Repeat Intracranial Expansion After Skull Regrowth in Hyperostotic Disease: Technical Note

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■ OBJECTIVE AND IMPORTANCE: **Camurati-Engelmann disease (CED) is a rare, autosomal-dominant genetic disorder resulting in hyperostosis of the long bones and skull. Patients often develop cranial nerve dysfunction and increased intracranial pressure secondary to stenosis of nerve foramina and hyperostosis. Surgical decompression may provide symptomatic relief in select patients; however, a small number of reports document the recurrence of symptoms due to bony regrowth. We present a patient who had been treated previously with bilateral frontal and parietal craniotomy who experienced recurrence of symptoms due to reossification of her cranial bones. This report underscores the progressive nature of CED and its influence on surgical management. Furthermore, we propose a novel surgical approach with multiple craniectomies and titanium mesh cranioplasties that could potentially offer long-term symptomatic relief.**

■ CLINICAL PRESENTATION: **A 46-year-old female patient with CED who was treated with ventriculoperitoneal shunting, posterior fossa decompression, and multiple craniotomies 2 decades prior presented with signs and symptoms of increased intracranial pressure. Studies of the skull at presentation demonstrated rethickening of cranial bones that resulted in severely decreased intracranial volume.**

■ INTERVENTION: **A radical craniectomy, requiring 4 separate bone flaps made up of bilateral frontal and parietal bones, was performed. The remaining coronal and sagittal bony struts were drilled to approximately 1 cm thick. Cranioplasties with 4 separate titanium meshes were performed to preserve the natural contour of the patient's skull.**

■ CONCLUSIONS: **Although surgical decompression could provide some patients with CED symptomatic relief, clinicians should consider managing CED as a chronic condition. To the authors' knowledge, this is one of few case reports documenting the recurrence of symptoms in a patient with CED treated by surgical intervention. Furthermore, we propose that multiple craniectomies with titanium mesh cranioplasties confer more permanent symptomatic control, and, more importantly, lower the risk of recurrence secondary to cranial hyperostosis.**

INTRODUCTION

Camurati-Engelmann disease (CED), also known as progressive diaphyseal dysplasia, is a rare autosomal-dominant disease. It is caused by mutations in transforming growth factor-β1 (TGF-β1).[1,2] Under normal physiological conditions, TGF-β1 appears to stimulate bone formation and suppress bone resorption.[3-5] TGF- β1 mutations in CED, however, result in excessive activity of the growth factor.[6,1,2,7,8] It, therefore, causes progressive hyperostosis of the cranium and symmetrical sclerosis of long bones cortical diaphysis.[6,1,2,9] Presenting symptoms are highly variable, but they commonly include headache, visual and auditory disturbances, and musculoskeletal pain.[2,10] While the natural course of CED is unpredictable, it has been reported that up to 50% of patients experience headache and cranial nerve dysfunction secondary to decreased cranial vault volume and cranial

Key words
- Hyperostosis
- Intracranial expansion
- Skull Remodeling

Abbreviations and Acronyms
CED: Camurati-Engelmann disease
CT: Computed tomography
ICE: Internal cranial expansion
ICP: Intracranial pressure
TGF-β1: Transforming growth factor-β1

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foraminal stenosis, respectively.[2] Although there is no curative treatment for calvarial hyperostosis secondary to CED, it has been reported that internal cranial expansion (ICE), namely craniotomy with thinning of the inner calvarial table and cancellous bone, could increase intracranial volume to effectively lower ICP and provide symptomatic relief to medically refractory patients.[6,11] Although the recurrence rate of intracranial hypertension following decompressive procedures remains unclear, it is well documented that a subset of surgically treated patients experienced bony regrowth and recurrence of symptoms.[10,12,13] Here, we present a patient with CED who underwent ICE 2 decades prior with increased ICP secondary to rethickening of her bone flaps. Furthermore, we propose a novel surgical procedure, involving a 4-piece decompressive craniectomy with titanium mesh cranioplasties, that could potentially provide long-term symptomatic relief.

CASE PRESENTATION

The patient was a 46-year-old woman with known CED treated with ventriculoperitoneal shunting, posterior fossa decompression, and ICE 20 years previously. At presentation, she demonstrated signs and symptoms of increased ICP, including proptosis, headaches, nausea, and vomiting. Computed tomography (CT) imaging revealed significant reossification of her skull with compression of underlying brain (**Figure 1**).

