Case Report

Pigmented Paravenous Chorioretinal Atrophy: A Fortuitous Finding in a Patient with Fronto-Ethmoidal Mucocoele

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Abstract

A 56-year-old Nigerian male patient presented to the eye clinic with a six-month history of progressive deterioration of vision in the right eye which was preceded by a one-month history of swelling above the right eye. He had no visual complaints in the left eye and no concurrent systemic illnesses. The best corrected visual acuity was hand movement with accurate light projection in the right eye and 6/6 in the left eye. On ocular examination, a right proptosis with inferotemporal displacement was present. A right relative afferent pupillary defect was noted while the left anterior segment examination was normal. Binocular indirect ophthalmoscopy of the right eye revealed a cup-to-disc ratio of 0.6, temporal disc pallor, peripapillary atrophy, and clumps of pigmentation along the venous arcades with generalized chorioretinal atrophy involving the area of the fovea. The cup-to-disc ratio was 0.4 with mild temporal disc pallor and iridescent spots at the macula with similar features of bony spicule-like clumps in the right eye present along the superotemporal arcade in the left eye. Optical coherence tomography (OCT) scan revealed disorganization of the inner retinal layers with intraretinal reflective foci with back-shadowing and both retinal pigment epithelium and choroidal atrophy. The OCT of the left eye showed a few intraretinal reflective foci and retinal pigment epithelium and choroidal atrophy. From the clinical and OCT findings, a diagnosis of pigmented paravenous chorioretinal atrophy and right fronto-ethmoidal mucocele is made.

Keywords: Bone spicule-like clumps, chorioretinal atrophy, fronto-ethmoidal mucocele, iridescent spots, paravenous pigmentation

Introduction

Pigmented paravenous chorioretinal atrophy (PPCRA) is a rare, unusual ocular condition with distinct features comprising of clumps of pigmentation mainly peripapillary with radiating, linear areas of chorioretinal atrophy. These clumps of pigment are typically present along the course of the retinal veins extending from the disc to the equator.[1] PPCRA was first described in a 47-year-old man with alopecia areata in 1937 and subsequently named retinochoroiditis radiata due to the peculiar distribution of the lesion.[2] Individuals with PPCRA usually have minor or no visual complaints, as it is often a contingent finding.[3] The etiology of PPCRA is largely unknown though it has been postulated to occur in association with inflammatory conditions such as syphilis, rubeola, measles, and uveitis.[4] It is thought to be an acquired retinal disease and usually bilateral and symmetrical though variations in symmetry have been described.[3,5] A mutation in the crumbs cell polarity complex component 1 gene which is also linked with other inherited retinal disorders has been associated with PPCRA.[6] This may probably be the first reported case of PPCRA in a Nigerian to the best of our knowledge. Consent was obtained from the patient.

Case Report

A 56-year-old man presented to the eye clinic with a 6-month history of deterioration of vision in the right eye. He noticed a swelling above the right eye with downward deviation of the eye about four weeks before the drop in vision. There was a positive history of headaches and eye

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aches but no antecedent history of trauma nor entry of any foreign body. He had no history of nyctalopia. He had no visual complaints in the left eye and no known concurrent systemic illnesses.

Visual loss in the right eye was discovered accidentally when he presented at the referral center to complain about the swelling around the right eye but has been progressively worsening. The patient believed that his vision was normal in both eyes before the onset of symptoms. There was no history of cough, rhinorrhea, ear discharge, inability to close the eyes nor drooping of the eyelids. There was no history of weight loss nor swelling in any other part of the body. Systemic review was not contributory. He had no family history of blinding eye disease nor similar ocular illnesses.

On general examination, our patient was a middle aged man, with asthenic build, afebrile, not pale, anicteric, good hydration status, and no palpable peripheral lymphadenopathy. He was conscious and alert and well oriented in time, place, and person. There was no sign of meningeal irritation and the muscle power and tone was grade 5 in all limbs. Cranial nerve examination was normal except for a deficit in the optic nerve on the right. On examination of the cardiovascular system, the pulse rate was 80/min, regular, good volume with normal first and second heart sounds and no murmurs. The chest was clear with a respiratory rate of 20 cycles per minute and vesicular breath sounds. Abdominal examination was normal and there was no palpable organomegaly. Examination of the ear, nose, and throat was essentially normal except for bilateral engorged turbinates which was more pronounced in the right nostril.

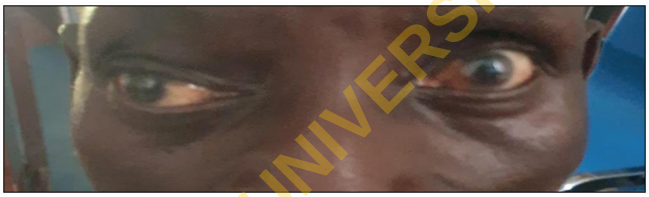

Figure 1: The right eye with non-axial proptosis deviated inferotemporally.

