NAEVUS OF OTA*

BY

G. P. GUPTA AND D. N. GANGWAR

From the Muslim University Institute of Ophthalmology and Gandhi Eye Hospital, Aligarh, India

THE naevus of Ota is characterized by benign mesodermal melanosis of the skin of the face with pigmentation of the eye and its adnexa. It is probably more familiar to the dermatologists.

Pusey (1916) was the first to describe the association of scleral and facial pigmentation in a young Chinese student. Later, in Japan, Ota and Tanino (1939) reported, under the name of naevus fusco-caeruleus maxillo-facialis, a case of "pigmented naevus" occurring in the skin areas supplied by the ophthalmic and maxillary divisions of the trigeminal nerve which was associated with the pigmentation of the eye. Jensen and Haffly (1948) also reported the occurrence of ocular and dermal melanosis in a 47-year-old Chinese man and suggested that oculo-cutaneous melanosis was a more descriptive term for this entity. Fitzpatrick, Zeller, Kukita, and Kitamura (1956) and Cowan and Balistocky (1961) preferred the term oculo-dermal melanocytosis. This syndrome of oculo-cutaneous pigmentation has subsequently been reported in the dermatological literature as the naevus of Ota. We report here a further two cases.

Case Reports

Case 1, a 5-year-old boy, was first examined in the out-patients' department of Gandhi Eye Hospital, Aligarh. The parents sought advice about the greyish pigmentation of the sclera of the left eye.

On general examination, the child was of average build. The left half of the scalp, as seen in the photograph, showed bluish-grey pigmentation which did not blanch on pressure (Fig. 1). The child had a Mongolian spot of bluish-grey colour in the lumbo-sacral region (Fig. 2) and the left arm. On ophthalmic examination vision appeared normal in both eyes. There was obvious pigmentation of the left eye, and the sclera of that eye showed scattered, irregular flecks of pigmentation all round the limbus (Figs 3 and 4). The parents said that the pigmentation had been present since birth.

Slit-lamp examination of the left eye showed the iris to be of a very deep brown colour that was darker than that of the iris of the other eye. The normal pattern of the iris was obscured. The iris crypts were obliterated and the surface of the iris was covered with minute, rounded elevations. The whole iris appeared thicker and more solid. The pupillary reaction in the left eye was less brisk than that of the other eye.

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FIG. 1.—Pigmentation in left half of scalp.

FIG. 2.—Mongolian spot in lumbosacral region.

FIGS 3 and 4.—Irregular flecks of pigmentation of sclera all round limbus in left eye.

On ophthalmoscopy, the fundus was a darker grey colour than in the right eye and pigment was deposited in the lamina cribrosa of the optic disc (Fig. 5).

Gonioscopy could not be done.

The child was re-examined after six months and it was found that the flecks of greyish pigmentation of sclera had been transformed into uniform diffuse pigmentation.

Case 2, a 52-year-old woman (Fig. 6), was first seen in the out-patients' department of Gandhi Eye Hospital, Aligarh. The patient sought consultation for her defective vision and needed refraction.

External examination revealed numerous discrete, pigmented spots over the right cheek and the bridge of the nose. The obvious pigmentation of the right eye was characterized by patchy dark brown discoloration (Figs 6 and 7).

Tests of vision gave the following results: right eye, 6/18; left eye, 6/60. The aided vision after refraction was: right eye, plus 1.00 D. Sph. plus 1.00 D. Cyl. Axis 180° 6/9; left eye, minus 8.00 D. Sph. 6/36.

Slit-lamp examination revealed pigmentation of the sclera and conjunctiva. The iris pattern was obscured and the crypts and collarette were not seen. The iris was heavily pigmented.

On gonioscopy the angle of the right eye appeared heavily pigmented as if smeared with a dark paint. The anterior and posterior trabeculae could not be differentiated. The normal architecture

FIG. 5.—Fundus photograph of left eye showing pigmentation at optic disc.

