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*Case Report*

# Traumatic subgaleal hematoma in patient with Ehlers-Danlos syndrome: A rare case report

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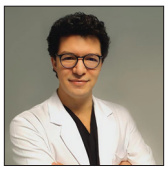

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## ABSTRACT

**Background:** A subgaleal hematoma (SGH) describes scalp bleeding in the potential space between the periosteum and the galea aponeurosis. This hematoma generally occurs after vacuum-assisted and forceps delivery, but may also be seen following head trauma. Despite its benign course, SGHs may complicate by life-threatening events.

**Case Description:** We report a case of a 10-year-old male with Ehlers-Danlos syndrome presenting with scalp swelling following minor head trauma. On examination, a small swelling was observed in the occipital region. During the follow up, as the volume of subgaleal hematoma was increasing, we performed needle aspiration to achieve volume reduction, and dressed with a cap like bandage that wrapped and compressed scalp. The patient was hospitalized due to hemodynamic instability and a blood transfusion was performed. Due to extended usage of compressive bandage, a large area of scalp tissue became necrotic. The necrotic scalp tissue was debrided and reconstructed by plastic and reconstructive surgery. After surgery, another hematoma formed extending from the front of the ear to the ipsilateral neck caused facial paralysis, this hematoma was evacuated and a drain was placed. The patient was followed up for 1 year and no recurrent cephalhematoma was observed.

**Conclusion:** Ehlers-Danlos is a rarely encountered connective tissue syndrome, this case underscores the importance for neurosurgery physicians to recognize the potential catastrophes, these patients may present with following even minor injury.

**Keywords:** Ehlers-Danlos syndrome, Pediatric neurosurgery, Subgaleal hematoma, Trauma

## INTRODUCTION

A subgaleal hematoma (SGH) is generally related to head trauma. The galea in a newborn can be pulled out in a vertical plane and slipped in a tangential direction when vulnerable to an external force as the scalp is tender, the subcutis and periosteum are sensitive, and there is a fragile connection between these tissues. In case of an injured vein, a subcutaneous hematoma may occur; therefore, the infant is sensitive to an SGH or subperiosteal hematoma. A few cases have been described in older children, occasionally as a result of minor head trauma such as hair braiding or hair pulling.[1-3,5,6,11] Despite its benign course, SGHs may complicate by life-threatening events.[1-3,5,6,9,11]

## CASE PRESENTATION

A 10-year-old boy was admitted to the Hacettepe University Department of Emergency following a minor head injury 1 h before arrival. He had fallen and striking the occipital region of his

head on the edge of the table. He presented to the emergency department with a chief complaint of minor scalp swelling. On initial presentation, the patient's vitals were within normal limits. He was awake and alert on admission. His neurological examination was normal excluding a minor swelling in the occipital region. His history also revealed joint hypermobility, fatigue, syncope, easy bruising, and hematoma formation resulting from minor trauma due to Ehlers-Danlos syndrome. The patient was consulted to our neurosurgery department within half an hour on admission. A stat computed tomography (CT) of the brain with the angiographic study was ordered. A CT showed no intracranial hemorrhage. The neurosurgery team decided to wrap a compressive bandage around his head in an attempt to stop the bleeding by compression. After around ½ h, the neurosurgery team was contacted again, only to be informed that the compressive bandage did not stop swelling. The scalp started to swell over the bandage. A CT has performed again. There was a massive increase in blood volume in subgaleal space [Figures 1a and b]. The neurosurgery team evacuated approximately 400 cc of blood with needle aspiration. The patient was still awake with a Glasgow Coma Score of 15 although, through the course of his admission, he slowly developed hemodynamic instability (MAP: 50 mmHg, HR: 140 bpm). The compressive bandage was revised and the whole scalp was compressed with a cap-like compressive bandage. The patient was hospitalized and he was kept under observation and support treatment with Vitamin K, intravenous immunoglobulin, and fresh frozen plasma. Due to extended usage of compressive bandage (day 10), a large area of scalp tissue extending from the occipital area to bilateral parietal bones became necrotic, the necrotic scalp tissue was debrided and reconstructed by plastic and reconstructive surgery. After surgery, another hematoma formed extending from the front of the ear to the ipsilateral neck causing facial palsy. A Jackson-Pratt drain was placed in combination with a pressure dressing after the evacuation of the hematoma was completed. The Jackson-Pratt drain was removed a week later. During this time, the facial palsy completely resolved. Twenty-nine days following admission, the patient was discharged. The patient was followed up for 2 years and is completely normal with no recurrence of cephalhematoma.

