

Adult presentation of retinoblastoma in Saudi patient

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Abstract:

To describe an unusual case of retinoblastoma in adult Saudi. A 21-year-old Saudi male developed acute unilateral visual loss due to a large intraocular lesion in his left eye. Within five days of presentation, the tumor enlarged rapidly causing angle closure glaucoma and orbital cellulitis like picture. The patient underwent enucleation and the histopathology specimen confirmed the presence of an undifferentiated type tumor cell that was consistent with retinoblastoma with choroidal and optic nerve invasion. Retinoblastoma should be considered in the differential diagnosis of an intraocular mass in adults. To the best of our knowledge, this is the first reported case of an adult presentation of retinoblastoma in Saudi patient.

Keywords:

Adult retinoblastoma, enucleation, glaucoma, orbital cellulitis, retinoblastoma, vision loss

INTRODUCTION

Retinoblastoma is the most common intraocular malignancy in childhood and infancy. Approximately 90% of cases are diagnosed before five years of age.^[1] Adult-onset retinoblastoma is extremely rare and hence is usually not considered in the top differential diagnosis of adults presenting with retinal or intraocular lesions. As per literature review, only 45 cases was reported till date^[2-6] and the oldest report was presented in 1919 from a 20-year-old white girl.^[2] The oldest reported age of RB in the Saudi population was 12 years.^[7] We present a histopathologically proven case of retinoblastoma in an adult Saudi male. This is likely the oldest presentation of retinoblastoma among Saudi population published to date.

CASE REPORT

A 21-year-old healthy male presented to the emergency department with a history of acute painless visual loss in his left eye for one month duration that was associated with redness. The patient was using 1% prednisolone acetate eye drops prescribed elsewhere. On examination, visual acuity was 20/20 in the

right eye and light perception in the left eye. The right eye was unremarkable. On examination, intraocular pressure (IOP) in the left eye was 16 mmHg, the cornea was clear, the anterior chamber did not show any active cells, the lens was clear and displaced. Fundus examination indicated a whitish reflex with a large intraocular mass pushing the iris-lens diaphragm anteriorly and superiorly [Figure 1a-c]. B-Scan ultrasonography revealed a large Choroidal lesion superiorly [Figure 1d]. This lesion measured 25.1 mm at the base and 15.0 mm in depth with a secondary total retinal detachment. A-scan ultrasonography indicated a lesion that was 16.8 mm in depth and low internal reflectivity with minimal vascularity. Magnetic resonance imaging (3 Tesla) of the globe and brain is presented in [Figure 1e]. The differential diagnosis included amelanotic melanoma, Metastatic carcinoma and intraocular infection. On the following day the patient developed severe pain in the left eye secondary to elevated IOP of 60 mmHg with no light perception vision. The anterior chamber was shallow and the patient was admitted and managed with antiglaucoma medications. Over the subsequent two days the patient developed cells resembling tumor cells in the anterior chamber with severe conjunctival chemosis lid edema with restriction extra ocular muscle movement which clinically was consistent

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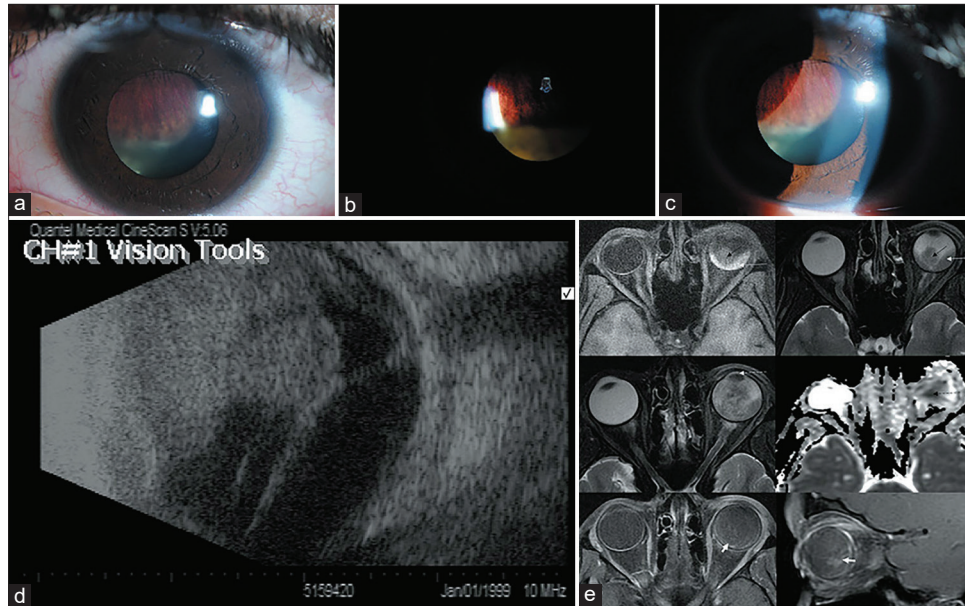


