LETTERS TO THE EDITOR

NEVUS SEBACEUS WITH CRIBRIFORM APPEARANCE SINCE BIRTH

Nevus sebaceus of Jadassohn is an uncommon lesion which manifests as a circumscribed, round or oval plaque on the scalp or face. In 60% of the cases, it is present at birth, while in the remaining, it appears by early childhood. During infancy and childhood, the lesion is a smooth, waxy plaque with alopecia. At puberty however, the surface becomes warty or covered with numerous, small, firm to rubbery nodules which persist throughout life.[1,2] We saw a patient with an extensive lesion and the unusual feature that the papillomatous processes were present right from birth.

An 8-month-old boy born after a full term normal delivery, had at birth, two ill-defined plaques covered with papillomatous processes

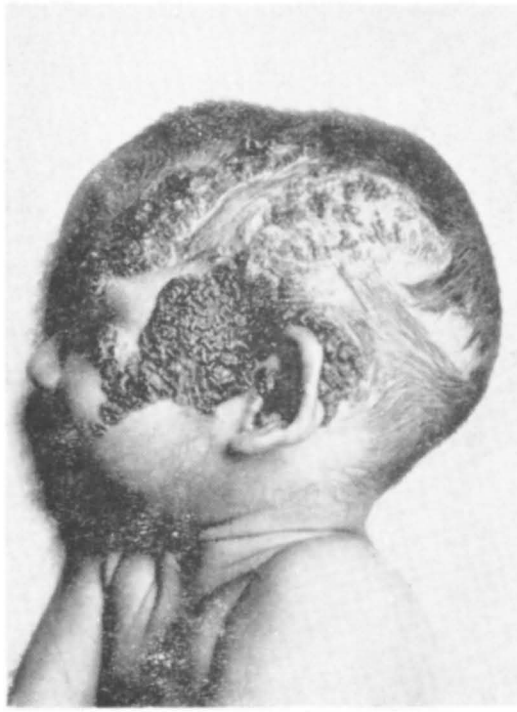

Fig. 1. Nevus sebaceus.

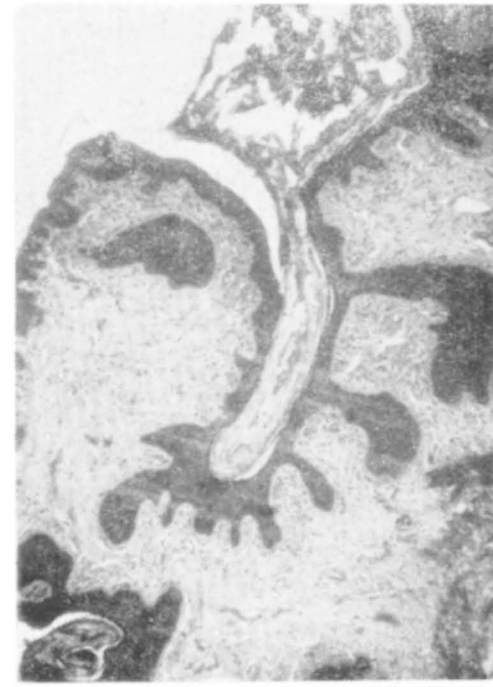

Fig. 2. Multiple, kerative-filled infundibula (H & E x 100).

on the left side of the scalp extending from the occipital region on to the left cheek. The parents had also noticed a few isolated papillomatous lesions on the left side of the nose. The lesions had been growing proportionately with the growth of the child. When seen by us, the plaques were 10×6 cm and 7×4 cm in size, present on the left side of the scalp, and extended from the occipital region to the lateral part of the forehead and the cheek. The surface of the lesion was covered with soft, skin-coloured papillomatous processes 0.3-0.5 cm in size, most prominent on the face and relatively sparse on the scalp, giving a cribriform appearance (Fig. 1). There were no hairs on the lesion. In addition, there was an irregular 0.5—1 cm wide area of

non-cicatricial alopecia running all around the lesion on the scalp. There were 6 papillomatous lesions arranged in a 2.5 cm linear band on the left side of the nose. There was no mental retardation or any history of epilepsy. There were no obvious cranial or ocular abnormalities. Histopathological examination of a biopsy from the scalp revealed several keratin-filled infundibula showing multiple buds (Fig. 2). The dermis contained many plump fibroblasts. The picture was highly suggestive of the childhood type of nevus sebaceus.

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References

1. Mehregan AH and Pinkus H : Life history of organoid nevi (Special reference to nevus sebaceus of Jadassohn), Arch Dermatol, 1965; 91 : 574-588.
2. Papali C, Aleykutty MA, Omana M et al : An unusual presentation of nevus sebaceus, Ind J Dermatol Venereol Leprol, 1984; 50 : 167-170.

