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Pemphigus Vegetans masquerading as Cutis Verticis Gyrata

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Introduction :

Pemphigus vegetans is an uncommon variant of pemphigus vulgaris comprising of only 1-2% of cases.

Case Report :

A 75 year old female presented with verrucous plaque with cerebriform surface over scalp involving parietal area and extending into frontal area of size 10 x08 cm with a surrounding depigmentation with perifollicular pigmentation(figure 1). The lesions started ten year back as a fluid filled bullae initially over scalp and later involving the chest and back and was diagnosed as a case of pemphigus vulgaris based on clinical and histopathological finding . She was treated with systemic corticosteroid with resolution of lesions with pigmentation except over scalp which underwent vegetation following which patient was lost for follow up . A month back she presented with persistent scalp lesion . Systemic findings were insignificant. Histopathology showed hyperkeratosis, keratotic plugging, irregular acanthosis, suprabasal bulla with basal cells arranged in row of tomb stone appearance with mixed inflammatory infiltrate containing eosinophils, neutrophils and mononuclear cells(Figure 2) . Direct immunoflorescence revealed intercellular IgG deposits in epidermis (Figure 3) . CT scan of chest and head were normal .

Ultrasonogram abdomen and pelvis was insignificant . Routine haemogram and liver and renal function test were within normal limits . She never had mucosal lesions any time during the course of her disease.

Discussion :

Cutis verticis gyrata(CVG) is a descriptive term for a condition of scalp in which deep furrows and convolutions are seen which cannot be flattened by traction or pressure, resembling the surface of cerebral cortex. The anatomy of scalp and its attachments are in such a way that infiltrate beneath the epidermis produces folds and gyri. The direction of the folds are usually anteroposterior but may be transverse over the occipital region[1] and also rarely irregular[2]. Based on the surface appearance it can be of two broad types, the first one, maximum folds in the midline and extending symmetrically but less marked in all directions peripherally, as in our case. The second type is a definitive uncircumscribed tumour with raised abrupt border with convolutions being more prominent and furrows more deeper more closely simulating cerebral surface.

It is classified by Diven et al as Primary essential (Aetiology not known, no associated features), Primary non-essential (Aetiology not known, associated with mental, cerebral and eye abnormalities) and Secondary described with underlying causes, such as inflammatory scalp condition (Eczema, psoriasis, pemphigus, folliculitis, erysipelas, impetigo, hamartoma, neurofibroma), nevoid abnormalities, acromegaly, post-trauma, idiopathic hypertension, osteoarthropathy (Pachydermoperiostitis), amyloidosis, syphilis, leukemia, fallopian tube carcinoma, acanthosis nigricans, tuberous sclerosis. A report of CVG following misuse of anabolic substances, hyper-IgE syndrome, Noonan Syndrome, Neurofibroma[3], has been described. In Polan and Butterwoth's review[4] of 195 cases of CVG, they classified 47.7% as Primary and 52.3% as Secondary. The largest segment of the latter group were associated with tumours/ naevi/ fibromata (17%), acromegaly (14%), and inflammatory conditions (10%). Our case comes under the latter.

Pemphigus vegetans a variant of pemphigus vulgaris historically has been divided into Neumann type, which begins and ends as Pemphigus vulgaris, which contains pustules in early stage, and Hallopeau type, which is relatively benign, having pustules as primary lesions instead of bullae. Then development is followed by gradually enlarging verrucous lesions in intertriginous areas. On histopathology,

suprabasal clefts with acantholysis along with numerous eosinophils with villi and verrucous epidermal hyperplasia are seen. Direct immunoflorescence examination reveals squamous intercellular Ig in all reported cases. The treatment of pemphigus vegetans is the same as that for pemphigus vulgaris with oral corticosteroids and various other drugs azathioprine, dapsone and parenteral gold. Our patient was managed with corticosteroids and was maintained with azathioprine. In view of her old age, we have not initiated pulse therapy with corticosteroids.

Pemphigus vegetans is more resistant to treatment than pemphigus vulgaris or pemphigus foliaceous. EGFR and TGF alpha are excessively expressed throughout the epidermis in pemphigus vegetans which may be responsible for the vegetating character[5.]

