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A Rare Case of Acromegaly Presenting with Cutis Vertics Gyrata

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Abstract- The morphologic changes in the scalp which characterizes cutis vertics gyrata was first described by Robert[1] in 1848 and McDowell[2] and Cown[3] in microcephalic idiots.

In 1906 Judassohn[4] called to the attention of dermatologists the condition was given the name as cutis vertics gyrata by Unna[5]

Fisher stated the belief that true cutis verticis gyrata occurs as a developmental anomaly, probably atavistic in type, representing a reversion to a lower form of life in which the muscles of the scalp could move at will. This is probably the type originally described as present in microcephalic idiots.

Index Terms- Acromegaly, Cutis, Gyrata, Skin, Scalp, Verticis

I. CASE REPORT

We present here a case of young man, who came to our clinic with initial features of thickening of the skin.

An 18 year old male patient, presented with complaints of

- Folds on the forehead since 18 months
- Thickening of the skin of the face since 18 months
- Thickening of the skin over the hands and legs since 18 months

There is no similar history in the family, no history of trauma, fever, hospitalization and medication.

On examination, deep folds of the skin present over the forehead and scalp, with deep groves and thickening of the skin were found to be present.

Figure 1 At Age 16

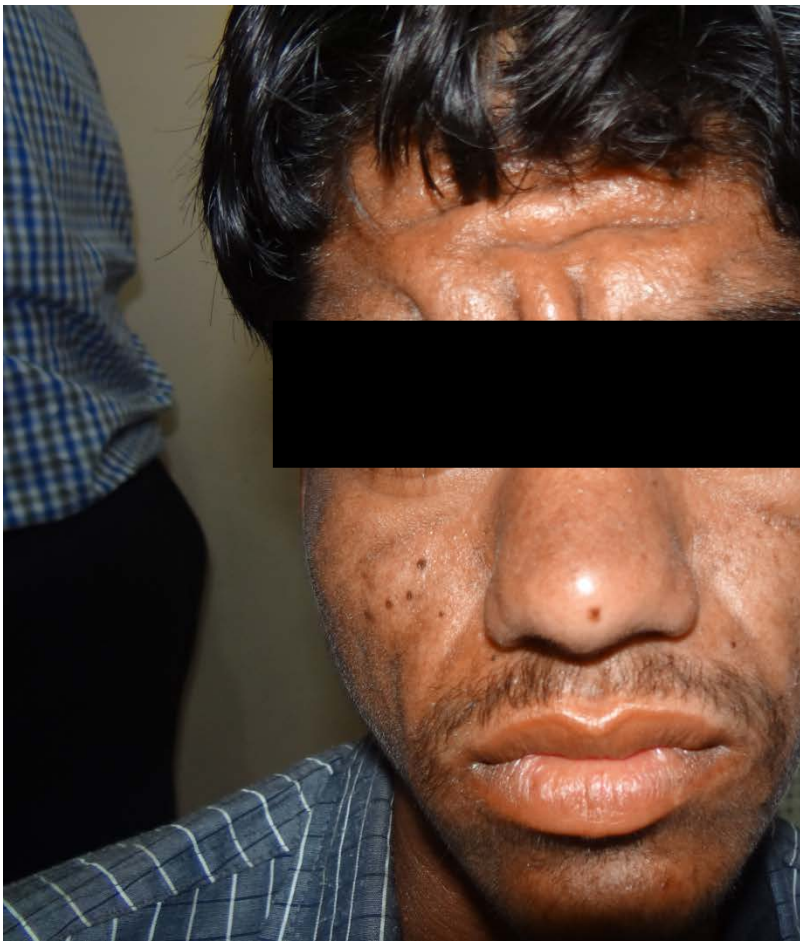

Figure 2 At Age 18

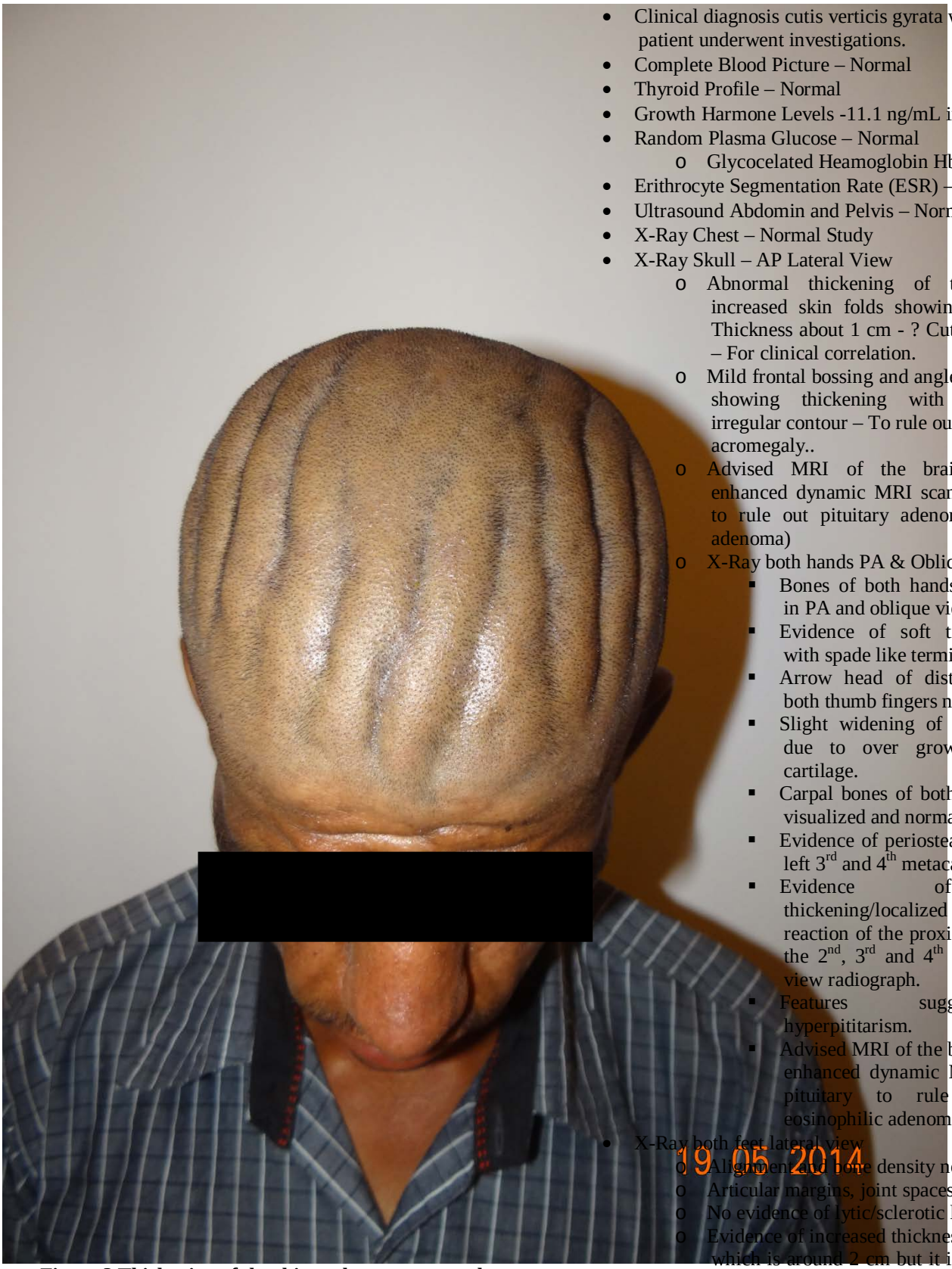

Figure 3 Thickening of the skin and grooves on scalp

- The patient has coarse facial features, wide nasal bridge and thick palms and soles.
- Clinical diagnosis cutis verticis gyrata was made and the patient underwent investigations.
- Complete Blood Picture – Normal
- Thyroid Profile – Normal
- Growth Harmone Levels -11.1 ng/mL increased
- Random Plasma Glucose – Normal
  - Glycocelated Heamoglobin HbA1c – Normal
- Erithrocyte Segmentation Rate (ESR) – Normal
- Ultrasound Abdomin and Pelvis – Normal Study
- X-Ray Chest – Normal Study
- X-Ray Skull – AP Lateral View
  - Abnormal thickening of the scalp with increased skin folds showing wavy margin. Thickness about 1 cm - ? Cutis verticis gyrata – For clinical correlation.
  - Mild frontal bossing and angle of the mandible showing thickening with splaying with irregular contour – To rule out mild features of acromegaly..
  - Advised MRI of the brain and contrast enhanced dynamic MRI scan of the pituitary to rule out pituitary adenoma (eosinophilic adenoma)
  - X-Ray both hands PA & Oblique View
    - Bones of both hands well visualized in PA and oblique views.
    - Evidence of soft tissue thickening with spade like terminal tufts noted.
