

PSEUDOHYPOPYON - AN UNUSUAL PRESENTING SIGN IN RETINOBLASTOMA

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ABSTRACT

Retinoblastoma is the most common intraocular malignancy of childhood. A case of pseudohypopyon as an unusual first sign of retinoblastoma is presented. More common clinical presentations and diagnosis of this disease are also discussed. As some cases of retinoblastoma can mimic non-malignant disease, it is important that the physician have a high index of suspicion for this tumour.

Keywords: Pseudohypopyon, retinoblastoma, childhood tumour, intraocular tumour.

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INTRODUCTION

Retinoblastoma, derived from primitive retinoblasts, is the most common intraocular malignancy of childhood. It occurs in 1 in 14,000 to 34,000 live births⁽¹⁻³⁾.

Five percent have a family history. Of the remaining 95%, 20% are thought to arise from germinal mutation with the affected person having the capacity to pass the gene to the offspring. It is now recognized that the inheritance is autosomal dominant with incomplete penetrance^(1-3,9).

In 25-30% of cases, the tumour is bilateral. All of these have the inherited form of disease.

The majority are diagnosed at under 3 years of age, with the average age quoted as 13 or 18 months, depending on the series^(1,4,5). Bilateral cases are usually recognized earlier, at an average age of 12 months, and the unilateral ones at about 24 months^(1,6,7). There is no sex predilection.

The affected child is thus usually too young to complain of any eye symptom and often does not present early. It therefore behoves the physician to be alert to this malignancy and to ensure that the child receives prompt attention as soon as retinoblastoma is suspected.

CASE REPORT

A three and a half year old Chinese boy was seen at the Department of Ophthalmology, Singapore General Hospital because his parents noticed "something white in the left eye" for four months.

On examination, he was found to have a quiet left eye with a small covergent squint. A 2 mm level of pseudohypopyon was noted in the anterior chamber of the left eye (Fig 1). The pseudohypopyon was chalky white in colour and consisted of migrated cells loosely packed together. It had a somewhat powdery appearance and the level moved with changes in the patient's head position. This, plus the conspicuous absence of any conjunctival injection or anterior chamber flare, suggested little, if any, inflammatory reaction in the anterior segment of the eye. The eye was of normal size. The cornea was clear and the pupil was round with no synechia but reacted with a rela-

tive afferent pupillary defect. Initially no leucocoria was noted but with the pupil well-dilated, a white reflex was seen through the nasal half of the left eye. The right eye was normal.

He was the only child in the family and had been a fullterm, normal baby with no significant illness before. There was no family history of any serious eye disease.

In view of the recent history, the clinical findings and his young age, the provisional diagnosis of retinoblastoma was made. He was admitted for investigations and examination under anaesthesia.

A plain skull radiograph did not show any abnormal intraocular calcification or bony erosion. CT Scan was normal.

Examination under anaesthesia with both pupils widely dilated was done by the authors, and revealed a large mass with areas of focal calcification in the left nasal retina. There was a separate mass in the temporal retina and scattered creamy white deposits over the other areas of the retina and vitreous. The vitreous was hazy with tumour cells.

The right eye was normal.

Paracentesis and aspiration of the layer of cells (pseudohypopyon) in the anterior chamber was done and sent for cytology.

Cytological analysis of the aqueous aspirate showed mostly necrotic cells and some tumour cells consistent with the diagnosis of retinoblastoma.

Enucleation of the left eye was carried out one week later and the pathologist reported undifferentiated endophytic retinoblastoma with pseudorosettes and areas of necrosis^(1,8). Tumour deposits were present over the ora serrata, iris and anterior chamber. The choroid and the cut end of the optic nerve were not involved by tumour. The patient was referred for a course of radiotherapy and later fitted with an ocular prosthesis.

He has been reviewed regularly and up to 7 years later, has had no recurrence, or any new tumour in the other eye.

