

Polymerase chain reaction-proven tuberculous anterior segment mass mimicking juvenile xanthogranuloma in a child

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Mass lesions arising from the anterior segment in children involving the iris and ciliary body can be of myogenic, neurogenic, or hematogenic/vascular origin. These include nevi, melanomas, adenoma, adenocarcinoma, cysts, metastatic tumours among others. Multiple iris mass lesions due to tuberculosis in children are rare. We present an uncommon atypical presentation of multiple anterior segment mass lesions referred to us as neoplasia. Although excision biopsy can be diagnostic, it was deferred and anterior chamber tap was done. Aqueous

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cytology was suspicious of juvenile xanthogranuloma (JXG) but polymerase chain reaction (PCR) confirmed tuberculous etiology. Treatment with antituberculous therapy (ATT) and steroids lead to complete resolution of the lesions.

Key words: Iris mass, iris melanoma, juvenile xanthogranuloma, pediatric tumors, tuberculoma

Mass lesions arising commonly from the anterior segment involving the iris and ciliary body in children include nevi, melanomas, melanocytoma, adenoma, adenocarcinoma, retinoblastoma, anterior segment cysts and metastatic tumours.^[1] Tumors of myogenic, neurogenic origin (such as neurilemmoma, neurofibroma) and hematogenic/vascular origin such as hemangioma, lymphoma, and leukemia are also rarely seen.^[2] Multiple iris mass lesions due to tuberculosis especially in children are an uncommon finding. Mass lesions usually need surgical removal for confirmatory histopathological diagnosis and management. We present an uncommon atypical presentation of polymerase chain reaction (PCR) proven tuberculous iris mass mimicking neoplasia in a child and its successful medical management.

Case Report

A 6-year-old Indian female child presented with a history of redness and diminution of vision in the right eye for 1 month. She had been treated elsewhere with one subtenon's (S/T) injection of triamcinolone acetonide and was referred to us as a tumor, due to the development of multiple iris mass lesions. The child was otherwise active with no other systemic complaints, such as fever or loss of weight. She was on topical steroids and systemic antibiotics when she presented to us.

On examination, her best-corrected visual acuity was 20 / 125 in the right eye (OD) and 20 / 20 in the left eye (OS) for distance. Slit-lamp examination of OD showed subconjunctival hemorrhage (due to S/T injection), anterior chamber reaction 2+, mutton fat keratic precipitates (KPs), deep corneal vascularization, posterior synechiae with festooned pupil, Busacca nodules, rubeosis iridis and multiple iris mass lesions [Fig. 1]. The lens showed early cataract changes and intraocular pressure (IOP) was 6 mm of Hg. The left eye was within normal limits. Fundus examination of OD revealed normal disc and macula while the rest of the fundus details could not be made out due to the small pupil. Ultrasonography (USG) of OD showed few low reflective dot echoes in the vitreous, attached retina throughout, normal choroid, and optic nerve head.

Investigations showed positive Quantiferon-TB Gold (QFT-G) and Mantoux (Mx) tests. High-resolution computed tomography (HRCT) scan of the chest showed few thin plate-like areas of atelectasis which involved the right middle lobe and the lingula with insignificant subcentral lymph nodes.

Although multiple iris mass lesions were noted, associated granulomatous inflammation and supportive lab investigations lead to a suspicion of a tubercular etiology. Other iris mass lesions in the pediatric age group were considered as differentials. Excision biopsy was deferred and anterior chamber tap was done under general anesthesia. Aqueous aspirate showed Gram-positive bacilli while PCR testing for mannose-binding protein-64 (MPB64) genome was positive and PCR for Insertion sequence-6110 (IS6110) [Fig. 2] and panfungal genomes were negative.

Aqueous aspirate cytology showed few lymphocytes and occasional neutrophils with the presence of uveal pigments. There were few red blood cells (RBCs) and plasma cells, and numerous histio-monocytic cells, some of them having cytoplasmic vacuoles. A suspicion of juvenile xanthogranulomatosis (JXG), histiocytosis, or a masquerade was considered [Fig. 3]. Although immunohistochemistry testing was planned, it could not be done as cell block showed sparse cells. But with PCR positivity, a course of antitubercular therapy (ATT) and tapering schedule of oral steroids was started with a close watch.

At 1-month follow-up, visual acuity of OD had improved to 20 / 30, IOP was 14 mmHg in both eyes. Slit-lamp examination of OD showed a quiet anterior chamber and resolution of the iris mass lesions [Fig. 4]. Her last follow-up was 4 months after the initial visit. She had completed the course of steroids and is continuing the ATT regimen with stable ocular and systemic conditions.

Discussion

Ocular TB can have varied presentations and mass lesions usually in the posterior segment can be a solitary tubercle or a choroidal granuloma/subretinal abscess.^[1] The common anterior segment presentation is granulomatous anterior uveitis. Iris mass lesions in children can be infective lesions such as due to TB or fungus and non-infective like sarcoidosis, Vogt-Koyanagi-Harada (VKH), and multiple sclerosis.^[2] It can also be a presentation of other neoplastic conditions. This child was referred to us as an iris tumor after the initial treating ophthalmologist found worsening of symptoms and an increase in the size and number of mass lesions with periocular steroid injection.

A possibility of TB as the underlying etiology was considered based on the presentation of granulomatous uveitis with positive Mantoux and QFT tests. Iris mass lesions being larger and multiple, anterior chamber tap was done although imaging and biopsy were deferred. Cytology from aqueous showed findings suspicious of JXG but was not confirmatory. IHC for CD68 staining or S-100 or CD1a helps in confirming or ruling out JXG.