Operative Approach

The patient was positioned supine with her head in neutral position on a horseshoe headrest. The patient's head was then fully shaved. A well-healed bicoronal zigzag scar from the patient's previous ICE was noted (**Figure 2**). The scalp was then prepared and draped in the standard neurosurgical fashion. An incision was made with a number 10 blade along the scar, and the scalp was reflected anteriorly and posteriorly to allow generous exposure of the patient's calvaria. Bifrontal and

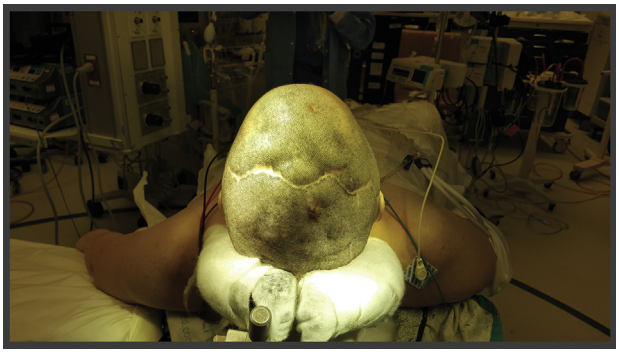

Figure 2. Previous surgical Scar. A zigzag, bicoronal scalp scar from the patient's previous internal cranial expansion was noted.

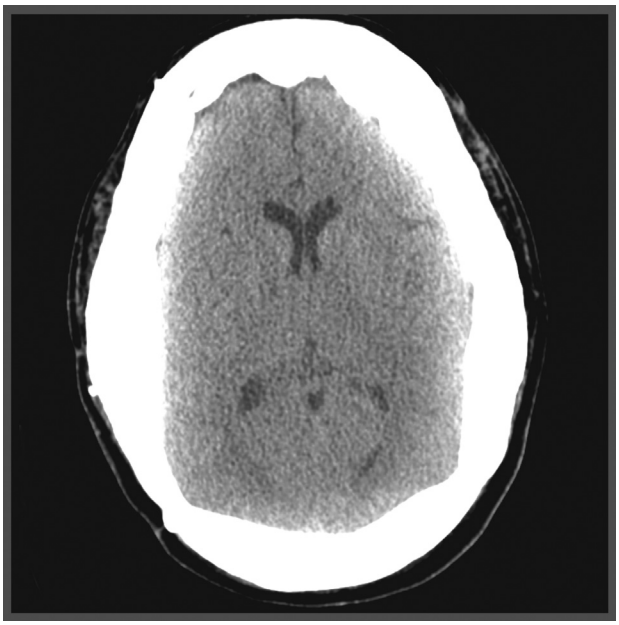

Figure 1. Preoperative computed tomography (CT) scan. Preoperative CT scan of the head showing diffuse hyperostosis and sclerosis of the calvarium. Effacement of underlying sulci was noted.

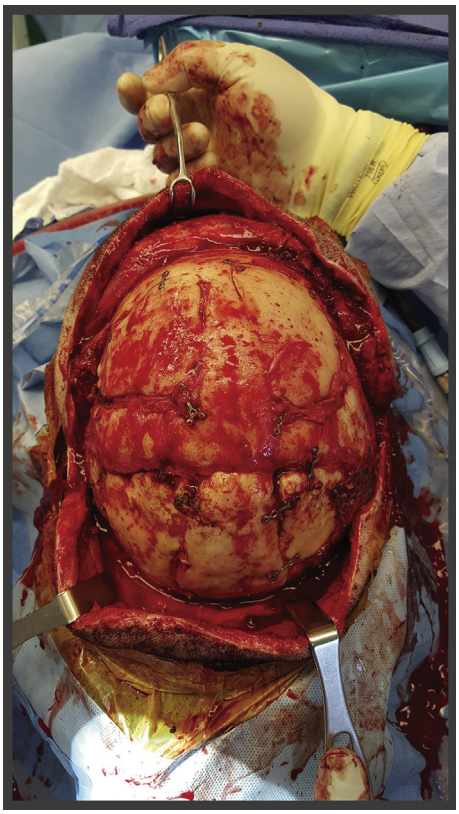

Figure 3. Operative Incision. An incision was made along the patient's scalp scar. Scalp was retracted anteriorly and posteriorly to expose the calvarium.