DISCUSSION

Clinical presentation

The most common presentation (60%)^(5,9-11) is that of leucocoria or "white pupil" (Fig 2). This is sometimes noted as a loss of ocular red reflex in coloured photographs of the child, when taken with a flashlight. The tumour occupies the vitreous cavity and appears as a white mass behind the pupil. It may be mistaken for a cataract. However, retinal vessels coursing over the tumour distinguish it from a lesion of the lens. A co-existing cataract is unusual. In contrast, persistent hyperplastic primary vitreous (PHPV) and nematode enophthalmitis, conditions which may mimic retinoblastoma, are commonly associated with a cataract⁽¹⁾.

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Fig 1 – Pseudohypopyon in the patient's eye

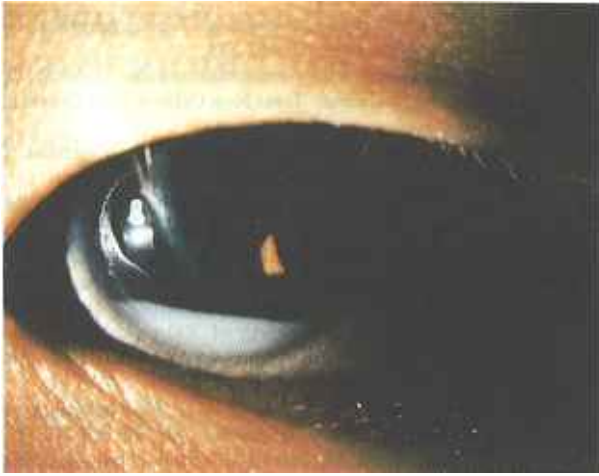


Fig 2 – Retinoblastoma presenting as leucocoria or "white pupil"

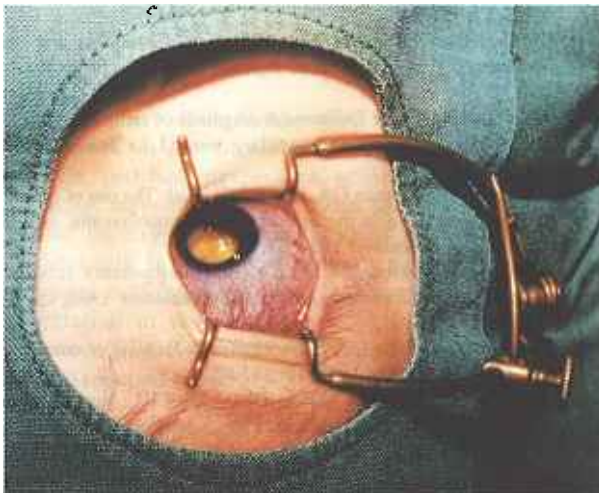
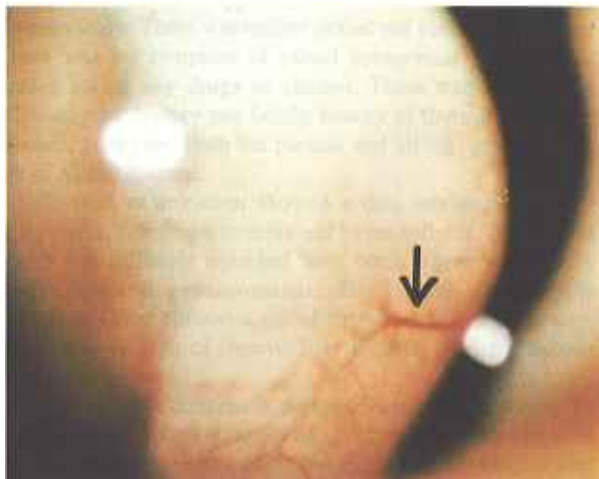


Fig 3 – Retinoblastoma exhibiting exophytic growth. Arrow shows retinal vessels coursing on the surface



The second most common presentation of the retinoblastoma is strabismus (20%), esotropia and exotropia occurring at equal frequency^(5,9-13). However, in infants esotropia may be more common⁽¹²⁾.

