



Clinical Image

Phacomatosis pigmentokeratotica



Fig. 1. (A) Yellowish-brown verrucous papules and plaques arranged linearly along lines of Blaschko, suggestive of naevus sebaceus. (B) Naevus sebaceus over the right pre-auricular area (inset showing similar lesion over anterior chest).



Fig. 2. Light brown-coloured patch of size 10 × 7 cm with overlying dark brown-to-black macules and papules, suggestive of speckled lentiginous naevus.

A 20 yr old female[†] presented to the department of Dermatology and Sexually Transmitted Diseases, Safdarjung Hospital, New Delhi, India, in February 2019, with progressive asymptomatic brownish-black lesions on the face, neck and chest since birth. Examination revealed yellowish-brown verrucous papules and plaques arranged linearly along the lines of Blaschko on the forehead, nose, right pre-auricular area, neck and chest, suggestive of naevus sebaceus (Fig. 1A and B). Light brown-coloured patch of size 10 × 7 cm with overlying dark brown-to-black macules and papules, suggestive of speckled lentiginous naevus, was present on the left side of the face, extending from forehead to mandibular region (Fig. 2). Biochemical, ophthalmic, neurological and skeletal evaluation revealed no abnormality. Histopathological examination from verrucous plaque

showed hyperkeratosis, acanthosis, papillomatosis and follicular plugging in the epidermis. Dermis showed increased number of enlarged sebaceous glands placed abnormally higher, with a few immature hair structures and dilated apocrine glands (Fig. 3). These features were consistent with naevus sebaceus.

Phacomatosis pigmentokeratotica (PPK) is a rare form of epidermal naevus syndrome characterized by coexistence of an organoid naevus with sebaceous differentiation along Blaschko's lines and speckled lentiginous naevus in a checkerboard pattern. Our case represents a rare form of PPK without any extra-cutaneous features. Patients with PPK are at risk of developing basal cell carcinoma and syringocystadenoma papilliferum in naevus sebaceous and malignant melanoma in speckled lentiginous

[†]Consent to publish clinical information and images obtained from the patient.

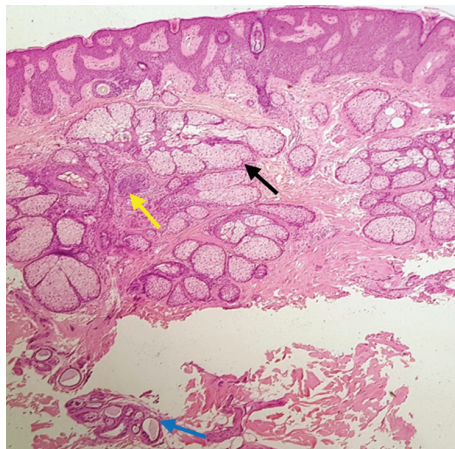


Fig. 3. Hyperkeratotic, acanthotic and papillomatous epidermis with follicular plugging. There are an increased number of closely set large sebaceous glands (black arrow) that are placed abnormally higher in the dermis along with a few immature hair structures (yellow arrow). Lower dermis shows dilated apocrine glands (blue arrow) (Hematoxylin and eosin, x100).

naevus; hence, regular follow up is necessary. The patient was planned for serial surgical excision of naevus sebaceous.

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Conflicts of Interest: None.

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