FIG. 6.—Pigmentation of upper half of sclera in right eye and pigmentation over bridge of nose.

FIG. 7.—Pigmentation of lower half of sclera in right eye and pigmentation over bridge of nose.

FIG. 8.—Fundus photograph of right eye showing pigmented spot at temporal margin of disc.

of the angle could not be determined. The angle was quite wide open. The intra-ocular pressure and water-drinking provocative test for both eyes were normal.

On ophthalmoscopic examination the fundus of the right eye appeared darker as compared with the left. There was a pigmented spot at the margin of the disc (Fig. 8).

Discussion

The naevus of Ota is a definite clinical entity of facial skin pigmentation commonly combined with pigmentation of the globe.

Dorsey and Montgomery (1954) state that the naevus of Ota is essentially an aberrant Mongolian spot located on the side of the face and involving the eye.

These Mongolian spots are most commonly seen in the sacral region but they may occur in other parts of the body; when they do the term aberrant Mongolian spot is used. The association described in our first case of a Mongolian spot in the sacral region and in the left arm with pigmentation of eye and the scalp strengthens the view that the naevus of Ota may be considered as an aberrant or misplaced Mongolian spot. This type of pigmentation is atavistic.

The naevus of Ota is common in Japan. No evidence of inheritance has yet been established, but by far the greatest number of cases occur in females (4:1). It is usually unilateral, and is found in all races but is more common in those that are darkly pigmented. Bilateral occurrence, although rare, is reported. Cutaneous pigmentation usually follows the distribution of first, second, and—rarely—the third divisions of the 5th cranial nerve, but there are cases in which pigmentation of the ear drum, buccal mucosa, palate, and nasopharynx was noted. The colour of the affected skin varies from light brown to dark brown, blue black, or slate colour. The boundaries of the skin lesion are usually poorly defined and variations in colour and extent may occur as a result of seasonal or hormonal disturbances such as puberty or the menstrual cycle. Usually, the skin pigmentation is flat, but in a few cases a nodular blue naevus has been present.

In the eye pigmentation is seen in the conjunctiva, sclera, cornea, iris, fundus, and optic disc. Any or all of these pigmentary changes may be present as a part of the ocular pigmentation of this syndrome.

The naevus of Ota is usually present at birth, but it may develop later in life at puberty or in pregnancy. The extent of melanosis varies from an almost completely grey or black pigmentation of the sclera to a small, single naevus like a grain of millet.

The condition is generally considered to be benign and harmless except from a cosmetic point of view. However, Dorsey and Montgomery (1954) reported two cases in which malignant changes in the skin in the naevus were seen. They described the histological appearance of the naevus of Ota as varying from that of a Mongolian spot to that of a cellular naevus. In a Mongolian spot, naevus of Ota, or blue naevus, the pigment consists of large melanin granules and is present in special dendritic or fusiform cells. These are buried deep in the corium and a layer of clear connective tissue and epithelium lies between them and the surface. The basal layer of epidermis which forms the pigment is not involved.

The naevus of Ota has to be differentiated from the condition of melanosis oculi and pre-cancerous melanosis. Though melanosis oculi is a similar condition, it is not associated with pigmentation of the face. As described by Reese (1951), melanosis oculi is a unilateral, congenital increase in the pigment cells throughout the globe and sometimes the conjunctiva and the lids and the extra-ocular muscles.

About 25 per cent. of cases of melanosis oculi turn malignant in middle age, while malignancy is rare in the naevus of Ota. Slit-lamp examination and biopsy may serve to distinguish the mesodermal or stromal melanoblast type of pigment found in the naevus of Ota from the granular, superficial, golden brown, ectodermic type found in pre-cancerous melanosis. Pre-cancerous melanosis is sensitive to radiation while the naevus of Ota is not.

Summary

Two cases of naevus of Ota are presented and discussed, and the literature is reviewed.

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