

An Unusual Skull with Acromegalic Features-A Case Study

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Abstract: *During routine osteology lectures, we came across a skull from osteology department which presented multiple unusual features, which closely resembled that of acromegaly. Inspired by the anomalous features of this rare endocrine disorder, the skull was given a closer look in this case study.*

Keywords: Skull, acromegaly

1. Introduction

Acromegaly is a very rare endocrine disorder, usually caused by a pituitary adenoma, leading to hyper secretion of growth hormone (GH) and consequently of insulin-like growth factor-I (IGF-1) after closure of epiphyseal growth plate, which plays an important role in regulation of bone metabolism. The anabolic actions of GH on many organ systems are well documented. GH and IGF-1 are important regulators of bone homeostasis through life. They are acting in autocrine and paracrine ways, stimulate proliferation, differentiation, and extracellular matrix production in osteoblastic like-cell lines and finally bone formation. During the childhood GH stimulates longitudinal bone growth. During the adolescence and early adulthood GH stimulates skeletal maturation till the achievement of peak bone mass-maximal bone mass (2). Its prevalence is 4 cases/million. Excess GH acts on osteocytes, chondrocytes and fibroblasts, stimulating the progressive growth of the skeletal system with anatomical and radiological changes. Musculoskeletal manifestations that occur in acromegaly are nonspecific and progressive (1). GH excess leads to increased bone turnover, defined by changes of bone markers. Articular manifestations are the main causes of morbidity and immobility of these patients, and they are persistent even after successful treatment. (5)

Symptoms of acromegaly include abnormal enlargement in bones of the extremities and head due to stimulated periosteal bone formation. Acral and soft tissue overgrowth, articular overgrowth, Enlarged viscera and Hypogonadism. Abnormalities of the bones of the skull are typically the most apparent bony changes. Acromegaly may also cause thickening of the soft tissues of the body, including the heart, lips, and tongue. (3, 4)

2. Case Report

The bony features of an acromegalic skull includes prominent mandible, enlarged frontal sinuses, increased calvarial thickness, prominent supraorbital ridge, prominent facial features, enlarged sella turcica and prominent occipital protuberance. Here, we got most of these features in the specified skull.

The Norma Frontalis shows Frontal bossing and prominent supra-orbital ridges with bilateral supra orbital foramen. The orbits were large and square shaped. Two accessory foramina are seen on the posterior aspect of medial wall of the orbit at the suture between orbital plate of frontal bone and lesser wing of sphenoid. The inferior aspect of nasal bones are protruding forwards anteriorly with a prominent anterior nasal spine. Body of Maxilla seems to be hollowed out with a prominent alveolar process which shows bilateral hollowing above lateral incisor and canine sockets. The body of zygomatic bone appears to be prominent and bulging.(fig.1)

Norma Occipitalis shows prominent tubercle like external occipital protuberance and a ridge like prominence superiorly just below the lambdoid suture lines.(fig.2)

Norma Lateralis shows Frontal prominence with frontal process of Zygomatic bone bilaterally presenting a bar like outgrowth in its posteromedial aspect which is directed backwards and medially. The Zygomatic arch is broad and the superior border of the zygomatic arch forms a prominent supra-mastoid crest the posterior aspect of which ends sharp and projecting as a spine over the parieto-mastoid suture. The inferior border presents a prominent tubercle followed posteriorly by an inverted -U shaped notch.(fig.3)

Norma Basalis shows a concave hard palate which is long antero-posteriorly than transversely with prominent palatine crests. Pterygoid hamulus is bilaterally prominent, more so in right side forming incomplete pterygo spinous bar. Body of Sphenoid is less broad anteriorly than posteriorly. Foramen Vesalius is present on the right side. Articular part of the Mandibular fossa is bilaterally absent with absent squamo-tympanic fissure. Occipital condyles are larger and oval, connected by a bony bar anteriorly. The left sided condyle appears to be grooved. The posterolateral borders of the Foramen magnum are raised and ends in three tubercles, two on right and one on left side of the midline posteriorly. Hypoglossal canals are bilaterally duplicated. Right posterior condylar canal is bridged by a bony bar and is intra-sinus type that opens into the sigmoid sinus and left posterior condylar canal is retro-sinus type opening behind the sigmoid sinus.(fig.4)

The base of the skull shows prominent frontal sinuses with sharp spine like anterior clinoid process. The pituitary fossa

appears to be destroyed. The vault shows prominent frontal sinuses and cranial vault thickening. The left side of the coronal suture shows synostosis. The vascular markings are bilaterally prominent with deep pits for arachnoid granulations. (fig.5)

The Mandible bone is missing, if available would have added much more information to this study.

