Bilateral acute keratouveitis in leptospirosis: A new entity

Dear Editor,

A 25-year-old male presented with history of acute onset of redness, photophobia and decrease in vision in both eyes two days ago. On ocular examination, visual acuity was 20/60 in both eyes. Anterior segment examination showed marked ciliary injection with acute central keratitis, 2×2 mm in size and minimal stromal edema of adjacent cornea. Slit-lamp examination showed fine keratitic precipitates with maximum concentration on the back of area of keratitis with preserved corneal sensation. Rest of the ocular examination was normal. He also had acute onset fever with severe generalized myalgia of three days' duration. Systemic symptoms included proximal muscle weakness.

Based on systemic features leptospirosis was suspected by the physician and the diagnosis was established by positive IgM and IgG serology for anti-leptospiral antibodies. Other laboratory findings, which were also consistent with the diagnosis included proteinuria, raised erythrocyte sedimentation rate, leucocytosis and raised liver enzymes.

He was treated with intravenous ceftriaxone 1g twice daily for a week. Ocular treatment included topical 0.1% dexamethasone six times and 0.3% ciprofloxacin eight times daily. He responded well to treatment and was symptom-free in a week. Topical medications were stopped by the second week, however, macular grade opacity had formed at the area of keratitis.

Leptospirosis presents as a biphasic illness. The first phase is the septicemic phase characterized by nonspecific features like fever, headache, myalgia and conjunctival congestion, while the second phase is the immune phase.1

During the acute phase, which is due to active bacterial invasion, ocular manifestations include subconjunctival hemorrhage, retinal hemorrhage, papillitis, however uveitis does not manifest in the acute phase.2 While, in the immune phase, the ocular signs include acute non-granulomatous panuveitis, vasculitis.2,3 Corneal involvement in leptospirosis has been described only in one report, in the form of interstitial keratitis.1 In tropical countries leptospiral uveitis is one of the commonest causes of hypopyon uveitis along with ankylosing spondylitis and Behçets disease.3

Leptospirosis is transmitted through human contact with surface waters or moist soil that harbors Leptospira interrogans.4 In our case, acute keratitis in both eyes can be due to direct inoculation of the organism on the cornea by contact with infected water. The presence of IgG anti-leptospiral antibodies suggests that the patient might have been exposed to leptospirosis in the past and the repeat exposure had mounted the immune response, which was seen in the form of uveitis.

Treatment of systemic leptospirosis includes systemic penicillin, amoxycillin or ceftriaxone.3 The ocular inflammation was treated with low-dose topical steroids and antibiotics, in view of the presence of keratitis and acute phase of the disease.2,3

Acute keratouveitis is considered to be caused by the Herpes simplex virus unless proved otherwise.6 It can involve any corneal layer but is commonly associated with stromal keratitis which is usually diffuse but rarely can be sectoral.6 There is decreased corneal sensation and dendritic pattern corneal ulcer may also be present. However, Herpes zoster keratouveitis is differentiated by the associated dermatomal distribution of shingles. Interstitial keratitis is the commonest form of corneal involvement in keratouveitis and is associated with tuberculosis, leprosy and syphilis.7

Our patient had systemic features of leptospirosis without dendritic pattern of keratitis and preserved corneal sensations. There was no corneal vascularization to suggest interstitial keratitis. Leptospirosis has been known to cause acute epithelial keratitis in animals like horses etc. and an immunologic component has been suggested as a possible reason in their pathogenesis. The immunologic component cannot be ruled out in our case.8

In conclusion, leptospirosis can have diverse clinical presentations and a high degree of suspicion is required for its diagnosis. Letospiral keratouveitis is a relatively new clinical entity to be kept in mind in patients with systemic features.

Arvind Gupta, MS; Datta Pandian Gulnar, MBBS; Renuka Srinivasan, MS; Subashini Kaliaperumal, MS, FRCS
Department of Ophthalmology, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry - 605 006, India.
E-mail: arvind_ophthal@yahoo.co.in

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Congenital or acquired Horner's?

Dear Editor,

It has been estimated that less than 5% of Horner’s syndrome (HS) are congenital in the pediatric population.3 Jeffery et al.
conducted a case series of 31 pediatric patients with HS and reported that 42% were congenital, 15% were acquired without surgical intervention and 42% were after a surgical procedure of the thorax, neck or central nervous system.2

A seven-month-old female baby presented to us with mild ptosis and an apparent enophthalmos (2 mm) of the right eye and right hemifacial anhidrosis. She was born full-term normal vaginal delivery.

On examination she had subtle anisocoria of approximately 2 mm with a right mitotic pupil, (pupil lag+) and brown colored iris in both eyes [Fig. 1]. She could follow light in all directions with each eye individually. Abdominal examination by pediatrician was normal.

A provisional diagnosis of HS was made and the same was confirmed by instillation of topical 0.5% apraclonidine eye drop which showed complete elimination of ptosis in the right eye and dilatation of the pupil. Further investigations were done to identify a possible cause for Horner’s syndrome. X-ray chest posterior-anterior view showed no mass in the thorax. Magnetic resonance imaging (MRI) of the neck showed a heterogeneously enhancing soft tissue lesion involving the root of neck that measured 2.26 x 1.80 cm [Fig. 2]. The lesion was located close to or arising from the right sympathetic chain, which could have been either a lymph node or paraganglioma. The MRI of the thorax and abdomen was normal. Urine vanillylmandelic acid (VMA) assay yielded normal results excluding the possibility of neuroblastoma (32 micro moles/24h of urine) although there are varied reports on the reliability on this test since elevation of urine catecholamine mainly depends on tumor bulk.3 It was opined in consultation with the neurosurgeon to observe the progression and avoid biopsy since the lesion was very close to major vessel and had a high chance of hemorrhage.

Though brachial plexus injury is the most common cause of congenital HS, any lesion along the sympathetic chain may result in HS. Among these lesions, timely diagnosis of primary thoracic or cervical neuroblastoma is very crucial.3,4,5

In our case, the presence of anhidrosis indicated preganglionic lesion and MRI showed a mass along the second order neuron between the celsiuspinal center of Budge and Waller and the superior cervical ganglion. Based on radiographic characteristics, the lesion was thought to be either an enlarged lymph node or a paraganglioma. Lymphadenopathy is an unlikely cause for this presentation, as there was no infectious or inflammatory lesion within the draining area of the cervical lymph node in the neck. Normal urinary VMA ruled out neuroblastoma, leaving us with the most probable diagnosis of the mass being nonchromaifin paraganglioma. Nonchromaifin paraganglioma is predominantly a benign tumor arising from paraganglionic cells anywhere in the body. They occur most commonly in the retroperitoneum or head and neck region and are usually not suspected at the time of presentation,6 due to their nonspecific nature of presentation.

Although delivery-related brachial plexus injury is the most common cause of congenital HS, from our case we feel it is important to do imaging of the entire sympathetic pathway, since life-threatening malignancy and abnormalities may be revealed.

Figure 1: Seven-month-old female baby with miosis, apparent enophthalmos of right eye and anhidrosis of right side of face

Figure 2: MRI of the neck showing a heterogeneously enhancing soft tissue lesion involving the root of neck

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