    - Arrow head of distal phalanges of both thumb fingers noted.
    - Slight widening of the joint spaces due to over growth of articular cartilage.
    - Carpal bones of both wrist joint well visualized and normal.
    - Evidence of periosteal reaction of the left 3rd and 4th metacarpal noted.
    - Evidence of cortical thickening/localized periosteal reaction of the proximal phalanges of the 2nd, 3rd and 4th noted in oblique view radiograph.
    - Features suggestive of hyperpititarism.
    - Advised MRI of the brain and contrast enhanced dynamic MRI scan of the pituitary to rule out pituitary eosinophilic adenoma.
- X-Ray both feet lateral view
  - Alignment and bone density normal
  - Articular margins, joint spaces normal.
  - No evidence of lytic/sclerotic lesions
  - Evidence of increased thickness of the heal pad which is around 2 cm but it is not recorded as a sign of acromegaly.
  - Evidence of cortical thickening of the lower 3rd of tibia and fibula noted.
- MRI Brain

- No significant anomaly seen in the brain
- Bilateral mastoiditis is seen with patchy mucosal disease in the paransal sinuses.
- Note is also made of the increased thickness of the skin of the scalp region.

II. Discussion

Cutis verticis gyrata is a problem mostly found in men. It is about five or six men for one woman.

Possible case of cutis verticis gyrate in this case of acromegaly with excess growth harmone.

The patient was very muscular and considered well built. Patient had features, indicating acromegaly, broadening of his facial features. An examination revealed gyriform oblong skin folds in the fronto parietal region.

Primary CVG, a syndrome occurring almost exclusively in males, has been associated with mental deficiency (the IQ is never greater than 35), cerebral palsy, seizures and ophthalmological abnormalities. It is present in 0.5% of the retarded population in Sweden, Scotland and the USA. There is no evidence for an endocrine influence in primary CVG. Pachydermoperiostosis has often been confused with primary CVG, but this affects not only the skin of the scalp but also the skin of the face, hands and feet; in addition there are accompanying periosteal features. Secondary CVG, has been associated with local disorders of the scalp, most commonly inflammatory conditions such as eczema and psoriasis and with hypertrophy and folding of the scalp as a result of congenetial naevi, acromegaly, amyloidosis, and acute myeloid leukaemia, It is treated by surgical reduction of the affected area and results are good. The approach to a patient with CVG should include not only a detailed clinical examination but also a scalp biopsy if an underlying naevoid disorder is suspected.

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