Figure 1: (a) SLE picture showed quite eye, anterior chamber did not show any active cells, the lens was clear and subluxated; (b and c) fundus showed a whitish reflex with large intraocular mass that pushing the iris lens diaphragm forward superiorly; (d) B-scan showed large choroidal lesion detected superiorly. This lesion measured 25.1 mm in base and 15.0 mm in depth by transverse there was secondary total retinal detachment; (e) MR Imaged at 3Tesla. Upper left; Axial T1-weighted image delineate the intraocular mass (black arrow) at the posterior globe near the optic nerve head in the background of hyper intense exudative retinal detachment (white arrow). Upper right and middle left; axial T2-weighted fat suppressed image shows a large hypo-intense mass at the postero-inferior left globe (black arrow) showed numerous punctate foci of signal loss presumably tiny calcification with the retinal leaves are elevated with subretinal fluid of hemorrhagic nature (white arrow) with marked narrowing of the anterior chamber (white dashed arrow). Middle right; ADC map shows lower signal measured ADC of $0.79 \times 10^{-3} \text{ mm}^2/\text{s}$ at the central mass markedly lower values with sharp contrast between the cellular mass and the subretinal exudate at the same section (black dashed arrow) with some degree of globe warping. Lower left, lower right; axial and sagittal fat suppressed contrast-enhanced T1-weighted image shows faint, contrast enhancement as plaque-like area (solid white arrow)

with orbital cellulites [Figure 2a and b]. Laboratory tests ruled out any infection. A repeated ultrasound showed extension of medium low reflective choroidal mass with markedly thickened sclera and T-sign posteriorly. There was suspicion of early sclera perforation, based on that the patient underwent urgent enucleation with silicone ball implantation. The results of histopathology specimen confirmed the presence of an undifferentiating cell type consistent with retinoblastoma with choroidal and optic nerve invasion [Figure 3c-g]. The patient was referred to oncology for chemotherapy and radiotherapy with the possibility of orbital metastasis.

DISCUSSION

The findings of the clinical and diagnostic image tests of this case are compatible with previous reports. The most common causes of secondary glaucoma in retinoblastoma is iris neovascularization (NVI) followed by pupillary block secondary to anterior displacement of the lens-iris diaphragm and tumor seeding of the anterior chamber.^[8] We believe that our patient had sudden increased intraocular pressure due to rapid growth of the tumor that caused anterior displacement of the lens-iris diaphragm with elements of pupillary block in addition to tumor seeding in the anterior chamber. Our case developed tumor related orbital cellulitis. In fact, orbital

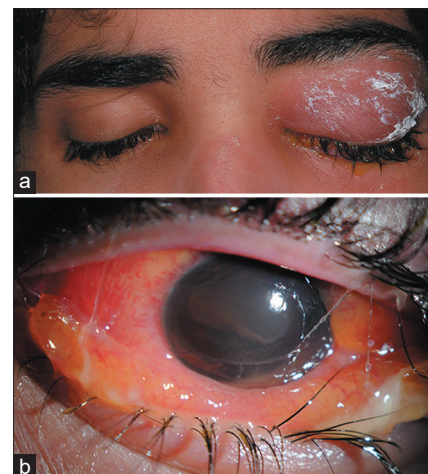


Figure 2: (a) Lid edema and frozen globe; (b) tumor cells in the anterior chamber with severe ciliary congestion and lid edema

cellulitis associated with retinoblastoma is uncommon and is characterized by noninfectious inflammation of the periorbital structures. The underlying mechanism is thought to be necrosis of the intraocular tumor, leading to intraocular and periorbital inflammation.^[9,10] Histopathology of the enucleated specimen in the majority of the reported adult retinoblastoma cases had differentiated, showing Flexner-Winter Steiner

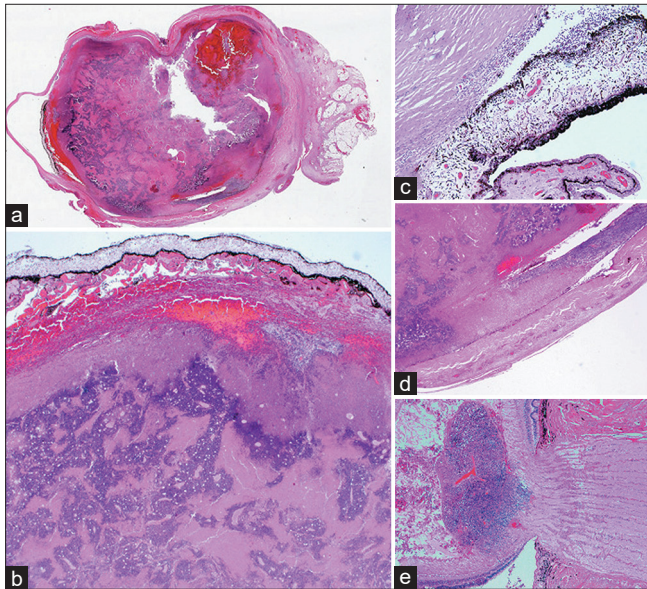


Figure 3: (a) Scanned slide showing a whole globe with an extensive necrotic tumor filling the posterior cavity. (b): The same tumor behind the iris tissue with extensive necrotic areas and smaller viable basophilic tumor areas (original magnification 200 Hematoxylin and Eosin); (c) The tumor showing anterior chamber seeding with angle invasion (original magnification x100 Hematoxylin and Eosin); (d) The tumor showing massive choroidal invasion (original magnification $\times 40$ Hematoxylin and Eosin); (e) The pre-laminar optic nerve invasion (original magnification $\times 40$, Hematoxylin and Eosin)

rosettes.^[6,11] In our case, the tumor was classified as undifferentiated retinoblastoma with no rosettes and fleurettes with massive necrosis and hemorrhage. In conclusion, although retinoblastoma in general is rare among adult population, it should be considered in the differential diagnosis of sudden

onset intraocular mass with unusual clinical presentation in Saudi adults.

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Conflicts of interest

There are no conflicts of interest.

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