JXG usually has extraocular involvement including nodular lesions^[3] in the skin which were not seen in our child. Ocular involvement is the most common extracutaneous manifestation of JXG. While iris JXG can present with spontaneous hyphema and secondary glaucoma, melanoma mimicking iris JXG in children has also been reported.^[4,5] The child was treated earlier with steroids alone and had worsened which is unusual in the case of JXG. Biopsy of the lesions has its attendant risks and there is a risk of bleeding and even loss of an eye. Before considering a biopsy of the lesion, based on aqueous positive PCR-MPB64,^[6] the child was started on ATT. Fungal granuloma was a differential, considering the worsening of ocular lesions with steroids alone, but aqueous did not reveal any KOH filaments, and PCR for fungal genome was negative. Iridociliary TB in adults mimicking melanoma has been noted,^[7] but multiple iris masses with suspected JXG based on equivocal histopathological findings are rare.

Conclusion

Our case demonstrates that TB can present with multiple large iris mass lesions, and can mimic iris tumors in children.

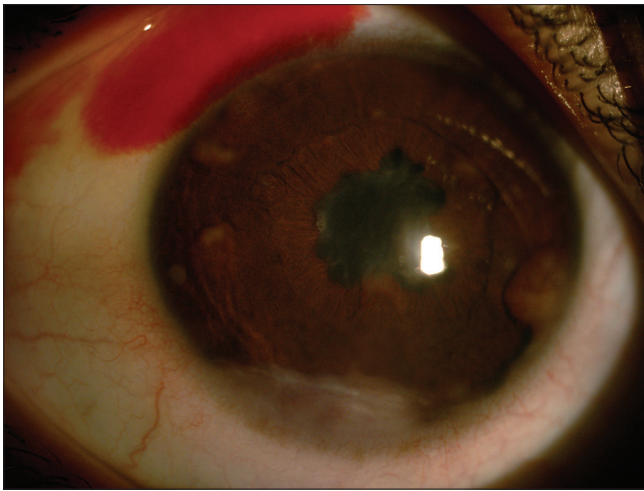


Figure 1: Slit-lamp picture of the right eye showed subconjunctival hemorrhage, corneal vascularization, mutton fat keratic precipitates, Busacca-like nodules, and rubeosis iridis. The mass lesion is noted over the iris between 3–4 clock h, two others at 9 and 10 o'clock positions. A festooned pupil with Peripheral anterior synechiae (PAS)/scarring noted at multiple clock hours

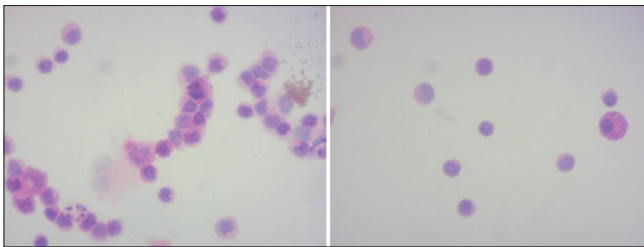


Figure 3: Cytology from the aqueous tap aspirate showed few lymphocytes and occasional neutrophils with the presence of uveal pigments. There were few red blood cells and plasma cells, and numerous histio-monocytic cells with some of them having cytoplasmic vacuoles

Aqueous testing can confirm the etiology and avoids unnecessary excision biopsy, especially in a child. In our case, suspicious cytology of JXG from aqueous lead to a diagnostic dilemma, although PCR clinched the diagnosis. Appropriate ATT helped in complete resolution of the lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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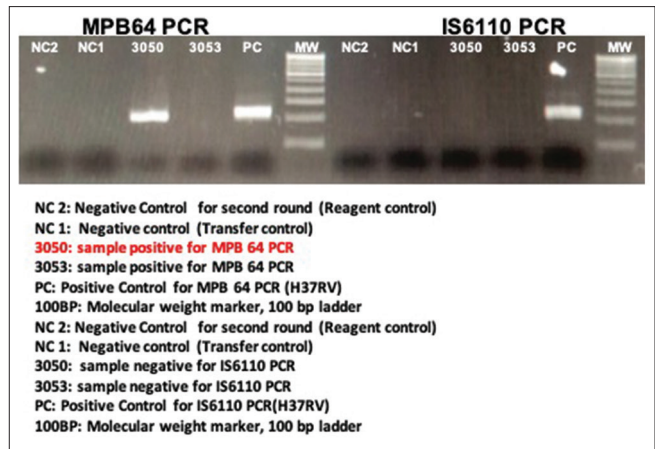


Figure 2: Agarose gel electro photogram showing II round amplified products of nested PCR targeting *MPB64* and *IS6110* gene of *Mycobacterium tuberculosis* complex genome

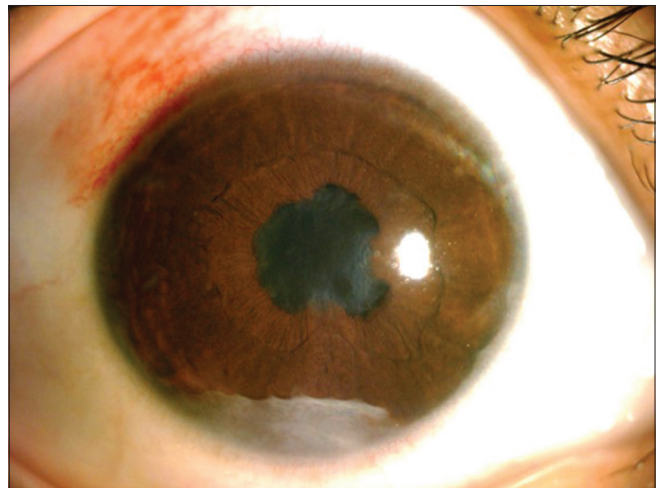


Figure 4: Posttreatment slit-lamp pictures showing improvement complete resolution of the iris mass lesions

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