OPEN

CASE REPORT
Breast

Breast Reduction in a Patient with Gorham–Stout Vanishing Bone Syndrome

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Summary: The authors describe a 23-year-old woman with Gorham's syndrome who underwent an uneventful bilateral reduction mammoplasty which has not been reported in the medical literature today. The patient had undergone multiple surgical and medical interventions before presentation in the senior author's clinic including a vascularized free fibular graft which ultimately disappeared due to disease progression. Preoperatively, the patient complained of debilitating neck and back pain secondary to her macromastia, which was noted to be asymmetric. A standard inferior pedicle breast reduction was performed with the removal of 600 g from the right breast and 400 g from the left. The patient healed well postoperatively without complication and was satisfied with her result. *(Plast Reconstr Surg Glob Open 2014;2:e181; doi: 10.1097/GOX.0000000000000146; Published online 9 July 2014.)*

Initially described as its own entity in 1955,[1] Gorham–Stout is an extremely rare entity characterized by rapid osteolysis of one or more bones by vascular ingrowth.[2] Although mortality is uncommon unless vital structures are involved,[3,4] partial or even complete bone disappearance often occurs causing significant morbidity. Relatively little is known about Gorham's disease due to the scarce incidence and rarity of described clinical cases.[5] There seems to be no predilection based on patient sex, age, or gender,[6] and investigators have been unable to detect any familial inheritance pattern.[7] Any bone(s) can be involved, although long bones, pelvic bones, and scapulae tend to be the most commonly reported sites of disease.[6] Treatment with medical therapy, radiation, and surgery have all been attempted with varying success,[8] but the disease process still presents a challenge for physicians. We present a case of a female patient with Gorham's syndrome involving the left upper extremity who underwent a bilateral reduction mammoplasty for symptomatic macromastia.

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PATIENT REPORT

The patient is a 23-year-old woman with a documented history of Gorham's syndrome involving the left humerus, left clavicle, left first through third ribs, left scapulae, and cervical and thoracic vertebrae. She was initially diagnosed in 1993 from a magnetic resonance imaging scan obtained for infantile "cystic hygroma" and recurring left extremity cellulitis.

Before her evaluation in the senior author's clinic, the patient had undergone several surgeries including debridement of her diseased humerus bone with the placement of a prosthesis, multiple fracture repairs of the left upper extremity in differing locations, free vascularized left fibular graft to the left forearm which ultimately disappeared from disease progression, and a left wrist arthrodesis. Additionally, she had received 2 cycles of vincristine therapy in an attempt to slow the disease progression and multiple sessions of interferon-α. Notably, in 2002, the patient was enrolled in a clinical trial for Marimastat, an experimental matrix metalloproteinase inhibitor, but was removed from the study due to pericardial effu-

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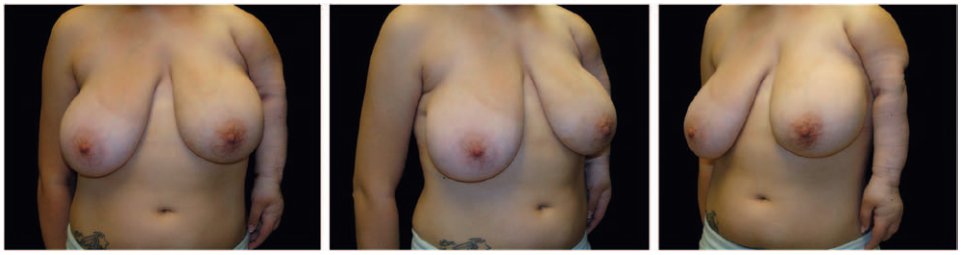

Fig. 1. Patient preoperatively showing asymmetric macromastia and Gorham's disease of the left upper extremity.

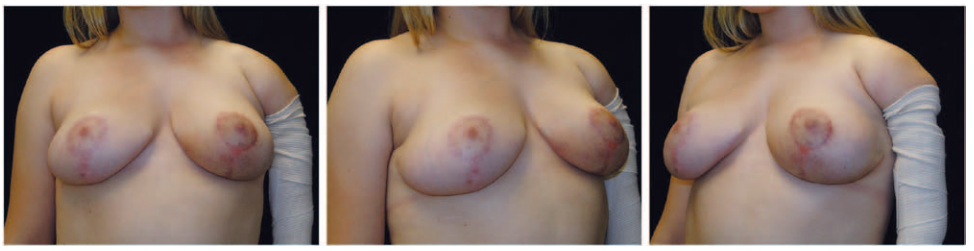

Fig. 2. Patient at 5 months postoperatively demonstrating reduction with good symmetry and healed scars. Notable hyperpigmentation of the skin of the left breast.

sion from mediastinal involvement which required interferon treatment. At the time of her recent presentation, her Gorham's disease was stable, but she was actively taking celecoxib, doxycycline, and peginterferon-2α for the antiangiogenic properties.

In the clinic, the patient reported debilitating neck and back pain secondary to her macromastia. Her preoperative breast size was 36 DD with significant asymmetry noted, likely resulting from her underlying Gorham's disease (Fig. 1). Operatively, a bilateral inferior pedicle technique with wise-pattern incisions was used to allow for ample reduction while simultaneously ensuring robust blood supply to the nipple. There were no operative complications, and a total of 600 g was removed from the right and 400 g from the left in an effort to correct her preoperative asymmetry.

The patient returned at 1 and 2 weeks after surgery with no complications and followed a standard postoperative course. She subsequently returned at 5 months, at which point it was determined that she was healing appropriately (Fig. 2). The patient expressed great satisfaction with her results, and she indicated that she had not experienced any complications from her reduction surgery as a result of her underlying Gorham's disease and associated medications.

CONCLUSIONS

We present for the first time a case of bilateral reduction mammoplasty in a patient with Gorham's vanishing bone disease, demonstrating efficacy and safety in this rare patient population. Additionally, the disease process may have resulted in significant parenchymal asymmetry requiring unequal reductions.

PATIENT CONSENT

Patients provided written consent for the use of their images.

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