Ocular examination revealed a best corrected visual acuity of hand movement with accurate light projection in the right eye and 6/6 in the left eye. There was a cystic swelling in the supraorbital region of the right eye with inferotemporal dystopia. The swelling was firm, mobile, non-tender, unattached to overlying structures, transilluminating with difficult finger insinuation. The extraocular muscle movements of the superior, lateral, and medial recti and inferior oblique muscles were restricted. Proptosis of the right eye measuring 3mm with Hertels' exophthalmometer was noted [Figure 1]. The anterior segment examination was essentially normal in both eyes except for a relative afferent pupillary defect present in the right eye and bilateral atrophic nasal pterygia.

The intraocular pressures measured by Goldmann applanation tonometry were 26 and 10 mmHg in the right and left eye respectively. Color vision tested with Ishihara pseudoachromatic chart was normal in the left eye but was not elicited in the right eye due to the diminished vision.

Binocular indirect ophthalmoscopy of the right eye revealed a slightly tilted disc with cup disc ratio 0.6, temporal disc pallor and peripapillary atrophy, clumps of bony spicule-like pigmentation on the posterior pole distributed along all the venous arcades with iridescent spots and marked chorioretinal atrophy at the macula. Vitreous strands and a posterior vitreous detachment were also present [Figure 2A]. The left eye had a pink disc with cup disc ratio of 0.4, peripapillary atrophy, pigment clumps distributed mainly along the superotemporal arcade, a single vitreous strand radiating from disc with iridescent spots and few pigmentary changes at the macula [Figure 2B].

Optical coherence tomography (OCT) scan revealed altered foveal contour, distortion of the retinal layers with loss of the ellipsoid zone, marked atrophy of the retinal pigment epithelium and choroid with intraretinal reflective foci, and back-shadowing in the right eye. The OCT scan of the left eye showed a normal foveal contour

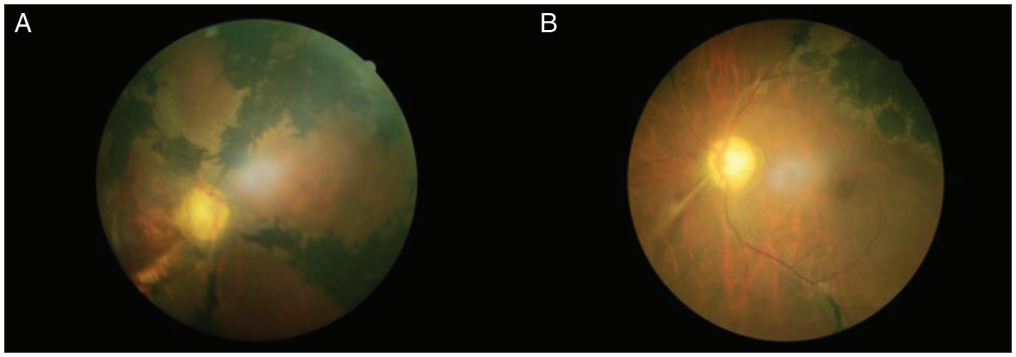

Figure 2: Fundus photographs of both eyes. (A) The right eye shows pigment clumps and areas of chorioretinal atrophy along all four venous arcades while in (B), the left eye there are clumps of pigmentation and chorioretinal atrophy more pronounced along the superotemporal arcade with few pigment clumps along the inferotemporal arcade

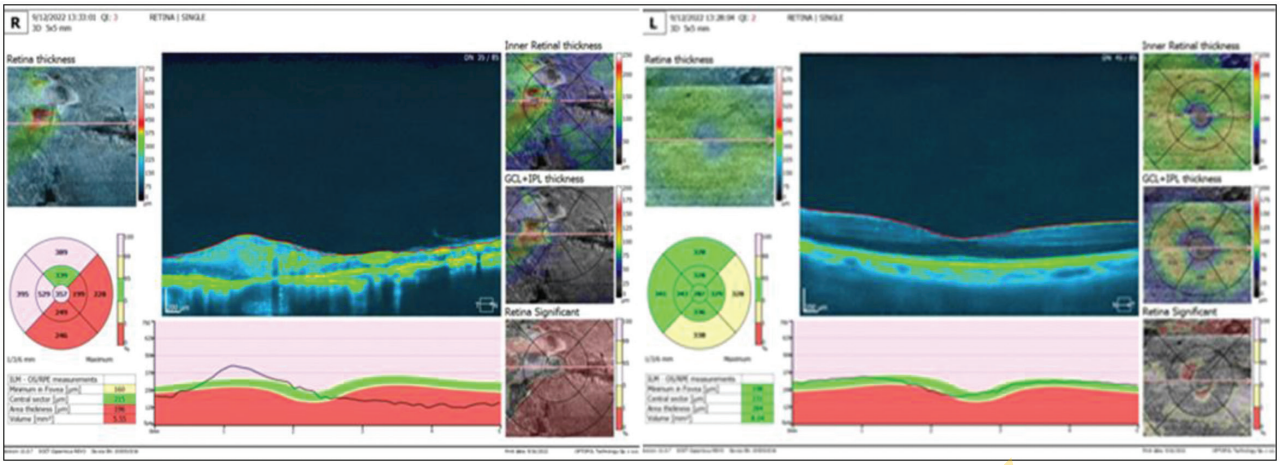

Figure 3: OCT scan of both eyes. In the right eye (R) there is disorganization of the inner retinal layers, intraretinal reflective foci with back-shadowing and retinal pigment epithelium and choroidal atrophy with lifting of the internal limiting membrane nasally. In the left eye (L) there are few intraretinal reflective foci with retinal pigment epithelium and choroidal atrophy

with retinal thinning and atrophy of the choroid with some visible choroidal vasculature with few intraretinal reflective foci [Figure 3]. Central visual field evaluation was not done as our patient was unable to fixate for the investigation.

