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Retinoblastoma in Phthisis Bulbi

Medical Science

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INTRODUCTION:

Phthisis bulbi represents an ocular end stage disease characterized by atrophy, shrinkage and disorganisation of the eye and intraocular contents. Phthisis bulbi is not uncommonly encountered in children, one of the most important causes being retinoblastoma. The case under study guides us, as to the approach to such a case.

CASE REPORT:

A 14 year old girl with right phthisical eye and a normal left eye presented to our hospital with the chief complaint of redness of RE with an unusual prominence that developed over the past 1 month within the otherwise phthisical eye. The RE ended being phthisical when she was 4 years old after an episode of redness, watering and ocular pain as informed by her parents. There was no history of trauma or any other ocular or systemic illnesses. The patient was using an artificial eye for cosmetic rehabilitation since last 5 years. The patient was born out of second degree consanguineous marriage. The parents of the child and an elder sibling were normal and there was no family history of retinoblastoma.

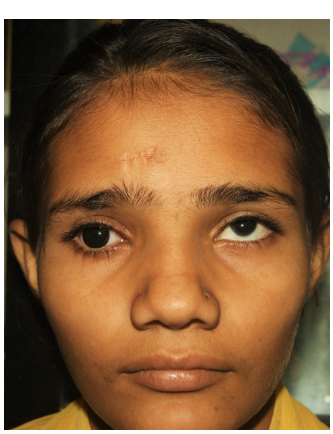
Upon examination, the general examination was normal and the vitals were within normal limits.

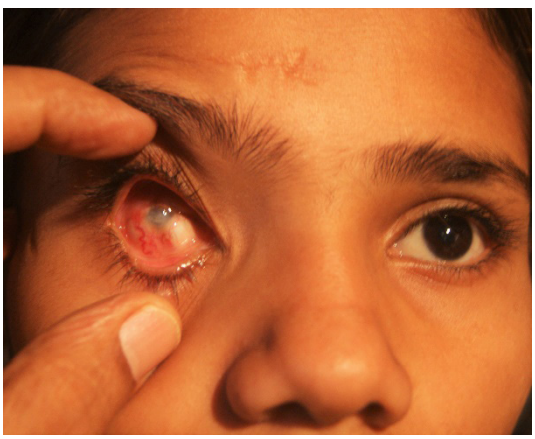
Upon ocular examination

V.A: RE: No PL

LE: 6/6

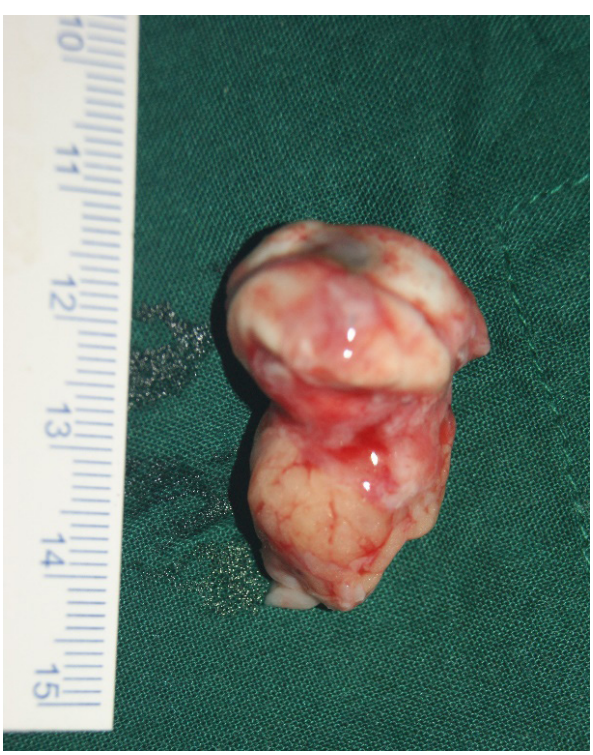
The RE was fitted with an artificial eye. Upon removal the underlying eye was phthisical with conjunctival and ciliary congestion and a very prominent episcleral vessel, with an elevation in the inferonasal quadrant (approximate size ~3*3 cm). The cornea showed diffuse haze and rest of the anterior and posterior segment structures were not seen. The anterior and posterior segments of the left eye were within normal limits.



