

# Management of Severe Cerebral Compression in Extensive Craniofacial Fibrous Dysplasia



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## Introduction

Fibrous dysplasia (FD) is a bone disorder marked by progressive substitution of normal bone with irregular fibro-connective tissue, resulting in expansion, fractures, pain, and functional impairment of the affected skeletal area. Although FD can affect any bone of the body, craniofacial involvement is common. Surgical intervention is the primary treatment. When feasible, demolitive and reconstructive procedures are preferred over less aggressive techniques, such as bone curettage, as they significantly reduce recurrence rates. The two-stage surgical approach was selected based on the lesion's size and the gradual progression of symptoms. This approach allowed for reduced risks associated with a single complex procedure, improved cerebral decompression, and the opportunity to plan a custom-made cranioplasty.

## Case Presentation

A 46-year-old woman with McCune-Albright syndrome presented with frontal lobe symptoms and facial deformity. The symptoms gradually progressed over three years, becoming severely debilitating in the past 12 months. Imaging revealed extensive craniofacial FD involving the frontal, ethmoid, sphenoid bones, as well as a large intracranial mass in the anterior and middle cranial fossae, causing cerebral mass effect.

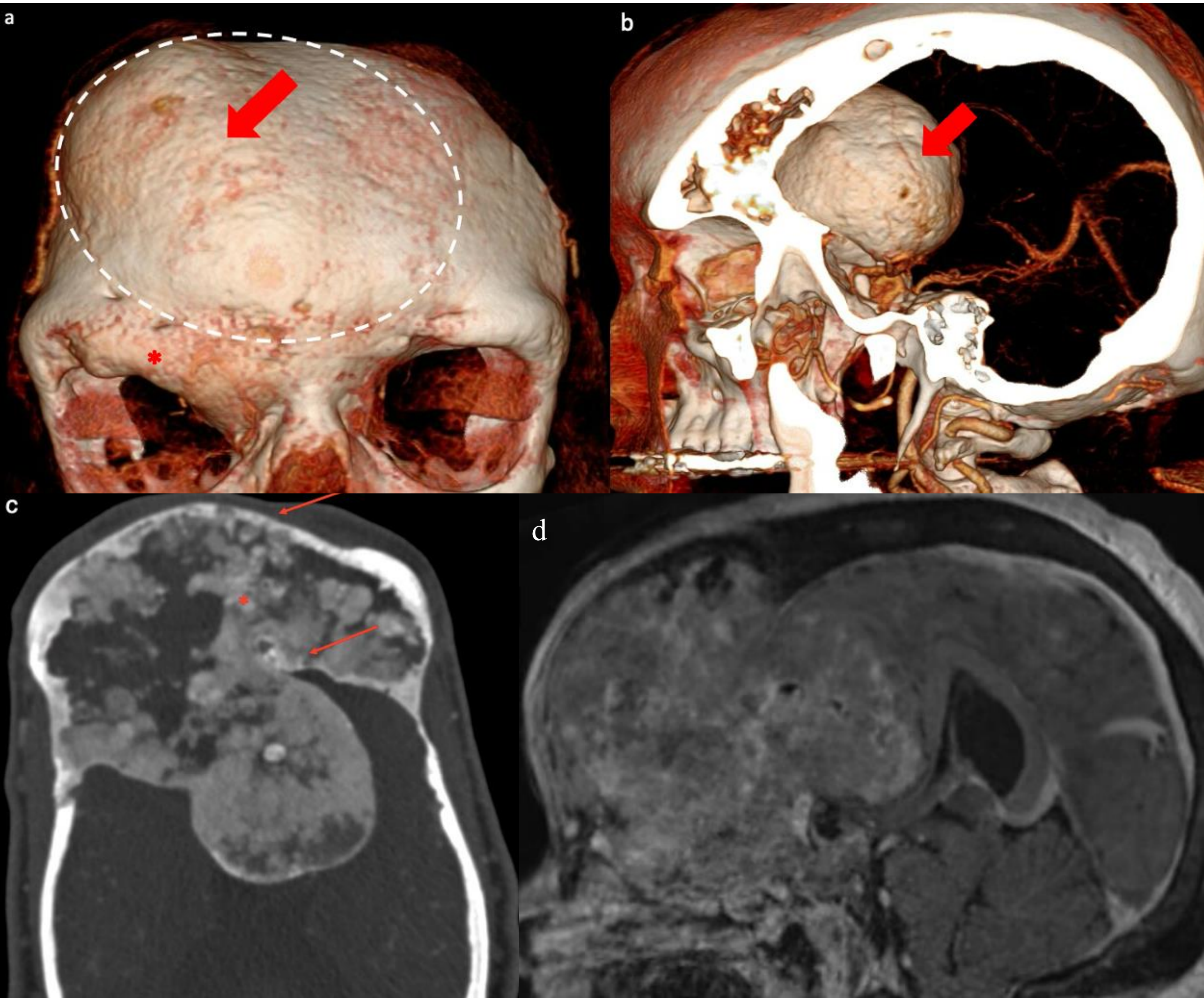


Figure 1. Preoperative images

We planned a 2-stage surgery with both demolitive and reconstructive purposes. During the first surgery, a bicoronal skin incision was made, and after mobilizing the scalp to the frontal prominence, two surgeons operated simultaneously on either side of the head to reduce surgical time. A bifrontal craniectomy was performed, providing access to the superficial lesion encased in the severely dilated frontal sinus. The lesion was carefully removed, and the right superior orbital prominence was recontoured to restore facial symmetry. Drilling of the pedunculated mass was initiated, remaining entirely intra-tumoral without exposing the dura.

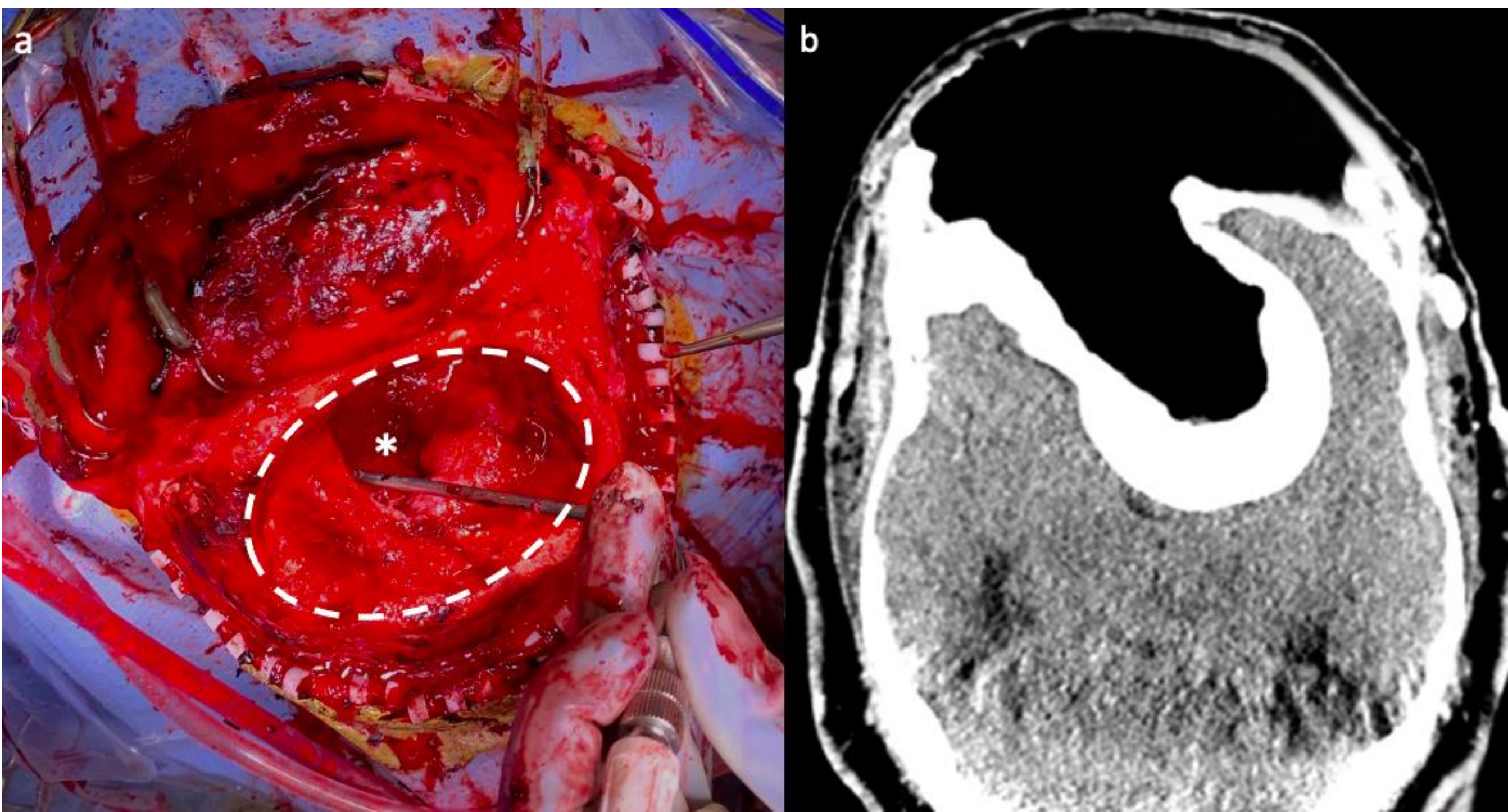


Figure 2. First surgery images

After the surgery, a fine-cut CT scan was performed to create a 3D custom-made polyetheretherketone (PEEK) cranioplasty. During the second surgery, the prior incision was reopened, tilting the flap forward to expose the bone around the craniectomy. We thinned the bone within the cavity and carefully lifted it. Under microscopic visualization, the resection in the deep portion, where the lesion was firmly attached to the dura and invaded the pia of the frontal lobes, was finalized. We reconstructed the anterior calvarium with the patient-specific implant.

