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Anesthetic and obstetric management of a parturient with Gorham-Stout Disease

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Abstract Body: Gorham-Stout disease (GSD) is a rare pathological condition of unknown etiology characterized by proliferation of vascular channels of hematic and lymphatic origin in bone and adjacent soft tissues. This results in progressive and often massive bone destruction.1 It occurs in young adults but only 2 cases have been described during pregnancy.2,3 Clinical features depend on the area of involvement and presence of complications e.g. pleural effusions, chylothorax and generalized lymphangiomatosis. Patients may present with pathological fractures, neurological and respiratory complications.

Case Report: LH is a 30 yr Caucasian woman with a 16 yr history of GSD who presented at 18 wks’ gestation with an unplanned singleton pregnancy. Seven years earlier, occipital bone loss led to tumor debulking, occipital to C5 fusion with instrumentation, and spinal cord decompression. A year later she had stereotactic radiotherapy to slow bone resorption. One year before pregnancy, she developed a massive cystic hygroma, which required tracheostomy to prevent airway closure followed by IMRT radiation. A perinatologist coordinated obstetric management with other specialists. Anesthetic consultation revealed an otherwise healthy woman who was well informed about GSD. However, she had minimal head extension or flexion due to cervical spine fixation (see X-ray) and limited mouth opening (see photograph). Pregnancy proceeded without complication but our concern was airway obstruction in late third trimester. Case reports suggested a risk of severe PIH with thrombocytopenia at delivery, 4 and consumptive coagulopathy with secondary Kasabach-Merritt syndrome - which could mimic severe PIH.5 An elective tracheostomy was recommended but declined by LH. The plan was a CS at 35 wks’ gestation and to have an ENT surgeon present. LH was admitted 2 days prior to her planned CS with a fetus in breech position, triple nuchal cord, and early labor. A semi-urgent CS using spinal anesthesia ( 10mg hyperbaric bupivacaine, 20mcg fentanyl and 0.2mg morphine in sitting position) produced a healthy baby. LH was stable and comfortable throughout the 25 min surgery with an ENT surgeon present throughout. She did well post-CS, going home after 3 days. A medical oncologist has recommended bisphosphonate therapy to reduce further bone resorption.

References:
1. Papadakis SA et al. Orthopedics 2008; 31: 278

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