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Cutis Verticis Gyrata in Acromegaly an infrequent finding

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Abstract

Cutis verticis gyrata is a rare clinical manifestation presenting with cerebral convolution like appearance of the scalp.It can be primary or secondary.We report a case of 41 year old male with acromegaly who had cutis verticis gyrata.

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Introduction

Cutis verticis gyrata (CVG) is a rare disease with an estimated prevalence of 1 in one hundred thousand in males and 0.026 in one hundred thousand in females[1]. It is characterised by excessive soft tissue proliferation resulting in redundadnt and thick scalp skin folds resulting in cerebriform appearance of scalp..We report a case of 41 year male who had acromegaly with cutis verticis gyrata.

Case Report

A 41 year male presented with insidious onset,slowly progressive acral enlargement,excessive sweating, joint pains,deepening of voice,proximal muscle weakness and headache. On physical examination he had skin thickening with cerebral convolution like appearance on the scalp (Figure 1), prominent supraorbital ridges, prognathism, widely spaced incisors, coarse facial features, multipe skin tags, enlarged hands and feet, sweaty palms, deep sonorous voice, bitemporal hemianopia & kyphosis.

On evaluation his Insulin like Growth Factor 1 (IGF 1) was 468 ng/mL, random Growth Hormone (GH) level was > 1600ng/mL. After 1 hour of 75g oral glucose tolerance test (OGTT) GH was 449 ng/mL. MRI pituitary showed a pituitary macroadenoma of 3.8(CC)x 2.6(AP) x 3.6 (ML) cm (Figure 2).

Discussion

Cutis verticis gyrata also known as paquidermia verticis gyrata, cutis verticis plicata, and "bulldog" scalp syndrome.It is characterized by thick skin folds on scalp giving the appearance of cerebral gyri and sulci[2]. Primary CVG can be essential or non essential.

Primary non essential form is associated with neurological & ophthalmological manifestations, whereas primary essential form is not associated with these manifestations. Secondary CVG can occur in pachydermoperiostosis, acromegaly, myxedema, acanthosis nigricans, amyloidosis, type 2 diabetes mellitus cerebriform intradermal nevi, neurofibromas, Touraine Solente Gole syndrome and leukemia.

In acromegaly, CVG occurs in 14 to 30%[3]. Excess IGF 1 and GH levels in acromegaly leads to diffuse soft tissue proliferation[4]. CVG in acromegaly can be managed by decreasing the GH and IGF1 levels. This patient was not willing for surgery and underwent conventional radiotherapy and now was on medical management with somatostatin analogues for acromegaly.

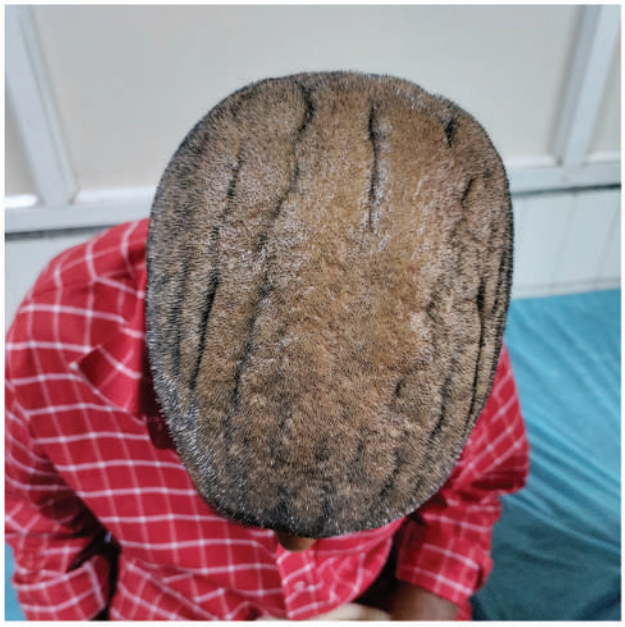

Figure 1: Scalp of patient with acromegaly showing cutis verticis gyrata.

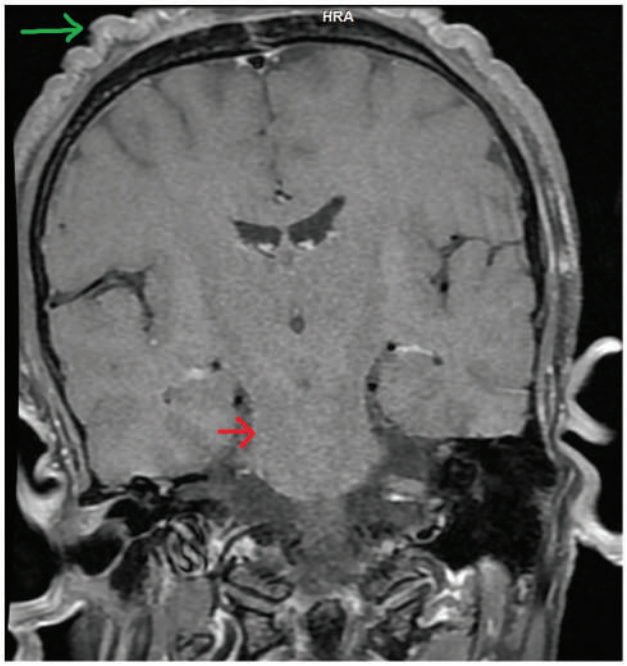

Figure 2 : MRI brain coronal section showing pituitary macroadenoma (red arrow) and cutis verticis gyrata (green arrow).

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