Introduction

Intraocular infestation of Gnathostoma spinigerum is quite rare and about 20 cases have been reported in literature so far. Anatomically, it can involve the eyelids, conjunctiva, cornea, anterior chamber, uvea, vitreous, and retina and can even present as orbital apex syndrome. Subretinal hemorrhages and subretinal tracks have been described after subretinal migration of worm. Here we are reporting for the first time, a case of intraocular Gnathostomiasis presenting with retinal vasculitis.

Case report

A 44 year old Indian male, working in Central Asia for the last 15 years presented to us with chief complaints of sudden onset watering, redness and pain in the right eye of one week duration. On examination his best corrected visual acuity was 6/6 OD and 6/6 OS. IOP was 26 and 16 mm Hg in the right and left eye respectively. Slit lamp examination revealed mild conjunctival congestion, 1+ cells in the anterior chamber, pigment on endothelium and anterior lens capsule in the right eye. Gonioscopy was done which showed the presence of open angles in both eyes with pigments on trabecular meshwork inferiorly in the right eye. Rest of the anterior examination was normal.

Fundus examination of the right eye showed normal disc, macula and vessels in the posterior pole. There was an area of vasculitis in the inferotemporal periphery with cuffing around the arterioles with multiple skip areas in between. There was an adjacent white necrotic area suggestive of retinitis. (Fig 1a, b). A small tinge of hemorrhage was visualized near the area of retinitis. Rest of retina was within normal limits. Left eye examination was also within normal limits. We made a provisional diagnosis of viral retinitis associated with vasculitis and so, investigations were directed on the same lines. Patient was put on symptomatic treatment.

2 days later patient had aggravation of symptoms. Vision had fallen to 6/18 and IOP had risen to 30 mm Hg in the right eye. Repeat examination showed worsening of anterior uveitis with 2+ cells in the anterior chamber. A live, brown, coloured, cylindrical, sluggishly motile worm measuring about 5 mm was seen in the supero temporal quadrant of the anterior surface of iris in the right eye (Fig 2 a, b); a small iris hole was appreciated on the temporal aspect of the iris. Patient was put on pilocarpine eyedrops and same day the worm was removed from the anterior chamber under topical anaesthesia.(Fig 3).

Histopathological evaluation of the worm showed it to be a third stage, male larva of Gnathostoma spinigerum, having the characteristic head bulb with 4 rows of hooklets and spines on the body surface.(Fig 4 a, b).

Except for a slightly raised C-reactive protein, all the investigations of the patient were within normal limits, with no evidence of eosinophilia; this could probably have been the result of the strict intraocular location of the worm, which could lead to decreased systemic response. Our patient denied any history of creeping, migratory rash or swellings. However, he had a definite history of frequent travel outside the country and gave a history of frequently consuming pickled fish both at home and abroad. We presumed he acquired the larva infestation because of that habit.

Postoperatively the patient was put on intensive topical steroid and mydriatics and his vision improved to 6/6 (B) and IOP came down to 22 mm Hg OD with persistent posterior synechiae in the nasal and temporal quadrants. The retinal vasculitis had disappeared and the area of retinitis had become a scar.

This case is being reported because to the best of our knowledge, this is the first case where retinal vasculitis and retinitis was associated with ocular Gnathostomiasis and also because there is a great need to generate health awareness particularly among the rural population who frequently consume pickled meats and fish, and drink fresh water from the rivers and ponds without processing it.

Discussion

Gnathostomiasis is a parasitic roundworm of domestic and wild carnivorous animals like cats, dogs and tigers . It was first isolated from stomach of tiger by Richard Owen in 1836. The definitive hosts excrete the eggs in faeces which then mature into first stage larvae in water. These are then eaten by Cyclops in which second stage larvae develop. These infected cyclops are eaten by the intermediate hosts like fresh water fish, frogs and snakes. Within the intermediate hosts, the infected larvae may in turn be eaten by pigs, poultry and other such animals. There are several species of Gnathostoma which infect humans, of which Gnathostoma spinigerum is the commonest. Humans serve as accidental hosts and are infested by eating raw, undercooked, pickled fish, poultry, pork or drinking infected water contaminated with the larva of Gnathostoma. Rarely, the larval worm can penetrate through the skin. In the stomach wall of the definitive hosts,
the third stage larvae attain maturity; however humans are
dead end hosts, therefore the larvae do not reach maturity
but instead migrate throughout the body (cutaneous
disease) or can become encysted in any tissue(visceral
disease), hence the myriad presentations. Human infection
can involve the GIT, CNS, genitourinary, respiratory system,
skin and eyes. Ocular involvement can occur many years after
being infested, and may sometimes be the only presenting
sign of the infestation, as in our patient.