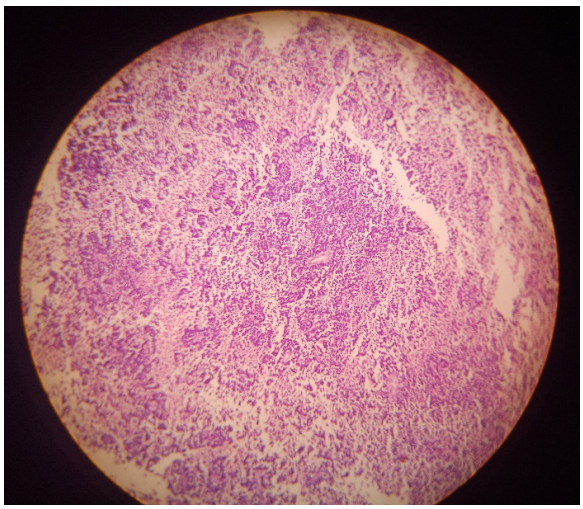


The ultrasonography of RE showed disorganized intraocular contents with no other remarkable finding with AL~12mm, whereas the LE USG was within normal limits with AL~23mm.

A probable diagnosis of retinoblastoma was made and the RE was enucleated and the specimen sent for histopathology examination. The enucleated eye showed a fusiform swelling involving the optic nerve.



The histopathology examination confirmed the diagnosis as retinoblastoma.



The patient was referred to tertiary cancer care centre for further evaluation and management.

DISCUSSION:

Reviewing the literature one finds that regression of retinoblastoma is not very common, but as STEWARD [10] SMITH AND ARNOLD rightly observed that in some non-familial unilateral regressed retinoblastoma cases with phthisical eyes it might have been overlooked, otherwise if all phthisical eyes be examined then the incidence might be even more significant. All the same the end result of the regression is not always a phthisical eye. The following modes of regression are observed in literature:

(i) Phthisical eye following pain and redness (? inflammation) in the eye - DeKLEIJN, [3] KNEIPER [2] DAS [7] , STEWARD ET AL [10] CARBAJAL [1] .

(ii) The tumour gets calcified

(iii) The previous tumour site shows choroidal atrophy with a small grey, green or pink area showing calcification - HINE [5] .

The diagnosis of regression has been made either (1) on clinical grounds and follow up - KNEIPER [5] STALLARD [8] , HINE [5] CARBAJAL [1] or (2) it has been made from family history e.g. if either of the parents have a phthisical eye with suggestive regression and if the children develop retinoblastoma, then the diagnosis of regression in parents becomes certain - SOVIK [9] , STEWARD ET AL [10] , or (3) it is proved histologically wherein calcification, even bone formation is observed (DEKLEIJN [3] , STEWARD et al [10] , SHERMAN [11] , Das [2] .

Whether all the eyes where retinoblastoma presents as inflammation lead to phthisis- is not definitely known. But one thing, definitely comes out in literature that those eyes that become phthisical do become irritable- at one stage or the other. On the other hand the visible tumours that undergo calcification under observation do not lead to phthisis of the eye. These are the tumours that apparently suffocate themselves to death while others make a-hue and cry before death. The latter category produce some inflammation probably toxic in nature. SHERMAN [11] had stressed the fact that retinoblastoma can occur in a small sized eye and even in a phthisical eye. The case under discussion also supports this concept.

Two types of hypothesis have been put forward to explain the aetiology of regression. Either the tumour outgrows its blood supply leading to necrosis and production of toxic products, which further lead to necrosis and a vicious circle is set up, or, as suggested by STEWARD [10] one eye develops a sort of `resistance' following a tumour in the other eye.

Due to this 'resistance' according to him the tumour regresses.

The latter hypothesis might be applicable to types of cases where one eye is treated for retinoblastoma and the other eye shows regression later as is seen in large majority of cases reported in literature.

We agree with Reese and Sharman that any phthisical eye in a child must be removed. It also is incumbent upon the attending surgeon to keep under observation the other eye regularly for any appearance of retinoblastoma later.

CONCLUSION:

Phthisis bulbi is not uncommonly encountered in children, one of the most important causes being retinoblastoma. The case under study guides us, as to the approach to such a case. It also tells us that it is incumbent upon every ophthalmologist to observe the other eve of the patient.

It is because of the rarity of proven cases and the important lesson that such type of cases deliver, the present study is being put on record.

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