Choroidal melanoma in phacomatosis pigmentovascularis cesioflammea

A 56-year-old white male patient was referred for management of a choroidal mass in his left eye. A cutaneous nevus flammeus involving the right side of his face extending to the right side of his neck and upper chest was noticed (Fig. 1A). On questioning, he mentioned that this nevus flammeus was previously treated with several sessions of dye laser. In addition, the eyelid skin showed a blue-grey hue that involved the upper and lower eyelids bilaterally (Fig. 1B, C). Slit-lamp examination revealed bilateral multiple flat, purple-coloured episcleral patches. Both irides and anterior chamber angles were heavily pigmented (Fig. 2A, B); however, the intraocular pressure was 15 mm Hg in both eyes. There were multiple anomalous vessels, with saccular dilatation, microaneurysms, and abnormal branching involving the right episclera and bulbar conjunctiva that were not discernible in the left (Fig. 2C). Funduscopy of the right eye was within normal limits, but in the left eye, there was a partially melanotic medium-sized choroidal mass located 2 mm superior to the foveola, associated with lipofuscin deposition (Fig. 2D). The mass appeared hypoechogenic on ultrasonography with associated subretinal fluid on optical coherence tomography. The funduscopy and imaging findings of the left eye were conclusive of a choroidal melanoma. Thus, this patient presented with bilateral oculodermal melanocytosis in association with a right oculocutaneous nevus flammeus and left choroidal melanoma. Ensuing systemic and neurologic computed tomography imaging revealed no relevant abnormalities.

The term *phacomatosis pigmentovascularis* (PPV) describes the coexistence of extensive cutaneous vascular malformation with melanocytic proliferation. This association encompasses several clinical presentations, which were first described and classified by Ota et al.[1] in 1947. PPV was initially classified into 4 types that were further subclassified according to absence (group A) or presence (group B) of systemic associations. To simplify, in 2005, Happle[2] reclassified PPV into 3 types: cesioflammea (blue nevus and nevus flammeus), *caesius* is Latin for bluish grey; spilorosea (nevus spilus and a pale pink telangiectatic nevus); and cesiomarmorata (blue spots and cutis marmorata telangiectatica congenita). The genetic model to explain PPV is "twin spotting," which assumes that PPV results from mosaic proliferation of 2 different clones of cells within a normal cutaneous area. It is thought to result from a postzygotic somatic mutational event, as evidenced in a reported case of monozygotic twins with discordant presentation.[3] PPV cesioflammea type represents 77% of PPV cases and could be associated with focal alopecia, deafness, and other neurologic and skeletal malformations; none of which manifested in our patient.[4] Because there was no evidence of a leptomeningeal angioma on neuroimaging or a choroidal hemangioma on funduscopy, the nevus flammeus in our patient was not thought to be a manifestation of Sturge–Weber syndrome. Nevertheless, similar to Sturge–Weber syndrome, anomalous conjunctival and episcleral vessels with nevus flammeus carries risk for glaucoma development from raised episcleral venous pressure or associated angle anomalies. This risk may be amplified because ocular melanosis carries a risk for pigmentary glaucoma. Teekhasaenee and Ritch[5] reported in their series of 9 PPV patients the presence of glaucoma in all the eyes that were simultaneously affected with melanocytosis and episcleral vascular malformation. Our patient experienced development of choroidal melanoma presumably as a consequence of the ocular melanocytosis component that carries 1:400 risk for development of uveal

