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PHACOMATOSIS PIGMENTOVASCULARIS- A RARE CASE REPORT

Dermatology

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ABSTRACT

Phacomatosis pigmentovascularis is a rare disorder which presents with a vascular and pigmentary anomaly. Here we report a case of phacomatosis pigmentovascularis who presented with a port wine stain, nevus of Ota and nevus of Ito.

KEYWORDS

Phacomatosis pigmentovascularis, port wine stain, nevus of Ota, nevus of Ito.

INTRODUCTION:

Phacomatosis pigmentovascularis is characterized by the concomitant occurrence of a vascular anomaly, i.e. a capillary malformation (port wine stain) and a pigmentary anomaly in the form of dermal melanocytosis, epidermal nevus, nevus spilus, nevus anemicus and café au lait macules[1]. We report a case of phacomatosis pigmentovascularis with a port wine stain and dermal melanocytosis.

CASE REPORT:

A 28-year-old male presented to the skin OPD with complaints of a red lesion on the right side of the face since birth which was asymptomatic. On dermatological examination, there was an erythematous mildly raised lesion on the right side of the face which blanched on pressure suggestive of a capillary malformation (port wine stain).There was a right sided facial hypertrophy. In addition he also had bluish discolouration of bilateral sclera and bluish discolouration of the skin on left cheek which was present from birth implying a nevus of Ota. Over the right shoulder, there was a brownish pigmentation present from birth extending down the arm and forearm indicating a nevus of Ito. He had a prominent right sided upper limb and pectoral muscle atrophy with pectoral weakness. He gave no history suggestive of any central nervous system and other ocular involvement.

Based on the above said findings, we came to a conclusion of phacomatosis pigmentovascularis.

DISCUSSION:

Phacomatosis pigmentovascularis first described by Ota was classified by both Happle[2] and Hasegawa[3] into five types depending on the type of pigmentary and vascular involvement. Each division is subdivided into "a" and "b" depending on the presence or absence of systemic involvement respectively.

Hasegawa's classification[3]	Happle's Classification[2]	Vascular anomaly	Pigmentary anomaly
I	Non-existent	Capillary malformation	Epidermal nevus
II	Cesioflammea	Capillary malformation	Dermal melanocytosis (Mongolian spots)
III	Spilorosea	Capillary malformation	Nevus spilus
IV	Non-classifiable	Capillary malformation	Dermal melanocytosis & Nevus spilus
V	Cesiomarmorata	Cutis marmorata telangiectatica congenita	Dermal melanocytosis

These conditions may occur with or without a nevus anemicus. Capillary malformation also known by the names nevus flammeus and port wine stain is one of the commonest vascular malformations usually present at birth and growing in size as the child ages. It is usually associated with hypertrophy of the underlying structure as is in our case where there is right sided facial hypertrophy.

The commonest dermal melanocytosis to occur in phacomatosis pigmentovascularis is Mongolian spots, but there have been few reports in literature regarding the concomitant occurrence of nevus of Ota and nevus of Ito with a capillary malformation[4]. Nevus of Ota and Ito are considered aberrant Mongolian spots[4].

Nevus of Ota also known as Nevus fuscoceruleus ophthalmomaxillaris is a dermal melanocytosis which presents from birth with bluish discolouration of unilateral cheek and sclera. It may rarely be bilateral. Nevus of Ito, another form of dermal melanocytosis goes by the name, nevus fuscoceruleus acromiodeltoideus. It presents as brown to bluish gray pigmentation over the shoulders.

When suspecting a case of phacomatosis pigmentovascularis, ocular and neurological abnormalities will have to be ruled out. Ocular complications include ocular melanosis, glaucoma, iris hamartomas, mammillations and nodules[5]. Neurological involvement is in the form of delayed development, seizures, hydrocephalus, intracranial calcifications and cerebral atrophy[6].

Phacomatosis pigmentovascularis has also been reported in association with Sturge-Weber syndrome and Klippel Trenauney syndrome[7].

Prognosis of the disease depends on whether systemic involvement is present or not. Treatment of cutaneous lesions is for aesthetic purposes and includes Q switched alexandrite laser for the pigmentary lesions and pulsed dye laser for the vascular lesions[8].

CONCLUSION

Our case can be classified as phacomatosis pigmentovascularis type IIa. Though Mongolian spots are the common presentation of dermal melanocytosis, our case reported with nevus of Ota and Ito and this is a rarity.

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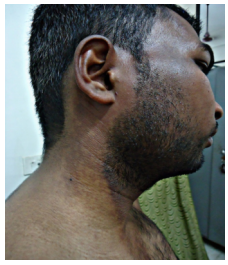

Figure 1: Port wine stain on the right side of the neck

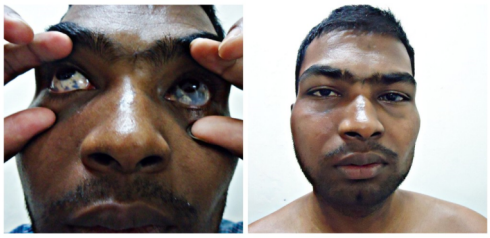

Figure 2: Clinical image showing bilateral nevus of Ota and Hypertrophy of the right side of the face

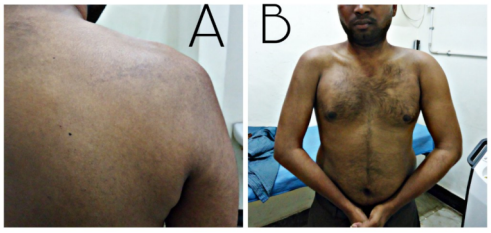

Figure 3: A- Nevus of Ito on the right shoulder
B- Right upper limb and pectoral muscle atrophy

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