Conclusion:

To our knowledge, there are no reports of pemphigus vegetans presenting as CVG .

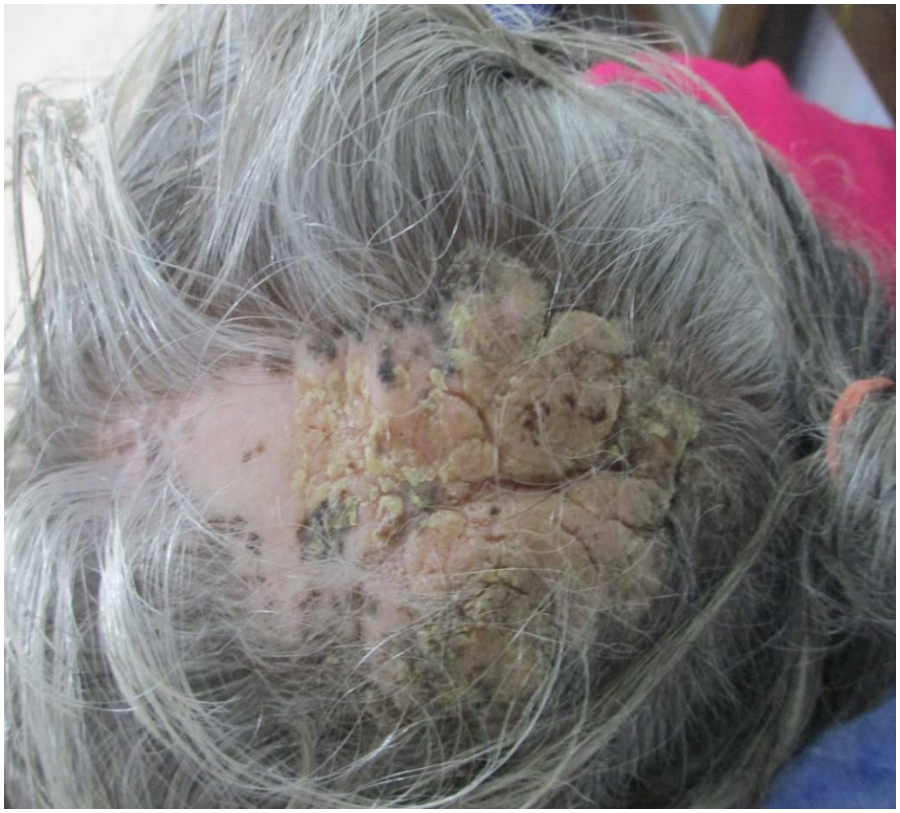

Figure 1 - verrucous plaque with cerebriform surface over scalp with a surrounding depigmentation with perifollicular pigmentation

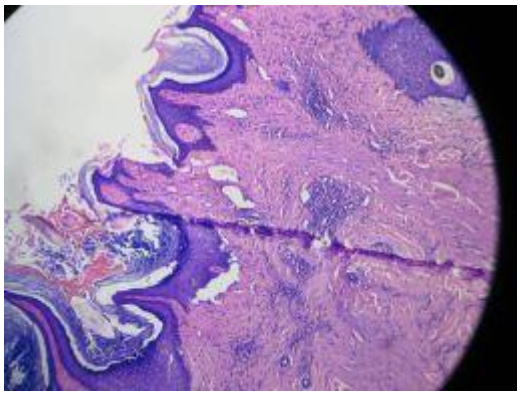

Figure 2 : Histopathology showing hyperkeratosis, keratotic plugging, irregular acanthosis, suprabasal bulla with basal cells arranged in row of tomb stone appearance with mixed inflammatory infiltrate . (H& E , 10 X)

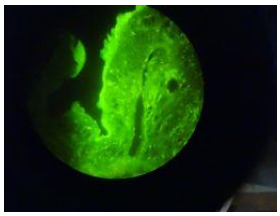

Figure 3 : Direct immunoflorescence showing intercellular IgG deposits in epidermis (10X)

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