AN UNUSUAL PRESENTATION OF NEVUS SEBACEUS

Cynthia Papali, M A Aleykutty, M Omana and James Joseph Karappally

A case of nevus sebaceus in a 12-year-old child showing an unusual honeycomb appearance, not described earlier is reported.

Key words : Nevus sebaceus, Honeycomb appearance.

The entity nevus sebaceus was described by Jadassohn[1] in the nineteenth century. It appears soon after birth[2] on the face or scalp as a smooth yellow patch. In childhood, it becomes a little raised with partial or complete hair loss, and by the onset of adolescence, it turns into a verrucous plaque. Many cases of nevus sebaceus develop benign and malignant tumours in adulthood. In one series, basal cell epithelioma was noted in 6.5% of the cases and squamous cell carcinoma and apocrine adenocarcinoma in a few others. Benign tumours noted were syringoma, syringocystadenoma and eccrine hamartomas.[3,4] Usually, nevus sebaceus is solitary and linear in configuration, but occasionally, they are multiple and may be rounded or irregular in shape. When multiple, the lesions are not only distributed on the head, but also on other parts of the body. At least a few of these lesions will be linear in shape.[5] Cases with extensive lesions have shown evidence of a neuro-cutaneous syndrome in the form of epilepsy, mental retardation, neurological defects and also skeletal abnormalities.[6,7] Some of the linear lesions may be combinations of a nevus sebaceus and epidermal nevus[8] which has also been reported to be associated with central nervous system abnormalities known as epidermal nevus syndrome.[9] Kuokkanen et al[10] have shown by computed tomography and X-ray studies that the central nervous system involvement in nevus sebaceus resembles that of tuberous sclerosis. Lansky et al[11] noticed involvement of other viscera like eyes, kidneys and heart in addition to the brain in nevus sebaceus, similar to that seen in tuberous sclerosis.

Histopathologically, nevus sebaceus shows the pattern of development of normal sebaceus glands in infancy, childhood and adolescence. In the first few years of life, the lesion shows well developed sebaceus glands.[12] In childhood the sebaceus glands appear greatly reduced in number and size. At this stage, incompletely differentiated hair structures are seen. Sometimes, they are in the form of cords resembling the embryonic stage of hair follicles. At times, the hair structures consist of a keratin-filled infundibulum showing buds of undifferentiated cells. At puberty, the lesion presents the most characteristic diagnostic features. The epidermis shows papillomatous hyperplasia and the dermis contains large numbers of mature and nearly mature sebaceus glands. In about 50% of the cases, buds of undifferentiated cells resembling basal cell epithelioma may be present in the dermis. They represent malformed hair germs. Ectopic apocrine glands develop in about 60% of cases. Sometimes, the apocrine glands are seen at a younger age. In adulthood, various types of appendageal tumors and malignant neoplasms are found to develop. Syringocystadenoma papilliferrum has been found in 8 to 19% of the cases. Less frequently, tumors like nodular hidradenoma, syringoma and sebaceus epithelioma are seen. Basal cell epithelioma is noted in about 5 to 7% of the cases. In many

From the Departments of Dermatology and Venereology, and Pathology, Medical College Hospital Trivandrum-695001, India.

Address correspondence to : Dr. Cynthia Papali.

cases, the basal cell epitheliomata are very small, clinically not apparent and show no aggressive growth pattern. Very rarely, squamous cell carcinoma and apocrine carcinoma are seen.[3,4]

Histogenetically, nevus sebaceus is considered to arise from the primary epithelial germ because of the frequent presence of abnormally developed pilo-sebaceus complexes, apocrine glands and the tumors derived from the apocrine glands like syringocystadenoma and syringoma.[13] Pinkus and Mehregan[14] use the term organoid nevus to such a combination when more than one adnexa and sometimes the epidermis and the connective tissue participate in the nevus formation. By the term nevus sebaceus of Jadassohn, attention is unduly focused on only one of the comopnents.[14]

Case Report

An orphan girl aged 12 years, moderately nourished, short statured and of average intelligence presented with asymptomatic lesions on the forehead and left cheek. She noticed two small lesions at about the age of four years and thereafter they were gradually increasing in size. There was no associated epilepsy. On examination, these lesions were in the form of two elongated, oval, smooth, yellowish plaques 4 cm×2 cm in size, situated on the right side of the forehead and left cheek. The surface of these plaques showed irregular septae which gave a striking honeycomb appearance to the lesion (Fig. 1). Multiple, small, firm, smooth papules were noted around the lesion on the left cheek and also on the left half of the upper lip. There was a linear, midline, firm, verrucous plaque involving both upper and lower lips and the chin. Another linear, verrucous, hyperpigmented plaque was found on the left ear lobule with slight extension upwards over the pinna and downwards on to the side of the face towards the angle of the mandible (Fig. 2).