All these bony changes could be correlated with the increased osteoblastic activity due to growth hormone hyper secretion which may be attributed to a pituitary adenoma as the pituitary fossa and area surrounding it seems to be deformed and destroyed.

3. Conclusion

The bony features of pituitary adenoma /acromegaly is often limited in textbook explanations as a hyperostosis due to increased osteoblastic activity. The clinical features are made out along with soft tissue changes in a living patient. No reports are there regarding the changes occurring in the cranium as an osteological entity. This case study looks into every possible details of the skulls in search for a shift from normal morphology. This throws new lights in the fields of osteology as well as aids in reasoning out various clinical features in these disease. Also it serves a guide to the radiologists in interpretation of acromegalic cases.

This skull is a still living signature, the disease had made over the patient who once lived and suffered it, telling us the story of most of the symptoms he had and what was behind all those, serving as a guide while coming across another case of the same, which makes this case study unique and valuable.

References

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Figures



Figure 1: Skull showing Norma Frontalis

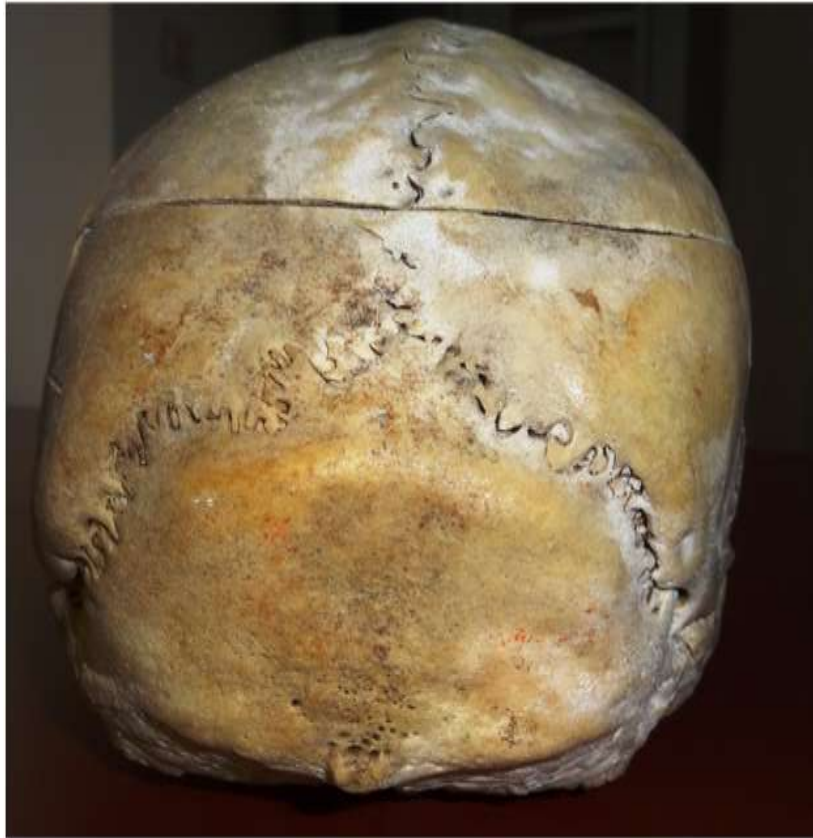


Figure 2: Skull showing Norma Occipitalis



Figure 3: Skull showing Norma Lateralis



Figure 4: Skull showing Norma Basalis



Figure 5: Skull showing the base with destroyed pituitary fossa