more commonly associated with anterior chamber intraocular lenses (IOLs),[1] and less commonly with posterior chamber IOLs.[2–4] We report a case of UGH syndrome in a patient with posterior chamber IOL who was on treatment with warfarin.

Case report

An 83-year-old male initially presented to eye casualty with an embolic left inferotemporal branch retinal occlusion. He had undergone bilateral extracapsular cataract surgery with posterior chamber IOLs 18 years previously. Both IOLs were noted to have subluxed inferiorly (Figure 1a**),** with iris atrophy superiorly in the left eye and a mild anterior uveitis. He had suffered a pulmonary embolus 1 month prior to his attendance and was on warfarin. His International Normalized Ratio (INR) was 1.7 (therapeutic range 2.0–3.0).

After 10 days, he reattended with loss of vision to perception of light in his left eye. Examination revealed a total hyphaema with an intraocular pressure (IOP) of 30 mmHg (Figure 1b**)**. The INR was elevated at 3.8. He was commenced on topical and systemic ocular hypotensives. After 2 days, his hyphaema remained unchanged but the IOP had normalized. The hyphaema resolved over 6 weeks and acuity improved to 6/9. IOP remained normal off treatment, but a residual anterior uveitis required topical corticosteroids. At review 3 months after the initial presentation, his INR was 2.1, the uveitis had virtually resolved and topical steroids were gradually stopped. His warfarin was stopped 1 month later and no further problems relating to UGH syndrome

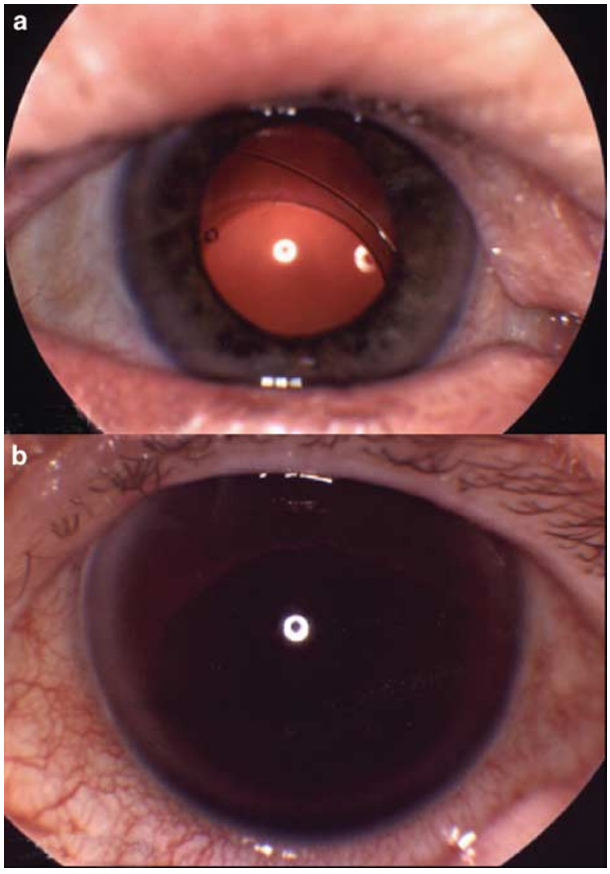

Figure 1 (a) The IOL is seen subluxed inferiorly; (b) 70% hyphaema in the same eye.