Fibrous Dysplasia of the Skull (Leontiasis Ossea)

Case History: A 12-year-old male complaining of skull expansion, facial deformity, nasal stuffiness, proptosis and visual impairment.

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![Figure A](image-url)
Figure B
Figure C
Figure D
Figure E
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Figure F

Findings: MDCT axial images (Figs. A, B) with sagittal (Fig. C), axial (Fig. D) bone window images and 3D reformatted images (Fig. E) show thick expanded all skull bones (sphenoid, frontal, maxillary, ethmoid bones, occipital, temporal bones) effacing the brain tissue and facial bones (mandible and maxilla), with diffuse ground glass appearance of the bones on bone window (Figs. C, D). Additionally noted encroachment upon the cranial nerves foraminae and orbital proptosis. Widened diploic space. Obliteration of paranasal sinuses. 3D reconstructed images show lion face appearance (Figs. E, F)

Diagnosis: Fibrous dysplasia of the skull (Leontiasis Ossea)

Discussion: Fibrous dysplasia is an abnormal bone growth where normal bone is replaced with fibrous bone tissue. The skull is a frequent site of the pathologic process now known as fibrous dysplasia and the skull changes are often the only manifestation of the disease. Fibrous dysplasia is a largely historical term used to describe a number of conditions which result in the affected patient's face resembling that of a lion. Although it is most frequently associated with craniofacial fibrous dysplasia, it has a broader meaning encompassing other lesions that have similar appearance.

In general the distinction is made between true leontiasis ossea (craniofacial fibrous dysplasia) and other conditions having similar external appearance (mimics), however it would be safe to say that as a term it is no longer of clinical use and should be avoided.

In addition to craniofacial fibrous dysplasia the following conditions may mimic leontiasis ossea:

• Paget disease
• gigantism
• craniosynostosis
• tumours of the paranasal sinuses
• syphilitic osteoperiostitis
• ureamia with secondary hyperparathyroidism

References

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