Introduction:
An encephalocele is most commonly secondary to a congenital anomaly of the bony structures of the skull leading to herniation of brain tissue. Infrequently encephaloceles are acquired because of trauma or infection. We present a previously not described case of an encephalocele secondary to Gorham’s disease.

Our Case:
A 17-year-old female presented for an encephalocele repair. She was diagnosed with Gorham's Disease 6 years prior, which led to the destruction of her left temporal bone with development of the encephalocele and extensive bony destruction in the head and neck area. The encephalocele had caused hearing loss and intractable seizures, which triggered the surgery. She underwent an uneventful induction and intubation with inline stabilization and placement of an arterial line and two large bone iv’s. Shortly after the resection of the encephalocele, the surgeon informed us that he noticed uncontrollable arterial bleeding, likely secondary to injury of a branch of the internal carotid artery. We immediately started the transfusion of PRBC and FFP. The surgeon packed the wound and interventional radiology performed a cerebral angiogram in the operating room to see if balloon occlusion of the internal carotid artery could be used to achieve hemostasis. The patient had abnormal posterior circulation and a balloon occlusion of the internal carotid artery would have resulted in a stroke. The surgeon was forced to pack again and close the cranium. The patient was cardiovascular stable throughout without any need for vasoressor support. A postoperative MRI demonstrated no signs of cerebral ischemia. The patient returned to the OR after 24 hours to remove the packing and evaluation. No active bleeding could be identified. The surgeon closed the cranium, the patient was extubated a day later and had no neurological deficits.

Discussion:
Gorham’s disease or vanishing bone disease is a poorly understood rare skeletal condition which manifests with massive progressive osteolysis along with a proliferation of thin walled vascular channels. There have been only 200 cases reported in the literature. The disease is idiopathic, non-familial without age, sexual or racial predilection. The diagnosis is based on clinical presentation, radiographic, and histological findings, specifically osteolysis with minimal osteoblast response, angiomatosis, and soft tissue swelling. Gorham's disease can involve the entire skeletal system and surgical intervention often becomes necessary to stabilize spine, facial bones or long bones. Tissue involvement can affect spleen, pleura and the lymphatic system. There are no standard treatment modalities available largely because of the continuous debate over its pathogenesis. Different strategies including early radiation, medical therapy and surgical resection have been attempted, with varying success. The prognosis is hard to predict because of a variable rates of progression. The symptoms may stabilize with time, regress or prove fatal especially when the spine or chest are involved. In cases of remission a recurrence is possible. Neurological complications as in our case can be seen when the skull or the spine are involved. Often times the disease leads to severe deformities and disability.

References: