Orbital Myiasis: An Unusual Presentation of Retinoblastoma

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Summary
A two year old child, presented with parents complaining of maggots with fungating mass in the right eye for two months. On examination, the child was debilitated. He had no perception of light in the right eye and only perception of light in the left eye. The right eye had a huge fungating mass invaded by maggots along with blood clots. The left eye had leukocoria. Ultrasonography of the right eye was not possible due to fungating mass and that of the left eye showed an intraocular mass with calcification. Diagnosis of bilateral retinoblastoma was made. Maggots were removed from the right eye after instillation of lignocaine and turpentine oil. Magnetic resonance imaging suggested extensive mass with cerebral metastasis. Systemic antibiotics, antifungal agents, palliative external beam radiotherapy followed by chemotherapy were given which led to reduction in tumor mass and improvement in the general condition. The mass in the left eye also demonstrated regression. Orbital myiasis in this case was a result of lack of awareness and negligence.

Keywords: Retinoblastoma, Orbital myiasis

Introduction
Ophthalmomyiasis refers to infestations of the eye and/or ocular adnexa. In humans, the orbital form of ophthalmomyiasis is particularly serious. It accounts for less than 5% of the cases of human myiasis. Keyt first reported it in 1900 and later on, Elliot reported it from India in 1910. It can occur in most regions of the world, particularly in rural areas of underdeveloped countries where the standard of hygiene is low and flies abound. Crowded conditions, debility, low socioeconomic status, and poor personal hygiene are the predisposing factors. Orbital myiasis can cause massive destruction of orbital tissues, accompanied by severe inflammations and secondary infections. In extreme cases, the larvae can cause destruction beyond the orbit with mucosal sinuses and intracranial invasion which can be life threatening. Cases of neonatal fatal cerebral myiasis, caused by the penetration of larva through the fibrous portion of the fontanelles, have been reported.

Case Report
The parents of a 2 year old child presented to us with complaints of fungating mass over the child’s right eye for 2 months which was infested with maggots for the past 1 month.

The child was asymptomatic 1 year ago, when his parents noted gradual swelling and protrusion of the right eye associated with watering. This gradually progressed to blood stained discharge from the fungating mass. The child’s health also deteriorated with weight loss. For the past 1 month, his parents noted maggots in the mass. They never consulted any local doctor for any of the complaints. On examination, the child was debilitated. He had no perception of light in the right eye and only perception of light in the left eye. The right eye had a huge fungating mass invaded by maggots along with blood clots. The left eye had leukocoria with pupil reacting to light. (Figure 1).

Ultrasonography of the left eye revealed a mass on the posterior pole with calcification and retinal detachment suggestive of retinoblastoma (RB).

Initial management consisted of manual removal of all maggots (fifty) after instillation of lignocaine and turpentine oil and local sterile dressing with antibiotic ointment. Systemic antibiotics, antifungals and nutritional supplements were initiated as advised by a pediatrician.

MRI showed a 10*7 cm exophytic lesion in the right orbit involving and not separate from the right eyeball, optic nerve and extraocular muscles with involvement of the overlying skin with surface ulceration. The lesion showed internal necrotic areas. There was no evidence of intracranial extension of the lesion or involvement of the maxillary sinus. A 10*10 mm altered signal intensity lesion was noted in the left frontal lobe suggestive of metastasis. (Figure 2).

A CSF tap was ordered which showed no evidence of malignant cells. Bone marrow biopsy was done and found to be normal.

Palliative external beam radiotherapy followed by chemotherapy was planned for the patient. A total dose of 30 Gy static radiations was given divided in 10 equal

Figure 1: (a) Right eye exophytic mass with left eye leukocoria; (b) Maggots after removal.
daily doses. The tumor showed a 60 percent reduction in size at the end of radiotherapy. The patient then received 6 cycles of Carboplatin, Etoposide and Vincristine. There was further reduction in the tumor mass with an improvement in the general condition of the patient. The left eye showed regression on ultrasonography as fundus examination was not possible due to mature cataract. Cataract extraction in the left eye was not planned due to the possibility of inadvertent extraocular spread. A significant weight gain (4 kg) with return of interest in routine activities was noted (Figure 3). The patient was kept on regular follow up. The parents were counseled and educated regarding importance of regular treatment and follow up.

Discussion
The main predisposing factor for the maggot infestation in our patient was probably the large bed of necrotic tissue offered by the malignancy, lack of education and awareness in the parents to be alarmed by the presenting features, lack of self care, communication and poverty. There are other case reports of the orbital myiasis complicating basal cell carcinoma. To the best of our knowledge, there is no reported case of retinoblastoma presenting as orbital myiasis so far.

The NCCT or MRI of orbit and brain is useful for delineating the extent of orbital involvement and excluding the intranasal and intracranial spread.

Management of orbital myiasis may range from simple manual removal of the maggots to destructive surgeries of the globe and orbit. The key step in the management of the less extensive orbital myiasis should be directed towards the manual removal of all the invading organisms. Solutions like hydrogen peroxide, chloroform, ether, ethanol, and turpentine have been used to facilitate the easy removal of the maggots. Controlling the secondary infection in orbital myiasis is of utmost importance. Some cases of extensive orbital myiasis and those associated with malignancy may require exenteration to prevent intracranial extension of the tissue destruction. In recent years, broad spectrum antiparasitic agent Ivermectin has been successfully used as a noninvasive means to treat orbital myiasis. Use of Ivermectin therapy has been recommended prior to surgical debridement to prevent destructive surgery and to reduce the difficulty associated with mechanical removal of the maggots with massive orbital invasion.

Conclusion
RB can present in various forms. Negligence and lack of awareness for treatment in early stage can lead to disastrous complication like orbital myiasis. Parental education and awareness is must to prevent such agony.

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