

conjunctiva to the underlying sclera, where it is usually mobile.³ In this patient, the lesion behaved like it arose from a PAM by spreading to contiguous areas, developing blood vessels that fed the tumor, and increasing in thickness and size over the years.

When the melanocytes remain in the epithelium, there is no possibility for metastasis. However, once intraepithelial pagetoid tumors involve 50% to 100% of the bulbar and palpebral conjunctiva, the chance for metastasis is larger.^{1,2} Advanced melanomas ultimately invade lymphatics and spread to preauricular, submandibular, and cervical nodes before disseminating to the parotid gland, liver, subcutaneous tissues, and brain.

Management of malignant melanoma of the conjunctiva is difficult and the subject of many controversies. Complete excision of the tumor and prevention of local recurrence, metastasis, and death are the goals.² Options include a wide excision of the melanoma with cryotherapy of the margins,⁵ wide excision with topical chemotherapy such as mitomycin-C,⁴ (Mitomycin C, Kyowa, Tokyo, Japan), wide excision with radiotherapy, and orbital exenteration. For small circumscribed melanomas, beta-irradiation with strontium-90 surface applicators, proton-beam therapy, gamma emissions from cobalt-60 or cobalt plaques have been found effective.⁴

Due to the extensive involvement of the conjunctivae, including the caruncle and upper lid, an orbital exenteration was done in this patient. Histopath results showed largely epithelioid configuration of spindle cells. The eyeball, optic nerve, lacrimal gland, and all surgical margins were negative for tumor.

Malignant melanoma is the most serious of the conjunctival malignancies. Nearly 50% of patients have recurrence after resection, while 26% develop metastasis after 10 years.¹ Prognosis is often poor for patients with malignant melanoma in the palpebral conjunctiva, fornices, plica, caruncle, or eyelid margin. Among these patients, mortality is 2.2 times higher.² Prognosis improves for patients with lesions found in the bulbar conjunctiva.¹

Proximity to the lymphatic system may explain why some locations are less favorable than others.² Survival rates are worse for the following: lesions arising from PAM, those with an initial thickness of more than 4 mm, lesions arising from an unfavorable site, multifocal disease in favorable sites, mixed spindle and pure epithelioid cells with lymphatic invasion, high mitotic index, and lesions that grow vertically.^{1,2}

In this patient, prognosis was considered poor because the tumor involved the palpebral conjunctiva, fornices, and caruncle, and the lesions arose from a PAM. Whether orbital exenteration can prevent metastasis is uncertain. Some studies have reported that the procedure did not lead to increased survival rate.⁴

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Retinal Dysplasia*

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ABSTRACT

Objective

To report a case of retinal dysplasia.

Method

This is a case report of retinal dysplasia, the first documented case seen at the University of the Philippines-Philippine General Hospital (UP-PGH).

Results

A three-month-old female presented with bilateral leukocoria and an intraocular mass in the right eye. The right eyeball was enucleated and histopathology results revealed retinal dysplasia.

Conclusion

Ocular ultrasonographic and cranial computerized tomography are helpful in the diagnosis of suspected retinal dysplasia.

Retinal dysplasia consists of an abnormal proliferation of developing retina, producing tubular structures with a rosette-like appearance in cross section.¹ It appears to be associated with a single pathogenetic basis: the separation of the retina during a critical stage of its differentiation from its underlying pigment epithelium.²

We are presenting the first reported case of retinal dysplasia seen at the UP-PGH.

A three-month-old female, the second of twins, born preterm at 7 months (1,400 grams) via Caesarean section,

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presented with bilateral leukocoria at 2 months of age. Supplemental oxygen was given for 7 days. Her development was at par with other infants her age. The patient's twin sister had retinopathy of prematurity in both eyes (OU).

Ophthalmologic examination revealed negative dazzle in OU. The right cornea measured 8 mm x 8 mm while the left cornea measured 10 mm x 10 mm. Pupils were 3 mm, slowly reactive to light with no afferent pupillary defect. Full range of ocular movements were present in OU. Intraocular pressure (IOP) was less than 4 mm Hg in the right eye (OD) and 7 in the left eye (OS). The anterior chambers in OU were shallow with posterior synechiae. A whitish mass was noted in OU with retinal vessels and hemorrhages. Gonioscopy revealed primarily open angles in OU with peripheral anterior synechiae interspersed inferiorly in OS.

The rest of the organ systems were normal. Cranial computerized tomography (CT) showed no intracranial lesions.

Ocular ultrasonography showed a right eyeball measuring 1.6 cm in anteroposterior diameter. The anterior half of the vitreous showed granular density with high narrow spikes and a midline irregular membranous density indicating detached retina. Behind the membrane was an irregularly shaped mass with spikes 50 to 70% of the anteriorly located densities. The choroid was markedly thickened (Figure 1). The left eyeball was 1.8 cm in length with findings of retinal detachment and thickened choroid.

CT showed a small right globe with calcifications in the posterior globe and enlarged optic nerve, and a larger left globe with irregular thickening and increased attenuation of the inferoposterior aspect accompanied by calcifications.

Differential diagnoses for leukocoria in infancy include retinopathy of prematurity (ROP), persistent hyperplastic primary vitreous (PHPV), Coats' disease, and retinoblastoma.

