Sir,

Bilateral Naevus of Ota in association with Klippel–Trenaunay Syndrome

Naevus of Ota, originally described by Ota,[1] is a hamartoma of dermal melanocytes. Naevi of Ota and Ito occur most frequently in Asian populations. Other ethnic groups with increased prevalence include Africans, African Americans, and East Indians. Women have a higher incidence than men. It is uncommon in Caucasians. Well-known associations include the Sturge–Weber syndrome, glaucoma, and rarely ocular melanoma. We describe a case of bilateral Naevus of Ota in association with Klippel–Trenaunay Syndrome.

Case report

A 25-year-old Caucasian women who was being investigated by the rheumatologists for multiple joint swelling and pain was noticed to have an unusual bilateral ocular discolouration. She was referred for ophthalmic review. On examination she had bilateral pigmentation affecting the anterior sclera bilaterally (Figure 1a). Both lateral lid margins were pigmented. All these had been present since birth. Her fundoscopy and intraocular pressure were normal on examination. A diagnosis of bilateral naevus of Ota was made.

Following review by the rheumatologists and vascular surgeons, she was noted to have multiple large areas of what the patient described as a birthmark over her scalp, neck, trunk, and leg. On her left leg she had a port-wine stain, extending over the whole lateral and posterior length of the leg associated with local gigantism and varicose veins (Figure 1b). A diagnosis of Klippel–Trenaunay Syndrome was made, for which conservative treatment was instituted.

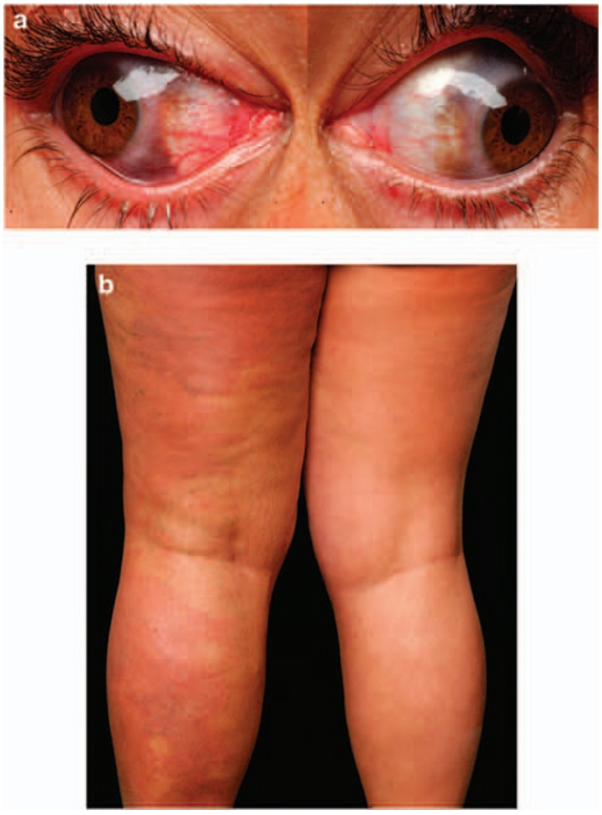

Figure 1 (a) Bilateral naevus of Ota with pigmentation also affecting both lid margins. (b) Left leg port-wine stain associated with local gigantism and varicose veins.

Discussion

The clinical findings in this patient are characteristic of Phacomatosis Cesioflammea with Klippel–Trenaunay Syndrome. Phacomatosis Cesioflammea,[2] was earlier called phacomatosis pigmentovascularis (PPV) Type II. PPV is a rare syndrome defined as an association of a widespread vascular naevus (port-wine stain) with an extensive pigmentary naevus,[3] such as a naevus of ota or Mongolian spots.

Klippel–Trenaunay Syndrome manifests as a triad of capillary malformation, congenital varicose veins, and hypertrophy of underlying tissues, particularly skeletal overgrowth. Unilateral lower limbs are frequently involved.

This case report, in addition to only one other, that of Talwar *et al*,[4] highlights that an association may exist between a bilateral Naevus of Ota, when phacomatosis cesioflammea exists in tandem with Klippel–Trenaunay Syndrome.

References

1 Ota M. Nevus fusco-caeruleus ophthalmo-maxillaris. *Jap J Dermatol* 1939; **46**: 369.
2 Happle R. Phacomatosis pigmentovascularis revisited and reclassified. *Arch Dermatol* 2005; **141**: 385–388.
3 Hasegawa Y, Yasuhara M. Phakomatosis pigmentovascularis type IVa. *Arch Dermatol* 1985; **121**: 651–655.
4 Talwar S, Jha PK, Suresh MS, Panvelkar VVN. Bilateral nevus of ota with klippel-trenaunay-weber syndrome. *Medical Journal of Armed forces India* 1992; **48**(3): 231–233.

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