

carcinomas (SCC). We are reporting this case because of rarity. Recently there was a case report of malignant melanoma of skin and SCC of the eye arising from limbus in an adult XP patient.²

A 6-year-old male, youngest child of a consanguineous parents had multiple freckles and hypopigmented atrophic macules on sun exposed parts of the body since 4 years of age. Parents have 2 male and 2 female children, 3 children developed XP, 1 male child is healthy.

Child had photophobia, blepharospasm and increased lacrimation. Developmental milestones were normal and no neurological manifestations.

Patient developed a small nodular growth 1 month back, situated at 5 O'clock position at the limbus of left eye. During 1 month, it attained the size of 1.5 cm X 1 cm grayish brown raised growth encroached upon cornea completely and growth was protruding out about 0.5 cm (Fig. 1). Child had pain,

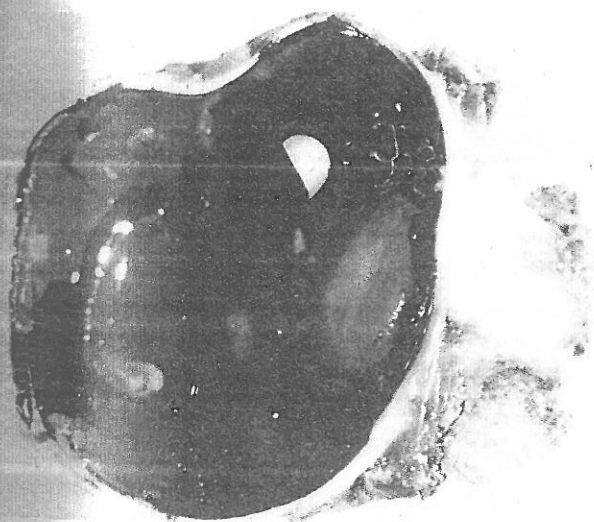


Fig. 1. Vertical section of eyeball with growth.

irritation and could not close the eye. There were no metastases.

Routine investigations were normal including LFT, X-ray chest was normal, skin

biopsy confirmed the diagnosis of XP. Enucleation of eyeball was inevitable. Histopathology of the growth revealed as well differentiated SCC.

Neoplasm of the eye in XP confined almost exclusively to the conjunctiva, cornea and eyelids, those portions of the eye exposed to ultraviolet radiation. These tissues shield the iris, lens and retina from ultraviolet radiation.

Unique review of 830 published cases of XP in a span of 108 years by Kraemer et al¹ revealed that neoplasms occurred most frequently at the limbus followed by the cornea and conjunctiva. The most frequent histologic type reported was SCC.

M M Udagani, V G Govekar
Consultants' Chambers, Shivaji Road,
Belgaum - 590 002, India.

References

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PAPILION-LEFEVRE SYNDROME

To the Editor,

In 2 cases of Papilion-Lefevre syndrome slightly different morphological features were seen by me. Both the patients are brothers; 1 is 6-years-old and other is 6-months-old. Parents are not consanguineous. Psoriasiform lesions are present not only on classical sites but on many other areas over the body in the elder child. Because of very rarity (only 10 cases are reported upto 1988 from our country).¹ These cases are discussed here.

A 6-year-old boy and his 6 months

brother were brought to skin department for thickness and scaliness of palms and soles. On examination, palmoplantar hyperkeratosis with erythema and scales are noticed in both children. In elder child - psoriasiform scales are present on palms, soles, both knees, elbows, over tibia and malleoli. Within 1 month after coming to us patient developed psoriasiform plaques in many areas on the body. All deciduous teeth are lost except deciduous canine on right side of lower jaw, and permanent 1st molar on left side of lower jaw. X-ray mandible showed permanent tooth bud of 1st and 2nd premolar. No calcification of falx cerebri was seen.

Papillon-Lefevre syndrome is an autosomal recessive disorder of keratinization characterised by erythematous scales, thickness of palms and soles, psoriasis like lesions on elbow, knees, weakness of periodontal ligaments and teeth loss with calcification of falx cerebri. Localized disorders of keratinization such as mal de maleda, Unna Thost, Papillon - Lefevre syndrome may have strong relation with psoriasis.

Not only the present thinking of disordered leucocyte function, disordered gingival fibroblast, and cementoblast function in P-L syndrome, some other etiological factor such as Zinc deficiency in Acrodermatitis enteropathica may come into our notice in future. In this contest, response of psoriasis to linolenic acid may be thought of. In my patients cutaneous lesions responded well with external application of MF3 ointment (moisturizing ointment) retinoic acid, ointment, oral beta carotene, oral alfacin capsules (Linolic + Linolenic acid) massage of sunflower oil before bath.

V V Narasimha Rao

*Skin Specialist, Civil Assistant Surgeon,
District Hospital, Machilipatnam - 521 001.*

KERATOTIC PAPULES ON CHIN A NEURODERMATITIS OR DYSMORPHOPHOBIA?

To the Editor,

The letter on keratotic papules on chin (KPC) by M M Udagani published in *IJDV* 1993: 59: 45 was interesting. I wish to report a case of KPC which may throw some light on the aetiopathogenesis of the condition. A female student aged 18 years presented with asymptomatic bilateral skin coloured follicular papules on the chin of 9 months' duration. Size was that of a rupee coin and the skin between the papules showed mild hyperpigmentation. though the patient denied any rubbing, her parents had noticed her constantly picking at the hairs on the chin. Further queries revealed that the girl was deeply worried about the unwanted hair growth on her face.

To reassure her, the prominent hairs on the chin were removed by electrolysis. Pimozide 2mg as a single morning dose along with topical Tretinoin (0.05%) produced almost complete clearing of her lesions in 6 weeks, leaving only a little residual hyperpigmentation. With only alternate day application for another 1 month and a follow up period of 2 months, there was no recurrence.

Predominance of the condition in female teenagers with one or other congenital or acquired blemishes on their face,¹ the particular localization on the chin, the typical appearance of chronic follicular keratinization due to constant rubbing, and the psychological disturbance all point towards the tentative diagnosis of a neurodermatitis. Chin is the most easily accessible area for students sitting with their elbows on the desktops and the chin resting between the thumb and forefinger. Modesty may be preventing the