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NEVOID BASAL CELL CARCINOMA SYNDROME

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A 65-year-old male developed nevoid basal cell carcinoma syndrome manifesting as multiple basal cell epitheliomas, marked mutilation of the face and characteristic pitting on the palms and soles. Calcification of the falx cerebri and scoliosis of the lumbar spine, were also seen.

Key words : Basal cell carcinoma, Palmo-plantar pits, Scoliosis, Calcification, Falx cerebri.

The nevoid basal cell carcinoma syndrome is a well established multiple system disorder. Majority of the patients have been white skinned. It is an autosomal dominant disorder with a low penetration and manifests as multiple skin tumours and palmo-plantar pits, with or without multiple skeletal and central nervous system anomalies, along with a variety of less common changes. There is paucity of reports from our country. We observed a case having calcification of the falx cerebri and scoliosis of the lumbar spine.

Case Report

A 65-year-old male patient developed an ulcerated, crusted, easily bleeding lesion, on the right side of his face for ten years. To begin with, the lesion was small, raised, pigmented and located on the right lateral aspect of the nose. Despite treatment which included excison, the lesion gradually increased in size over the years, and about seven years back, it became ulcerated with a serous discharge and bleeding on trauma. For the last four months, the lesion had started extending onto the nose also. There was no tendency to subside. He also noticed small, circumscribed depressions on the palms for the last six years and multiple naevi on the face for the last fifteen years. Family history was non-contributory. Patient had above average intelligence and venereophobia. The spine showed scoliosis to the right in the lumbar region. The cutaneous lesions were bilateral but asymmetrical and confined mainly to the right side of the face, nose and forehead (Fig. 1). There were multiple naevi

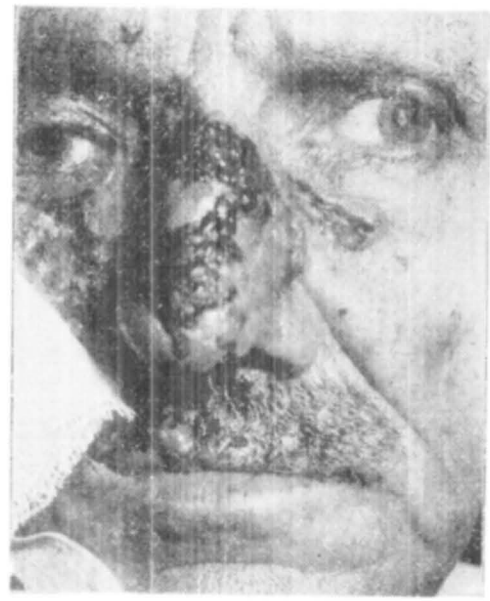

Fig. 1. Multiple naevi on face, involvement of right lower eyelid and destruction of right alae nasi.

seen as smooth surfaced, pigmented, raised oval papules and nodules 2 mm to 1 cm in size. There was no telangiectasis. One lesion on the forehead showed a central depression. A few similar lesions were also seen on left side of the face. One linear lesion along the left lower eyelid showed a central depression and crusting. The main lesion was situated over the right

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maxilla in the form of an ulcer with a well defined margin which was raised but not rolled out. Its base was indurated and non-tender and the floor showed haemorrhagic crusting and a sero-purulent discharge. The ulcer had invaded the right lower eyelid and the right side of the nose destroying the alae nasi. Destruction of the deeper tissues was seen in the lower half of the lesion. Both the palms and soles had multiple, more or less circular pits about 1 mm deep and 1 mm to 4 mm in diameter with almost perpendicular edges (Fig. 2). Floor of the pits was skin-coloured.

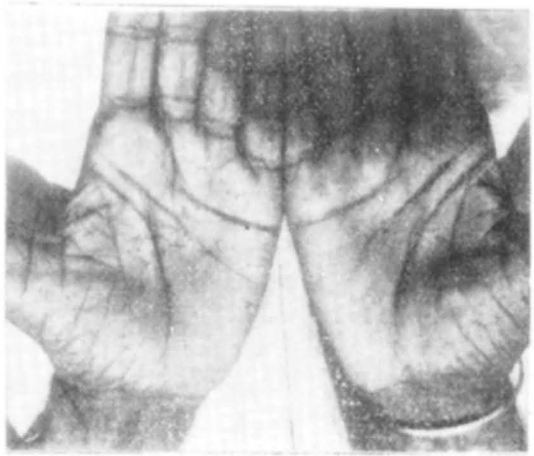

Fig. 2. Characteristic multiple palmar pits.