Gnathostomiasis is endemic in Southeast Asian countries,
particularly Thailand where it happens to be the most
common tissue parasitic infestation. Though reported,
intraocular Gnathostoma infestation is a rare presentation.
The first report of intraocular Gnathostoma infestation was
reported from Thailand. Though India is an endemic zone,
there are only a few reports of intraocular Gnathostoma
infection from India. In India, the first ever case of intraocular
Gnathostoma infestation was reported in 1945.

Diverse presentation of ocular infestation has been reported
like orbital cellulitis, anterior uveitis, iris holes, secondary
apex syndrome. It is believed that the larva gains entry into
the eye through the optic nerve or by directly penetrating
the sclera. However in our patient there were no retinal holes
or any tract near the optic disc. The worm could have gained
entry into the eye by penetrating the sclera and choroid in an
anterior location. Most of the earlier reports have remarked
about multiple iris holes and fenestrations, yet there was
only a solitary iris hole in our patient which we believe can
explain the presence of the worm in the anterior chamber in
our patient.

Retinal vasculitis or retinitis has not been described before in
eyes infested with Gnathostoma. Subretinal tracks suggestive
of movement of the worm within the retina or subretinal
hemorrhage secondary to movement trauma or vessel
damage have been reported before. After initial infection, it
may take years for the larva to reach the eye. We postulate
that during this latent period, our patient was sensitized to
the Gnathostoma antigen, and when the worm entered the
eye, a hypersensitivity response was mounted resulting in
retinitis and vasculitis. Thus the vasculitis and retinitis in the
retinal periphery made us tilt our diagnosis more towards
viral retinitis or early acute retinal necrosis and only after
seeing the adult worm in the anterior chamber could we
come to a definitive diagnosis.

Retinal vasculitis has been described in various parasitic
infections like Schistosomiasis, Wuchereria Bancroftii
and Toxoplasmosis, but not in Gnathostomiasis. However
eosinophilic vasculitis involving the skin has been reported
in Gnathostomiasis.

Though there are several tests like ELISA, precipitation and
indirect fluorescent antibody to detect the antibody against
the parasite, definitive diagnosis is based on recovery and
identification of the worm from body tissues. Recently,
diagnostic value of IgG4 antibodies from patients of
Gnathostomiasis has been assessed and recognition of 21
kDa antigenic extract of third stage Gnathostoma larvae by
infected human sera has been found to be a diagnostic
marker for human Gnathostomiasis, with 100% specificity
and sensitivity.

Various case reports of intraocular Gnathostoma infection
have mentioned about the absence of systemic clinical
findings and eosinophilia, just as in the case of this patient;
nevertheless suggestive clinical history, recurrent migratory
swellings and eosinophilia in a patient who comes from an
endemic zone and presenting with ocular symptoms should
arouse a suspicion of intraocular infestation. The raised CRP
may be a contributory evidence for systemic vasculitis for
which we do not have any definite proof.

Till date no antiparasitic drugs have been reported effective
against the intraocular parasite. Hence the cure is based
only upon early identification and removal of the worm
from the eye before it migrates into the CNS and causes
fatal cerebrovascular accidents. Prevention is based upon
avoiding raw, undercooked food and untreated water.
Greater community health awareness about the method of
transmission, disease manifestation and prevention is also
warranted.
References
1) Manson-Bahr PEC, Bell DR,Manson’s Tropical Diseases.9th ed,ELBS:Bailliere Tindall;1987;Appendix II.pp 1348-50.