Cutis Verticis Gyrata in a Patient with Hyper-IgE Syndrome

Hei Sung Kim, Rachael Yu Lin Teo and Audrey Wei-Hsia Tan
National Skin Centre, Singapore, 1 Mandalay Road, Singapore 308205. E-mail: hazelkimhoho@gmail.com
Accepted February 2, 2009.

Sir,

Cutis verticis gyrata (CVG) is a rare morphological condition of the scalp characterized by ridges and furrows resembling the surface of the brain. We report here a case of CVG in a patient with hyper-IgE syndrome, which is a multisystem disorder known to affect the dentition, skeleton, connective tissue and immune system. The presence of CVG in hyper-IgE syndrome has not been described previously.

CASE REPORT

A 50-year-old Malay man presented with a deformity of the scalp, which had started during adolescence. He had no personal history of mental retardation, seizures or head injury, and denied a family history of a similar scalp condition. Physical examination revealed scaly folds and grooves arranged in an antero-posterior direction over the scalp, which could not be flattened with pressure (Fig. 1). Hair growth was sparse overlying the folds. The patient refused a skin biopsy.

The patient had had a history of sensitive skin since childhood, which subsequently evolved into extensive atopic dermatitis. The eczema was poorly controlled and he had been hospitalized once for eczema herpeticum, which responded well to acyclovir. There was no associated asthma or allergic rhinitis. Coarse facial features with deep furrows on the forehead/glabella area had been noted for more than 30 years (Fig. 2) and dental extraction was performed to remove a double row of teeth. He had a history of recurrent infections, including soft tissue abscesses and septic arthritis (left ankle, fingers), interstitial keratitis and pulmonary tuberculosis. Persistent eosinophilia and raised IgE were noted during routine medical check-ups. No other members of his family had been diagnosed with hyper-IgE syndrome.

Investigations revealed leukocytosis (27.3×10^9/l, normal range: $6–14 \times 10^9$/l, 65% neutrophils) with mild eosinophilia (1.40×10^9/l, normal range: $0.04–0.4 \times 10^9$/l) and markedly elevated IgE (> 5000 IU/ml, normal range: 0–29.2 IU/ml). C-reactive protein (21.7 mg/dl, normal range: 0–1 mg/l) was also raised. Chest radiograph showed right upper lobe fibrosis consistent with old pulmonary tuberculosis, but did not show any active focus of infection. Findings (urinalysis, thyroid function test, skull radiographs) were otherwise within normal limits.

We diagnosed the patient with hyper-IgE syndrome with CVG of the scalp. The CVG is currently managed with meticulous shampooing.

DISCUSSION

CVG describes a scalp condition in which there is hypertrophy and folding of the skin that resembles the surface of the cerebral cortex (1). It is classified as primary essential (aetiology not known, no associated features), primary non-essential (aetiology not known, associated with mental, cerebral and eye abnormalities) and secondary (2). Secondary CVG has been described with underlying causes, such as inflammatory scalp conditions (eczema, psoriasis, pemphigus), nevoid abnormalities and acquired tumours (3). A variety of endocrine disorders, such as acromegaly, myxoedema

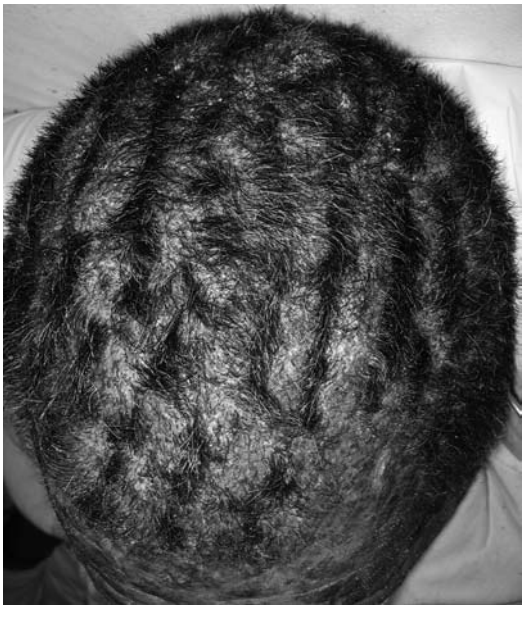

Fig. 1. Tight grooves and folds of the scalp resembling that of the cerebral cortex.