ROP is a proliferative retinopathy seen in premature infants weighing less than 1,500 g at birth. The younger the gestational age and the lower the birth weight, the more severe is the case. Administration of supplemental oxygen to the newborn has been implicated as a cause. It is typically a bilateral disease that affects the vitreous and peripheral retina. Even though the patient was born preterm with low birth weight, the presence of an intraocular mass and calcification ruled out this condition.

PHPV is a congenital, nonhereditary malformation of the eye that is usually unilateral and not associated with systemic defects. Severe cases may present with microphthalmic eyes with shallowing of the anterior chamber and retinal detachment. The bilateral involvement in this patient and the presence of calcifications make this condition unlikely. In less severe PHPV, a

membranous band extending from the posterior lens capsule to the optic disc may be present.

Coats' disease is an idiopathic condition characterized by telangiectatic and aneurysmal retinal vessels with intraretinal and subretinal exudates. It is unilateral in 80% or more of cases,³ affecting more males (3:1 ratio). Gradual progression with increasing exudation occurs over time. Massive exudative retinal detachment can occur with cholesterol crystals producing characteristic echograms that were not seen in this patient.

Retinoblastoma is a neuroblastic tumor that is the most common primary intraocular malignancy of childhood. The most common initial sign is leukocoria. Exophytic tumors are usually yellow-white and occur in the subretinal space with subretinal fluid accumulation and exudative retinal detachment. Calcification is a common finding in areas of necrosis of tumor cells producing echograms of extremely high reflectivity on A-scan and very bright signals on B-scan.⁴ Retinoblastoma was considered in this patient because of the early presentation of bilateral leukocoria and findings on ultrasonography confirmed by CT.

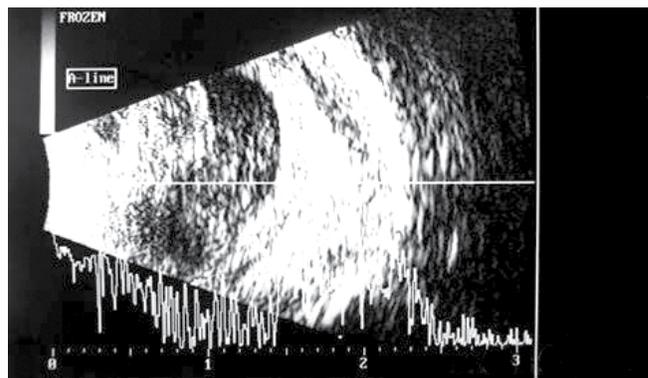


Figure 1. Ultrasound of the right eye.

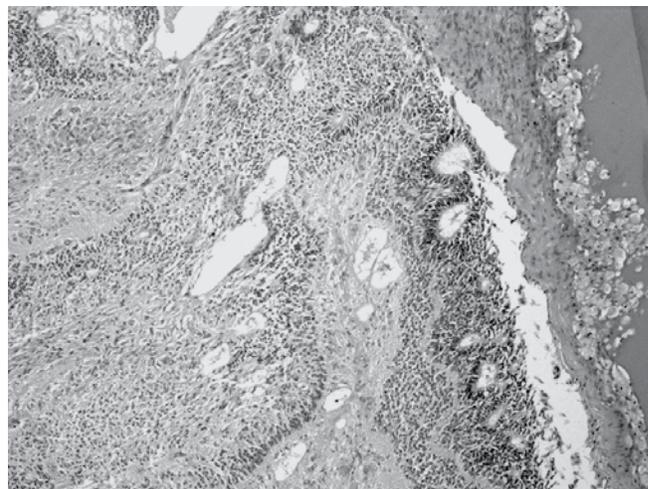


Figure 1. Rosettes of retinal dysplasia.

The right eye was enucleated. Dissection of the globe showed a white retrolental mass occupying the anterior one half of the vitreous cavity. Microscopically, a completely dysplastic detached retina was totally separated from the choroid by eosinophilic, serous subretinal fluid and hemorrhage. There were numerous macrophages ingesting the red blood cells and cholesterol clefts in the subretinal fluid denoting an old vitreous hemorrhage. Focal retinal dysplasia includes extensive glial formation, cellular proliferation and rosette-like formation (Figure 2). The optic nerve was unremarkable.

The pathogenesis of retinal dysplasia include the following:

- resulting from hyperplastic extension of the retina into “abnormal” sites away from its pigment epithelium,
- secondary to detachment of the retina from the pigment epithelium,
- occurring in an otherwise normal location over areas devoid of pigment epithelium, and
- *in situ* dysplastic process with no evidence that the dysplastic retina has ever been separated from its underlying epithelium.⁵

The underlying feature of each of these processes is the absence of a presumed normal histogenetic control by the pigment epithelium on the developing retina.⁵ Dysplastic changes appear to be intermediate between a receptor that has developed normally and one that has

followed uncontrolled, neoplastic growth. Muller fibers contribute to dysplastic rosette formation but not retinoblastoma rosette formation.

Retinal dysplasia may also be associated with congenital anomalies (13%),⁶ or with chromosomal abnormalities⁷ like Trisomy 13, or systemic findings such as Norrie’s disease (x-linked recessive).⁷

Retinal dysplasia should be considered as a differential diagnosis in patients presenting with leukocoria. The ultrasonographic and tomographic findings are helpful in the diagnosis of suspected retinal dysplasia. A thorough physical examination is needed to rule out systemic abnormalities associated with the disease. Early screening will be of help in genetic counseling of the parents.

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