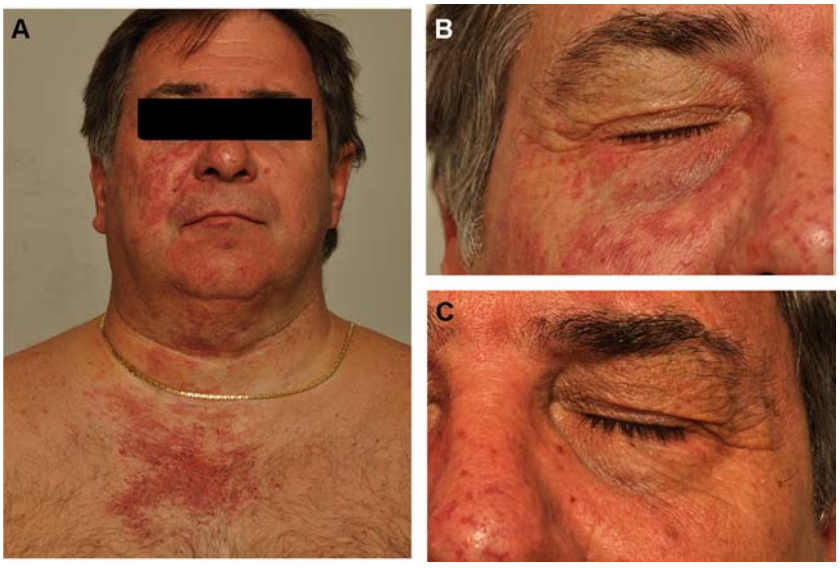

Fig. 1—(A) Nevus flammeus affecting skin of the right side of face, neck, and upper chest. (B, C) Skin of the right and left eyelids shows blue-grey hue.

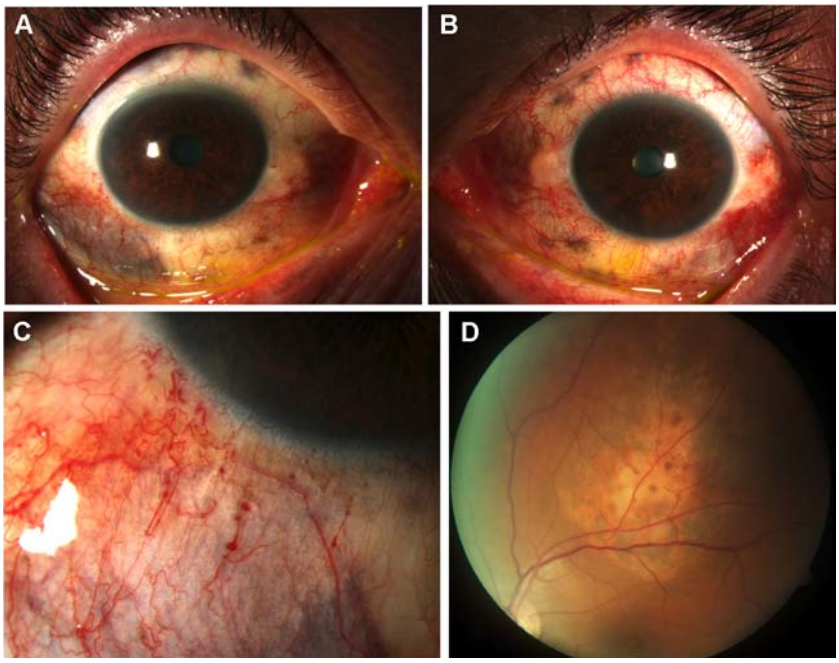

Fig. 2—(A, B) Slit-lamp photos show multiple flat, purple-coloured episcleral patches and heavily pigmented irides. (C) Right episclera and bulbar conjunctiva showing multiple anomalous vessels with saccular dilatation, microaneurysms, and abnormal branching. (D) Fundus photo of the left eye shows a choroidal melanoma.

melanoma. However, Tran and Zografos[6] could not exclude the possibility that the pathogenesis of PPV may play a role in ocular melanoma. Shields et al.,[7] in a series of 7 PPV patients, suggested dilated funduscopy for all cases of nevus flammeus, because choroidal hyperpigmentation was the only detectable manifestation of ocular melanocytosis in 50% of their cases. Because of the scarcity of ophthalmic reporting on this rare phacomatosis, PPV may be confused with more common syndromes such as oculodermal melanocytosis or Sturge–Weber syndrome, and it is to be recognized for its potential ocular and systemic implications.

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Exacerbation of central serous chorioretinopathy following PASCAL photocoagulation

Central serous chorioretinopathy (CSC) is a common retinopathy characterized by serous detachment of the neurosensory retina with or without pigment epithelial detachment (PED). The published risk factors of CSC include type A personality, pregnancy, systemic steroid use, collagen vascular disease, obstructive sleep apnea, alcohol use, hypertension, psychopharmacologic medication, *Helicobacter pylori*, and tadalafil.[1,2] We report a case of atypical CSC that developed from a preexisting PED immediately after panretinal photocoagulation (PRP).