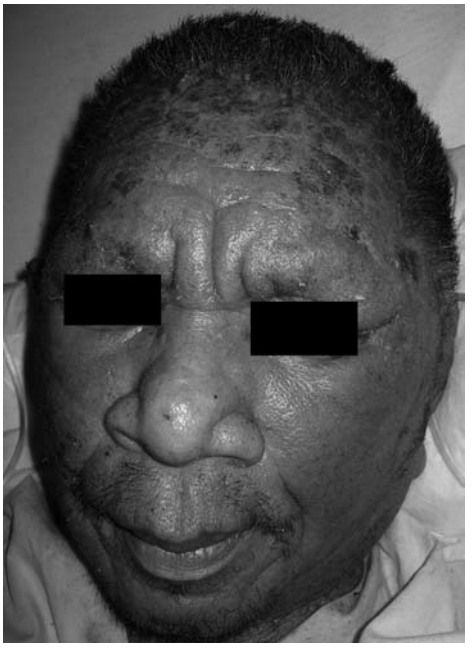

Fig. 2. Characteristic coarse facies of hyper-IgE syndrome: a prominent forehead with folds on the glabella, deep-set eyes, a broad nasal bridge, fleshy nasal tip and oily skin.

and insulin resistance, have also been reported in association with CVG (4).

Secondary CVG can also occur as a part of pachydermoperiostosis (idiopathic hypertrophic osteoarthropathy), a syndrome of digital clubbing, periosteal new bone formation, coarse facial features, furrows of the face/scalp and hyperhidrosis of the palms and soles (5). It is classified into primary (hereditary) or secondary (provoked by an underlying pulmonary disease). With the coarse facial features and deep furrows on the forehead/glabella, we initially considered pachydermoperiostosis as one of our differentials, but the lack of palmoplantar hyperhidrosis and the absence of the characteristic bone changes (thickening of the phalanges, clubbing, acro-osteolysis) made the diagnosis less likely. Such facial features were later found to be associated with the patient's concurrent hyper-IgE syndrome.

The recommended approach to the patient with CVG includes a personal and family history, physical examination and skin biopsy (to identify an underlying nevoid abnormality or tumour) from the affected area. Laboratory (complete blood count, blood chemistry, IgE) and further haematological, endocrinological (thyroid function, glucose/ insulin level) or radiological studies are required depending on the presenting features of the individual patient.

Hyper-IgE syndrome is a rare multisystem disorder with approximately 250 cases previously reported in the literature. It was first described in 1966 by Davis et al. (6) when 2 patients were reported with eczematous dermatitis, recurrent staphylococcal abscesses, hyperextensible joints and coarse faces. The diagnosis is usually made clinically as there is no specific genetic or confirmatory test available. Eczematous or atopic dermatitis-like eruptions, abnormal dentition (retained primary teeth, double rows of teeth), coarse faces (prominent forehead, deep set eyes, broad nasal bridge, fleshy nasal tip), skeletal abnormalities (osteoporosis, scoliosis), recurrent infections (lung-recurrent pneumonias and pneumatocele formation, skin-cold abscesses) and immunological abnormalities (raised IgE, peripheral eosinophilia) are the characteristic features of hyper-IgE syndrome and can be found in a variable degree (7).

The relationship between CVG and hyper-IgE syndrome is not clear. The fact that CVG has never been reported as a feature of hyper-IgE syndrome suggests a chance association, but extension of the coarse facial folds to the scalp may have caused the development of CVG in the case described here.

REFERENCES

1. Larsen F, Birchall N. Cutis verticis gyrata: three cases with different aetiologies that demonstrate the classification system. Australas J Dermatol 2007; 48: 91–94.
2. Garden JM, Robinson JK. Essential primary cutis verticis gyrata: treatment with the scalp reduction procedure. Arch Dermatol 1984; 120: 1480–1483.
3. Diven DG, Tanus T, Raimer SS. Cutis verticis gyrata. Int J Dermatol 1991; 30: 710–712.
4. Woollons A, Darley CR, Lee PJ, Brenton DP, Sonksen PH, Black MM. Cutis verticis gyrate of the scalp in a patient with autosomal dominant insulin resistant syndrome. Clin Exp Dermatol 2000; 25: 125–128.
5. Rimoin DL. Pachydermoperiostosis (idiopathic clubbing and periostosis): genetic and physiologic considerations. N Engl J Med 1965; 272: 923–931.
6. Davis SD, Schaller J, Wedgewood RJ. Job's syndrome: recurrent, "cold" staphylococcal abscesses. Lancet 1966; 1: 1013–1015.
7. Grimbacher B, Holland SM, Gallin JI, Greenberg F, Hill SC, Malech HL, et al. Hyper-IgE syndrome with recurrent infections – an autosomal dominant multisystem disorder. N Engl J Med 1999; 340: 692–702.