Other clinical presentations are a red eye with or without glaucoma (7%), poor vision (5%), change in iris colour (1%), or in very late cases, a mass extruding from the eye with proptosis^(5,9-12).

Pseudohypopyon, which is a level of tumour cells in the anterior chamber, is a very rare occurrence^(14,15). The Oncology Service at Will's Eye Hospital, Philadelphia, reviewed 60 consecutive new retinoblastoma patients between 1974 and 1978 and reported none presenting with pseudohypopyon⁽⁶⁾.

In another study of 504 cases of retinoblastoma seen from 1960 till 1983 in the Eye Clinic of Essen, West Germany, only 2 cases presented this way⁽¹²⁾.

A larger series of 1500 retinoblastoma patients seen at the New York Hospital, Cornell Medical Center Ophthalmic Oncology Center revealed only 17 patients who either had pseudohypopyon at initial presentation or developed it at a later stage⁽¹⁵⁾.

Pseudohypopyon may be mistaken for inflammatory disease of the eye. Several cases have been reported to be misdiagnosed and treated as uveitis, especially if the vitreous also has tumour seedings obscuring a view of the retinal mass. However, a quiet eye in the absence of the injected red eye of inflammatory disease should alert one of a masquerading tumour^(15,16).

In the New York Hospital study, those patients initially misdiagnosed as uveitis were the oldest patients in the study, and therefore not in the typical age group of under 3s. They had diffuse infiltrating disease with no distinct mass identified ophthalmoscopically. Such tumours are rapidly progressive with a poorer prognosis and also fail to calcify sufficiently for detection by radiography^(15,17,18).

A subtle differentiating sign that was present in our patient was the powdery appearance and consistency of the pseudohypopyon. A hypopyon of inflammatory material is usually more tenacious and the level unlikely to move freely with changes in head position.

Diagnosis

Ophthalmoscopy

Diagnosis can be made in the majority of cases by careful ophthalmoscopy through a dilated pupil^(6,11,20). The pupil is easily dilated with 0.5% or 1% Mydracyl instilled twice 5 minutes apart. Maximum dilation occurs 20 to 25 minutes later. Other tests are frequently just aids in diagnosis.

The tumour may demonstrate endophytic or exophytic growth, or a combination of both.

The endophytic tumour grows inwards towards the vitreous, breaking through the retina. It appears as a white chalky mass with no retinal vessels on the surface. It seeds the vitreous with yellow-white tumour clumps. Often, glistening white areas of calcification are seen on the tumour mass as well, as was found in this patient. An inflammatory lesion, particularly a nematode ophthalmitis, may be confused with the endophytic retinoblastoma.

The exophytic type of tumour grows beneath the retina, pushing the latter forwards in retinal detachment. Retinal vessels would then be seen coursing on its surface (Fig 3). In this instance, Coat's disease and other causes of exudative retinal detachment are differential diagnoses to consider.

Ophthalmoscopically, the 2 classical signs of retinoblastoma are seeding and calcification⁽¹⁰⁾.

Radiography

In 75% of patients, delicate flocculent calcification is apparent in plain X-rays of the orbit^(10,11).

The CT scan is important in picking up gross optic nerve or orbital extension of the tumour. It is also useful in detecting any associated pinealoblastoma (trilateral retinoblastoma) in cases of bilateral retinoblastomas^(4,10,19).

Paracentesis

Aspiration for cytological analysis in instances of pseudohypopyon is sometimes useful but occasionally may yield only necrotic cells and no tumour cells.

Apart from the above, ultrasonography, magnetic resonance imaging, enzyme analysis, fine needle biopsy and immunological methods have been used in diagnosing retinoblastoma.

CONCLUSION

This article highlights pseudohypopyon as an uncommon presentation of retinoblastoma. It is important to bear in mind that *leucocoria* and *strabismus* are the 2 most common presenting complaints. In a child under 5 years of age with these signs and symptoms, retinoblastoma should always be considered, as early referral and treatment can save sight and life.

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