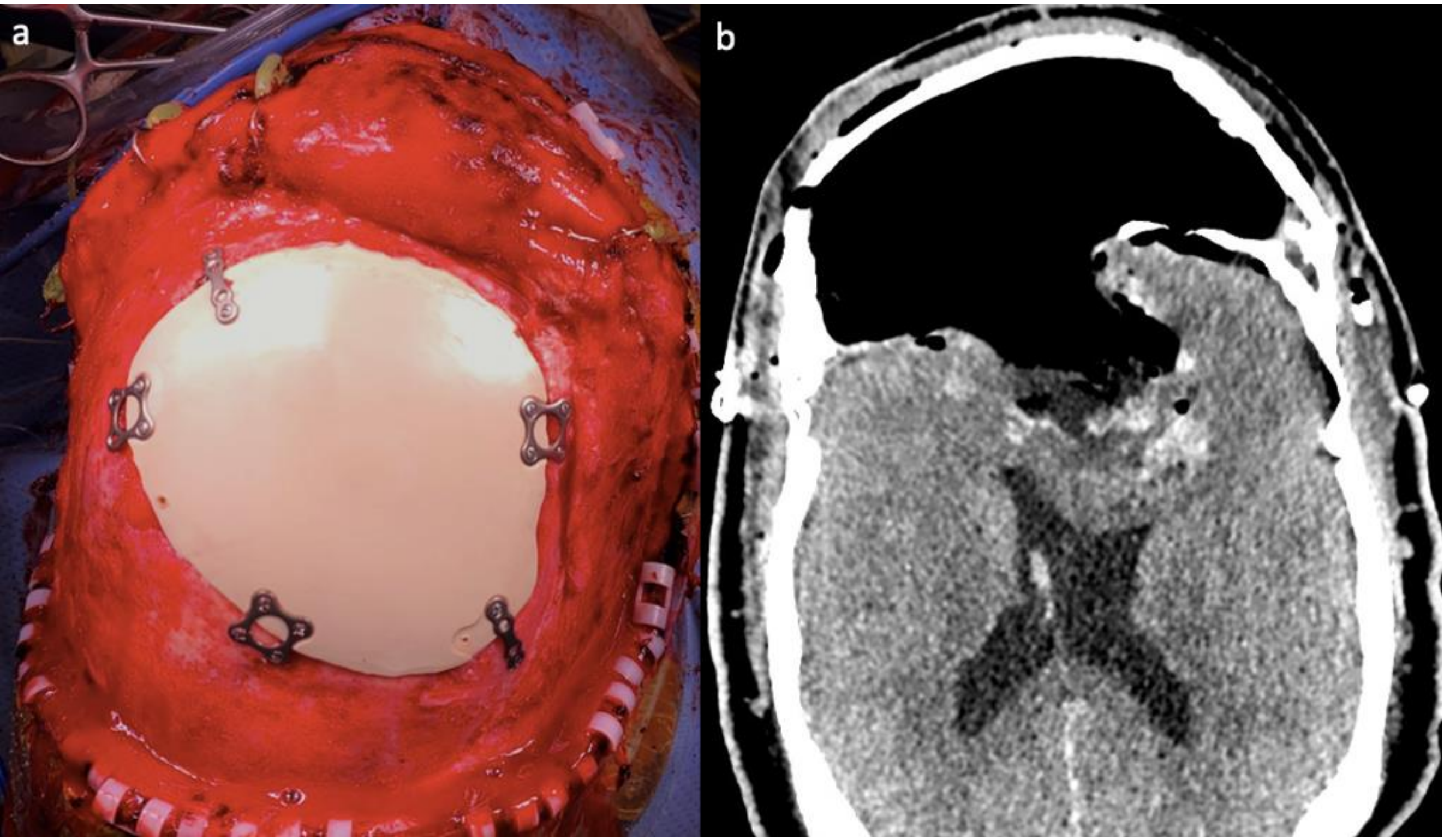


Figure 3. Second surgery images

## Conclusion

Two-staged surgical strategy successfully mitigated the risks associated with a single extensive surgery, particularly the potential for high blood loss and those inherent to the prolonged operative time. This approach also allows for planning a custom-made cranioplasty during the second stage, based on the extent of the lesion removal on the first intervention.