Routine investigations on blood and urine were normal. VDRL test was negative. X-ray skull and face showed calcification of the falx cerebri (Fig. 3) but no bony erosion. X-ray of the spine showed scoliosis of the lumbar spine to the right. Skin biopsy from the margin of the ulcer showed tumour masses of various shapes and sized embedded in the dermis (Fig. 4) the peripheral cell layer showing a palisade arrangement. No connection of the tumour masses to the surface epidermis could be shown. This picture was consistent with solid basal cell epithelioma.

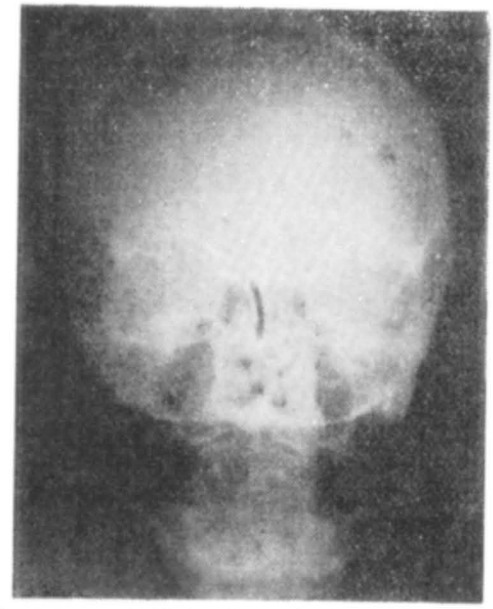

Fig. 3. X-ray skull showing calfication of falx cerebri.

Comments

Naevoid basal cell carcinoma syndrome is a genetically determined disorder but positive family history was lacking in our patient. Though skin tumours usually start appearing in childhood or at the latest at puberty, our patient developed lesions at the age of fifty years. Similar late onset was reported by Kamath et al.[1] All the cutaneous lesions

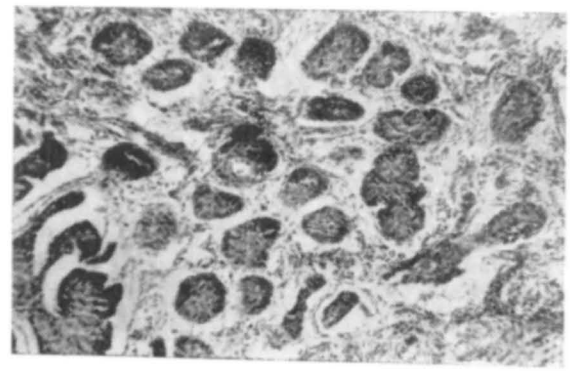

Fig. 4. Solid basal cell carcinoma (H and E 60 X).

in our case were localised to the face and most of them were in the nevoid stage with a few showing a neoplastic change, the lesion over the maxilla had become invasive, destructive and mutilating.

Pits of the hands and feet are characteristic and diagnostic of this syndrome and are present in approximately one half of the adult patients with this disorder.[3] Nearly all patients with this syndrome have been shown to have multiple skeletal and central nervous system abnormalities,[4] important among these being odontogenic cysts of the jaw, anomalies of the ribs, scoliosis or kyphosis, spina bifida occulta, mental retardation; calcification of the falx cerebri,[5] cerebellar medulloblastoma, fibrosarcoma of the mandible/maxilla, hypertelorism with broad nasal root and frontal bossing. Abnormalities of uterus and ovaries, mesenteric cysts and hypogonadism in males, have also been reported. Mason et al[2] reported that all the diverse features of basal cell carcinoma such as solid, adenoid, cystic, keratotic, superficial and fibrosing formations may be seen in this syndrome. This rare syndrome is far less common in the dark races, and this is the second confirmed case report from India. It is necessary that all cases having multiple naevi or basal cell carcinomas must be properly followed up over long periods to search for other components of the syndrome.

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