Cranial computed tomography scan from the referral center revealed osteolytic, expansile lesion of right frontal bone with outer and inner table erosion with extension of the mass into the right orbit. Both optic nerves appeared intact. The features of mass effect of the lesion with inferolateral displacement of the globe and lack of intracranial extension were in keeping with a right fronto-ethmoidal mucocele.

A diagnosis of bilateral pigmented paravenous chorioretinal atrophy and a right fronto-ethmoidal mucocele with orbital extension and right glaucoma was made based on the history and clinical findings. The patient was counseled about the retinal diagnosis and visual prognosis and was referred to the orbit and oculoplastic surgeon and otolaryngologists for further review and management.

Discussion

PPCRA was an incidental finding in this index patient. The reason for the presentation at the eye clinic was the swelling above the right eye. The clinical features of this swelling were in keeping with features of a fronto-ethmoidal mucocele. Peripapillary and radial chorioretinal atrophy with pigment clumps along the course of the venous network which are among the classical clinical features of PPCRA[1,7,8] was present in both eyes of our index patient on indirect ophthalmoscopy. PPCRA may be asymptomatic and discovery was made on routine ocular examination.

Reports of familial cases of PPCRA have been documented in literature though our patient had no family history of visual complaints hence the high likelihood of it being sporadic.[8-11] A detailed ophthalmic evaluation of family members would rule out a hereditary component.[1,2,7,10] A unilateral case of PPCRA has also been reported though our patient had asymmetric presentation.[3,5,12,13]

The retinal findings were asymmetrical with affectation being more severe in the right eye, hence the poorer visual acuity in the right eye. Though rare, macula involvement is the most likely etiology of the poor vision in the right eye.[3] Furthermore, the deficient vision in the right eye may be attributable to marked chorioretinal atrophy at the macula confirmed on OCT and glaucomatous optic atrophy and elevated intraocular pressure in that eye. As the initial name for this entity retinochoroiditis radiata implies, the perivascular lesions were typical and dense in our patient especially in the right eye.[2] Asymmetric retinal lesions in PPCRA as seen in this index patient have been documented in other reports.[3,5,10]

The diagnosis in this patient was based on the characteristic perivascular hyperpigmentation and associated chorioretinal atrophy. The OCT scan findings of thinning of the retinal layers, loss of ellipsoid zone and choroidal atrophy with intraretinal foci associated with back-shadowing are documented features of PPCRA.[11,13,14] OCT signs such as cystic spaces and serous detachment may occur in PPCRA but was absent in this patient.[10,14]

A useful diagnostic tool in PPCRA is fundus autofluorescence (FAF). This reveals hypofluorescent areas correspond to areas of RPE loss and retinal thinning. Likewise, electroretinography (ERG) is also helpful in making the diagnosis with reduction in amplitude of the scotopic and photopic stimulation being characteristic findings. Fundus fluorescein angiography (FFA) may also be beneficial in making a diagnosis as window defects in the early phase

correspond with areas of choriocapillary atrophy and prominent choroidal vessels are seen. Central visual field examination will show constricted fields.[11,13-15] Our patient was unable to fixate for central visual field evaluation. ERG, FAF and FFA are not available at our hospital; nevertheless the characteristic retinal signs and the OCT scan findings were sufficient to arrive at the diagnosis of PPCRA.

It has been postulated that PPCRA may be of inflammatory origin and cases co-existing with Behchet disease and Vogt-Koyanagi-Harada syndrome has been described.[4,11,13] Although our patient presented with a right fronto-ethmoidal mucocele probably secondary to a chronic rhinosinusitis, the PPCRA is more likely to be a pre-existing ocular disorder in this case.

Differential diagnoses considered include retinitis pigmentosa which was ruled out as there was no history of nyctalopia and the presence of radial paravenous pigment clumps in the patient. Serpigous tuberculous chorioretinitis was ruled out as he had no history of cough nor clinical ocular and systemic signs in keeping with tuberculosis. Pseudo retinitis pigmentosa disorders such as toxoplasmosis and other uveitic conditions were also ruled out.

Conclusion

Pigmented paravenous chorioretinal atrophy may be an incidental finding in ophthalmic patients. These patients may present to the ophthalmologist with other signs of other ocular disorders or on routine ocular evaluation. A detailed history and ocular examination including a binocular indirect ophthalmoscopy are essential for prompt diagnosis.

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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Conflict of interest

There are no conflicts of interest.

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