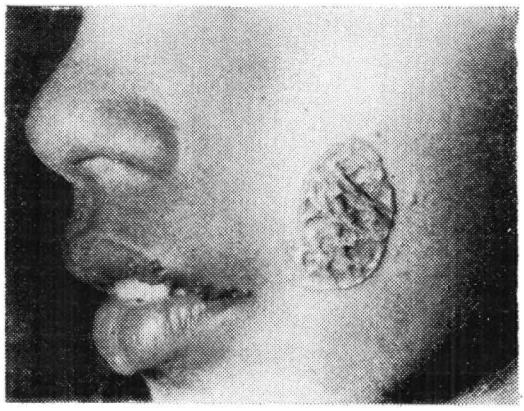

Fig. 1. The striking honeycomb appearance of the lesion.

Fig. 2. The verrucous lesion on the ear.

Several skin-coloured plaques were seen covering a major part of the scalp, the surfaces of which were smooth in some and uneven in others. One of the lesions was hairless and easily visible but the rest were covered with scanty hair and were not readily discernible (Fig. 3). A biopsy from the honeycomb like lesion on the forehead showed the following histopathological features.

The epidermis was atrophic at most of the places with flattening of the rete ridges. A few places

Fig. 3. A typical lesion on the scalp with alopecia.

showed acanthosis with irregular downgrowth of the rete ridges. At some places, there were keratin filled infundibula having multiple buds of undifferentiated cell groups (Fig. 4). In the upper dermis there were numerous mature sebaceus glands and a few immature hair stru-

Fig. 4. Skin section showing keratin-filled infundibulum with multiple buds of undifferentiated cell groups.

ctures resembling basal cells. There were also cords of undifferentiated cells resembling embryologic hair follicles. In the lower dermis, many mature apocrine glands were noted. Based on these histological features a diagnosis of nevus sebaceus was made.

Comments

The uneven and smooth, hairless and partially hairless plaques on the scalp seen in this case, are characteristic of nevus sebaceus and they undoubtedly point to the diagnosis. Lesions which are linear and verrucous, as seen in this case over the chin and on the left ear are also described in nevus sebaceus. The localisation of lesions on the face and scalp is also generally observed in nevus sebaceus. But the striking honeycomb appearance of the two lesions on the face, has not been described in the literature. In this case, the presence of characteristic lesions elsewhere gave a clue to the correct diagnosis, which was established by the biopsy. A slight variation present in the histopathological features was in the epidermal atrophy noted at most of the places, which probably corresponded to the depressions in between the septae. It is worth recording that a solitary, purely honeycomb-like lesion as seen in this case can mislead the clinician to a wrong diagnosis if one is not aware of such a presentation in nevus sebaceus.

References

1. Jadassohn J : Bemerkungen Zur histologie der systematisierten naevi and uber "Talgdrusen-Naevi", Arch Dermatol Syphilol, 1895; 33 : 355-394.
2. Robinson SS : Nevus sebaceus (Jadassohn), Arch Dermatol Syphilol, 1932; 26 : 663-670.
3. Jones EW and Heyl T : Nevus sebaceus, A report of 140 cases with special regard to the development of secondary malignant tumors, Brit J Dermatol, 1970; 82 : 99-117.
4. Mehregan AH and Pinkus H : Life history of organoid nevi : Special reference to nevus sebaceus of Jadassohn, Arch Dermatol, 1965; 91 : 574-588.
5. Lentz CL , Altman J and Mopper C : Nevus sebaceus of Jadassohn : Report of a case with multiple and extensive lesions and an unusual linear distribution, Arch Dermatol, 1968; 97 : 294-296.
6. Feurstein RC and Mims LC : Linear nevus sebaceus with convulsions and mental retardation, Am J Dis Child, 1962; 104 : 675-679.
7. Marden PM and Venters HD : A new neurocutaneous syndrome, Am J Dis Child, 1982; 112 : 79-81.

8. Wauschkuhn J and Rohde B : Systematisierte Talgdrusen: Pigment, und epitheliale Naevi mit neurologischer symptomatik; Feuerstein—Mimscheo neuroektodermale syndrom, Hautarzt, 1971; 22 : 10-33.

9. Solomon LW, Fretzin DF and Dewald RL : The epidermal nevus syndrome, Arch Dermatol, 1968; 97 : 273-285.

10. Kuokkanen K, Koivikko M and Alavaikko M : Organoid nevus phakomatosis, Acta Dermatol Venereol (Stockh), 1980; 60 : 534-537.

11. Lansky LL, Funderburk S, Cuppage FE et al : Linear sebaceus nevus syndrome : A hamartoma variant, Am J Dis Child, 1972; 123 : 587.

12. Lantis S, Leyden J, Thew M et al : Nevus sebaceus of Jadassohn, part of a new neurocutaneous syndrome Arch Dermatol, 1968; 98 : 117-123.

13. Haber H : Verrucous naevi, Trans St. John's Hosp Derm Soc, London, 1955; 34 : 20.

14. Pinkus H and Mehregan AH : Adnexal nevi and benign adnexoid tumours, in : A Guide to: Dermatohistopathology, Second ed, Appleton Century Crofts, New York, 1976, p 519.