MASSIVE CRANIOFACIAL BONE DESTRUCTION WITH UNILATERAL FACIAL PARALYSIS IN GORHAM SYNDROME

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Objective
To report a case of Gorham–Stout Syndrome (GSS), presenting with multiple facial masses and unilateral right facial paralysis, its diagnostic criteria, and management.

Materials and Methods
Design: Case report
Setting: Tertiary government hospital
Patient: One

Results
- 55-year-old female
- 7-month history of masses in the right frontal and temporal areas with facial nerve paralysis from destruction of the temporal bone
- Histopathology: Osteolysis with vascular channel proliferation
- Management: Radiotherapy and antibiotics for concurrent pneumonia and possible CNS infection
- After 3 sessions of RT and 17 days of antibiotics, she succumbed to septic shock

Discussion
- GSS was defined by Gorham and Stout as an idiopathic condition with destruction of the osseous matrix of the bone due to angiomatosis
- Usual sites affected: pelvis, scapula and the skull
- Clinical presentation: pain, swelling, and functional impairment of the affected region
- Treatment options: surgical resection with bone grafting, anti-osteoclastic and anti-angiogenic medications, and radiotherapy
- Only 2 cases of GSS were published in the Philippines, prevalence is not identified

Table 1. Diagnostic Criteria by Heffez et. al. 5
- Positive biopsy
- Absence of cellular atypia
- Minimal or no osteoblastic response and absence of dystrophic calcification
- Evidence of local, progressive, osseous resorption
- Nonexpansible, nonulcerative lesion
- Absence of visceral involvement
- Osteolytic radiographic pattern
- Negative hereditary, metabolic, neoplastic, immunologic or infectious etiology

The case fulfilled 8/8 of the diagnostic criteria

Conclusion
Symptoms and presentation of GSS vary greatly on the site of the bones involved. The rarity and idiopathic nature of this case makes it difficult to catch. A good correlation of the clinical, histopathologic, and radiologic findings are essential in early detection of the disease to achieve control and limit functional problems for these patients.

References

Fig. 1a.
Fig. 1b.
Fig. 2a.
Fig. 2b.
Fig. 3a.
Fig. 3b.

Fig. 1a. Anterior view, evident asymmetry of the face at rest, 1b. Right lateral view.

Fig. 2.
3D reconstruction of the CT scan of the cranium showing massive osteolysis of the facial and skull bones. A. Anterior view, B. Posterior view.

Fig. 3.
Irregularly expanded nerve bundles with nodular appearance, prominent myxoid and vascular stroma, with overlying thick fibrovascular delineation, with no mitosis or atypia. A. LPO, B. HPO.

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