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SKULL RADIOGRAPHY OF PATIENT WITH ACROMEGALY- UNUSUAL HAIR ON END APPEARANCE.

Neurosurgery

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ABSTRACT

Acromegaly is clinical disorder caused by excess secretion of growth hormone by mostly pituitary macroadenoma. Growth hormone has significant effect on bone growth and remodeling which is evident on X-ray. We report hair on end appearance as a new radiological image finding of longstanding untreated acromegaly, which is not previously described in this disorder.

KEYWORDS

Acromegaly, Skull Radiology, X-Ray, Hair on end.

Clinical case Summary: Acromegaly is a hormonal disorder that results from post-pubertal chronic excess of growth hormone (GH) secretion, which most commonly occurs due to GH-secreting pituitary macroadenoma. It is a rare disorder affecting both sexes equally, with recorded prevalence of >13 cases per 100,000 individuals [1]. Because of insidious onset of clinical features, patients are usually diagnosed 5-10 years later after the first symptom develops[2].

31 years old female diagnosed to have acromegaly 2.5 years back when she presented with diabetic ketoacidosis. History of headache, acral enlargement and coarse facial features evaluation revealed excess IGF-1, non-suppressible growth hormone levels and pituitary macroadenoma. After two and half years of untreated acromegaly, X ray Skull revealed enlarged Sella turcica, thick calvarium, occipital perturbance, hair on end appearance of skull, frontal bossing, enlarged fontal sinuses, prognathism. This appearance with prognathism is many a times called as a "lantern jaw[3]" or "Habsburg jaw [4]", which can be seen in many other conditions and is not specific to acromegaly. Many a times the interdental space also widen, which is clinically known as a diastema.

The **hair on end sign**[5] indicated thick trabeculae which lies in perpendicular direction to skull vault interspersed with radiolucent expanded diploic space due to hyperplasia of marrow. This sign is classically described in conditions that results in extensive RBC turnover due to various conditions like hereditary spherocytosis, iron deficiency anaemia, enzyme deficiency, e.g. G-6-PD deficiency causing haemolytic anaemia, sickle cell disease and thalassemia major resulting in extramedullary haematopoiesis. However, this sign has not been earlier described in patients of acromegaly. Our patient had normal hemogram, serum iron levels, peripheral blood film, negative direct and indirect coomb's test, normal LDH levels and normal cardiac ejection fraction. Hence, this is a unique finding that can be observed with a patient of long-standing untreated acromegaly resulting in thick skull vault trabeculae and hair on end sign.

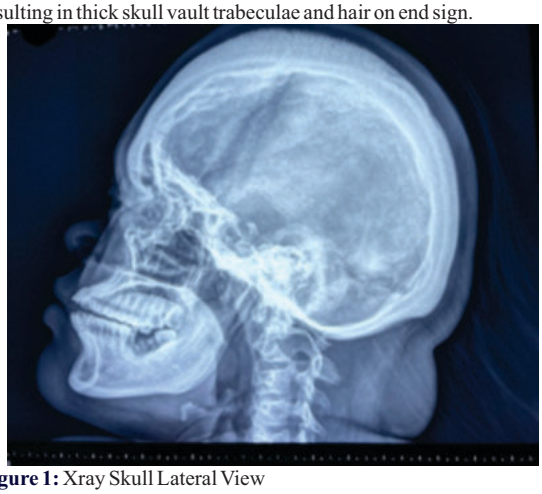

Figure 1: Xray Skull Lateral View

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