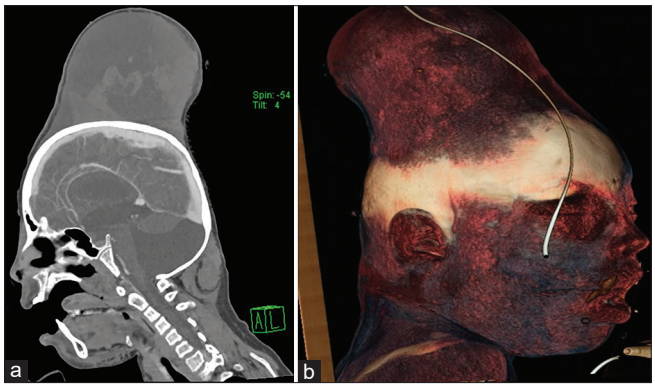


**Figure 1:** CT obtained after compressive bandage. (a) Sagittal view and (b) 3D reconstruction of emergency CT.

## DISCUSSION

Ehlers-Danlos syndrome is a collagen tissue disorder described by Job van Meekeren.[14] Ehlers-Danlos syndrome has been associated with perforated viscous, vascular catastrophes, ruptured uterus, pneumothorax, aneurysms, arteriovenous fistulas, intracranial aneurysms, and arterial dissections.[1-3,5-9,11,12,15] Due to the compromised nature of connective tissues in Ehlers-Danlos syndrome, even minor traumas may lead to catastrophic results.[1,2]

Anatomically, the subgaleal area binds the cranial periosteum and the galea aponeurotica. The subgaleal area includes loose connective tissue and minor emissary vessels connecting the extracranial venous system and the intracranial venous sinuses. SGH results from the impairment of these emissary veins. SGH expressed as a gathering of blood under the galea aponeurotica layer can be classified as nontraumatic or traumatic depending on the mechanism of the event. Traumatic SGH can be caused by minor head trauma such as hair pulling[1,2,5,11,12] whereas nontraumatic cases occur because of coagulopathies or rupture of arteries or arteriovenous fistulas.[8,9,12,15] A review of the literature revealed few reported cases of SGHs in patients with Ehlers-Danlos syndrome. A case review by Morel *et al*. describes a case very similar to ours, which occurs after minor trauma and increases in size with needle aspiration, the patient was treated with compressive bandages and blood transfusion.[11] In their case, angiography also did not show any vascular abnormalities, and the authors concluded that the underlying pathological mechanism led to a massive increase in hematoma; skin hyperelasticity, and a bleeding diathesis resulting from vascular and perivascular tissue fragility. In another case reported by Felton *et al*., a more invasive method of treatment was utilized and a drain was placed under the scalp, due to the fast, uncontrolled, and massive increase in the size of hematoma.[3] The patients were recovered in both cases.

In our case, the patient is 10 years old with a SGH in the occipital region. This is a rare case of traumatic SGH presenting in adolescence and this report describes a giant occipital SGH with protrusion and anemia in a 10-year-old boy for which we utilized a more conservative approach, which leads to compression necrosis and further complications. The clinical features of the patient were found to be compatible with the EDS Kosho type (EDSKT). The new Ehlers-Danlos syndrome, also known as EDSKT, is a connective tissue disorder that causes massive subcutaneous

hematomas in individuals.[4,10,13] Genetic testing is required for the diagnosis of EDKST. Unfortunately, the patient's family refused to allow us to do the test. The ideal treatment for a similar case remains debatable. Our patient's initial presentation resulting from an otherwise benign trauma highlights the importance of rapidly initiating aggressive treatment modalities when conservative measures fail with SGH in a patient with Ehlers-Danlos syndrome.

## CONCLUSION

Ehlers-Danlos syndrome is an uncommon connective tissue disorder. Our case underlines the importance for neurosurgeons to recognize the potential catastrophes of minor trauma in these patients.

## DECLARATIONS

### Statement of ethics

The authors have no ethical conflicts to disclose. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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