A teenage girl with bilateral hyperpigmentation of cheeks and sclera and a vascular stain on her cheek: What is your diagnosis?

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CLINICAL PRESENTATION

A 16 year old girl was referred to dermatology clinic with bilateral blue gray hyperpigmented patches on her cheeks since birth. Upon close physical examination, scleral blue-gray hyperpigmentation was observed in both her eyes (Figure 1). She also had a blanchable red patch with poorly defined borders on her right cheek (Figure 2). Ophthalmology consultation further revealed an increased intraocular pressure with optic nerve damage. She had neither systemic problems nor a history of a similar condition in her family.

What is your diagnosis?

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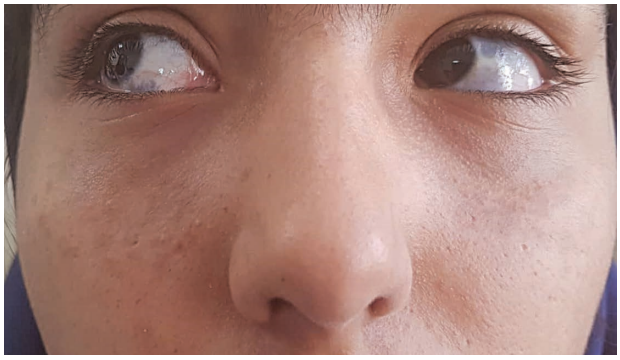

Figure 1. Bilateral blue-gray periocular and scleral patches with poorly defined borders

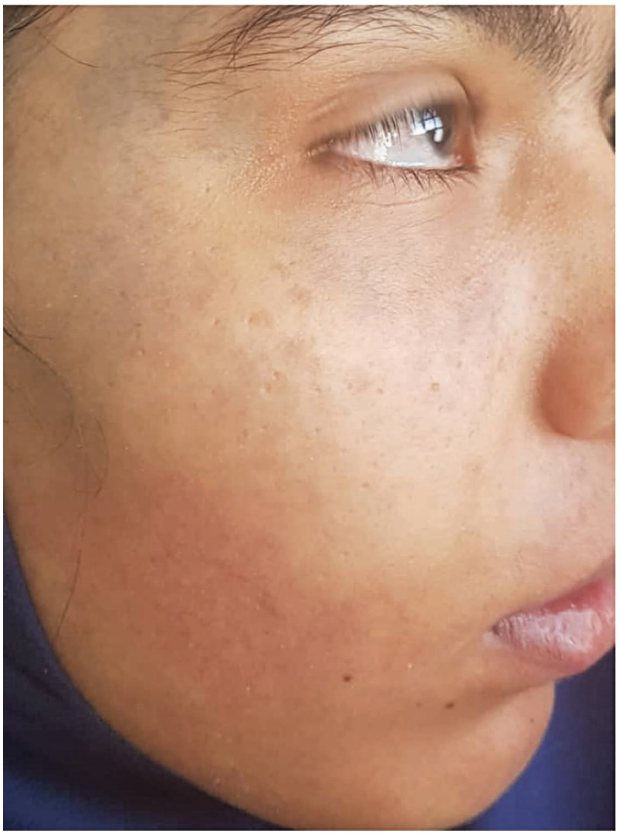

Figure 2. A blanchable red patch with poorly defined borders on her right cheek

Diagnosis

Bilateral Ota nevus and a capillary stain: Phacomatosis pigmentovascularis (PPV) type 2a

DISCUSSION

In phacomatosis pigmentovascularis (PPV), vascular and pigmentary skin lesions exist alongside one another since birth [1]. PPV is more often than not associated with different types of vascular or melanocytic lesions such as port wine stain, melanocytic or epideraml nevi, nevus of Ota and Mongolian spots. Near to 50% of affected people have systemic involvement such as neurologic or ophthalmologic abnormalities [2,3]. According to different types of skin manifestation, this skin condition entails several subtypes [1,2].

Ophthalmologic problems such as ocular melanosis often occur along with nevus of Ota and may affect one or both eyes [4]. People with nevus of Ota should undergo serial ophthalmologic examinations because they are at higher risks of golucoma or melanoma development [5]. There are reports of PPV with Sturge-Weber or Klippel-Trenaunay syndrome [6].

If PPV has no association with systemic complications, it does not require any treatment. However, due to body image and self-esteem problems caused by skin lesions, laser treatments can be helpful [4,7].

Yang et al in 2015 presented three cases of PPV with port wine stains (Sturge-Weber Syndrome), nevus of Ota and congenital golucoam [8].

In 2017, Namiki et al, reported a case with Klippel-Trenáunay syndrome, extensive dermal melanocytosis as nevus of Ota, nevus of Ito and ectopic Mongolian spots with a final diagnosis of Phakomatosis pigmentovascularis type IIb [9].

To our knowledge, this is the first report on PPV including bilateral nevus of Ota, capillary satain and bilateral golucoma.

Conflict of Interest: None declared.

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