LEG ULCERS IN KLINEFELTER'S SYNDROME

Chronic leg ulcers are commonly associated with a large number of systemic and dermatological conditions. We observed a patient with recurrent leg ulcers who had features of Klinefelter's syndrome. An 18-year-old male refugee from Bangladesh presented with two large ulcers, one on each leg of 2 months duration. Past history revealed that during the last 8 years, he had similar recurrent ulcers preceded by oedema and tenderness of both the legs. Bed rest and symptomatic therapy resulted in healing of the ulcers. Patient was ill-nourished and had bilateral, discrete, non-tender, inguinal lymphadenopathy. IQ was normal. Peripheral pulses and blood pressure were normal. He had no beard and had a feminine distribution of pubic hair and fat, and left-sided gynaecomastia. Right testis was absent and the left was pea-sized. The ulcer was irregularly oval, 6×4 cm in size, with pigmented borders and unhealthy granulation tissue discharging sero-sanguineous material, situated over the lower lateral part of the right leg. A similar ulcer, 8×8 cm in size, was seen over the anterior aspect of the middle of the left leg. Multiple scars of previous ulcers were present on both the legs. There was no evidence of varicose veins or peripheral vascular disease. Complete hemogram, platelet count and adhesiveness, peripheral smear for abnormal RBCs, X-rays of the chest and both legs, and arteriovenograms of the lower limbs were normal. Blood VDRL was non-reactive. Biopsy from the edge of an ulcer showed epidermal hyperplasia and a chronic inflammatory infiltrate in the dermis. Buccal smear for Barr bodies was positive.

Klinefelter's syndrome is characterised by hypogonadism, that includes gynaecomastia, eunuchoidism, elevated serum and urinary gonadotrophins and dysgenesis of seminiferous tubules. Recurrent leg ulcers may be one of the presenting symptoms of Klinefelter's syndrome. This heightened susceptibility to hypostatic ulceration is associated with increased frequency of varicose veins, arterial insufficiency and deep vein thrombosis.[1-3] Varicose veins were not present in our patient and the sites were also atypical for it. Past history suggestive of deep vein thrombosis was present. However, in one case of Klinefelter's syndrome with leg ulcers, there was no evidence of underlying arterial or venous disease.[1] The reason for the increased frequency of vascular abnormalities in Klinefelter's syndrome is not known. It has been hypothesised that abnormal levels of sex-hormones[3] may play a role or the increased number of X-chromosomes may interfere in some way with the clotting mechanism.[5,6] Recent reports of subarachnoid haemorrhage in

Klinefelter's syndrome[7,8] point to an inherent weakness of the blood vessels. Since Klinefelter's syndrome is an uncommon condition, care must be taken to exclude this possibility in males with unexplained recurrent leg ulcers.

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References

1. Howell R : Hypostatic ulceration and Klinefelter's syndrome, Brit Med J, 1978; ii : 95-96.
2. Campbell WA, Newton MS and Price WH : Hypostatic leg ulceration and Klinefelter's syndrome, J Ment Def Res, 1980; 24 : 115-117.
3. Campbell WA and Price WH : Venous thromboembolic disease in Klinefelter's syndrome, Clin Genet, 1981; 19 : 275-280.
4. Leigh IM and Sanderson KV : Klinefelter's syndrome and leg ulceration, Brit J Dermatol, 1979; 101 : 34-35.
5. Mantle DJ, Pye C, Hardisty RM et al : Plasma factor VIII concentrations in XXX women, Lancet, 1971; i : 58-59.
6. Clayton JK, Anderson JA and Mc Nicol GP : Preoperative prediction of postoperative deep vein thrombosis, Brit Med J, 1976; ii : 910-912.
7. Price WH, Steer AJW and Wilson J : Subarachnoid hemorrhage in Klinefelter's syndrome, Lancet, 1980; ii : 300.
8. Mark B : Vascular abnormality in Klinefelter's syndrome, Lancet, 1982; ii : 491.

GIANT MOLLUSCUM CONTAGIOSUM

We observed an interesting clinical presentation of molluscum contagiosum which is described here. A 5-year-old, apparently normal male child had eruptions over the neck of 4 months' duration. The lesions started as mildly pruritic, flesh-coloured, papular lesions which gradually progressed over the same site to form nodular masses. The lesions were soft, non-tender, well circumscribed, irregularly lobulated elevations, disposed in a linear pattern over the sides and back of neck. A solitary lesion, 3 mm in size, showing the typical features of molluscum contagiosum was seen over the centre of the nape of neck. Excision biopsy of one of the bigger lesions confirmed the diagnosis.

The average size of the lesions in molluscum contagiosum is 3-5 mm, though these can gradually increase upto 10 mm. Other uncommon variations in morphology include pedunculation in older lesions, or formation of lobulated masses by coalescence. Rarely, tumor masses of sizes as large as 2 or 3 cm have been observe.[1,2] It is worthy of note that giant lesions of molluscum contagiosum may be confused with keratoacanthomas or sometimes even basal cell epithelioma[3] and our case illustrates this point well.

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References

1. Friedman-Kien A : Molluscum contagiosum, in : Clinical Dermatology, Editors, Demis DJ, Crounse RG, Dobson RL et al: Harper and Row, Maryland, Unit 14-13; 1972; p 1-6.
2. Ormsby OS and Montgomery H : Molluscum contagiosum, in : Diseases of the Skin, Lea and Febiger, Philadelphia, 1954; p 622.
3. Pillsbury DM, Shelley WB and Kligman AM : Molluscum contagiosum, in : Dermatology, WB Saunders, Philadelphia, 1956; p 702.