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Case Report

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Giant cephalhematoma following minor head injury in a patient with Ehlers-Danlos Syndrome: A case report and literature review

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Abstract

Ehlers-Danlos syndrome (EDS) is an extremely rare disease characterized by connective tissue fragility. It is characterized by laxity of ligaments, leading to joint hypermobility and hyper extensibility of the skin. The fragility of the vessels and hyper extensibility of the skin may complicate the development of large soft tissue hematomas after minor trauma, which may result in serious morbidity and mortality. We report a case of giant cephalhematoma in an 11--year-old boy with Ehlers-Danlos syndrome following a minor head injury. The cephalhematoma increased in size within hours and extended to involve the neck and upper chest wall, resulting in hypovolemic shock and cardiac arrest.

Keywords: cephalhaematoma, Ehler-Danlos syndrome, head injury, mortality

1. Introduction and importance

Ehlers-Danlos syndrome (EDS) is a rare connective tissue disease resulting from abnormal collagen production. The "classic" triad of hyper elasticity of the skin, joints hyper flexibility, and fragility of the skin and vessels are the common features of this pathology (1). This may lead to the development of large hematomas after minor trauma, which may result in life-threatening conditions (2). Therefore, emergency physicians should focus on the identification of life-threatening manifestations related to the great vessels, such as aneurysms, great vessel ruptures, and dissection. The systemic nature of the disease, however, can lead to vessel wall fragility in areas outside of the great vessels, resulting in ecchymosis and hematoma formation following minor trauma (3). In this study, we present a case of a large scalp hematoma in a patient with EDS following a minor head injury, resulting in hypovolemic shock and cardiac arrest.

2. Case presentation

An 11-year-old boy, known case of EDS, was rushed to the surgical ER with his mother, as he had fallen and struck the occipital region of his head on the floor. The patient developed a rapidly enlarging occipital swelling after the incident. The mother reported no loss of consciousness, fits, or vomiting. The patient has a previous history of multiple hematomas after minor traumas in the knee and buttocks.

The initial assessment in ER revealed a fully conscious state and a huge scalp swelling over the occipital and left parietal areas (**Fig. 1**). Airways were patent with good air entry bilaterally, the abdomen was soft and lax, the Glasgow coma scale was 15/15, pupils equally reacted to light with no spine tenderness nor limitation in movement. There was no neurological deficit.

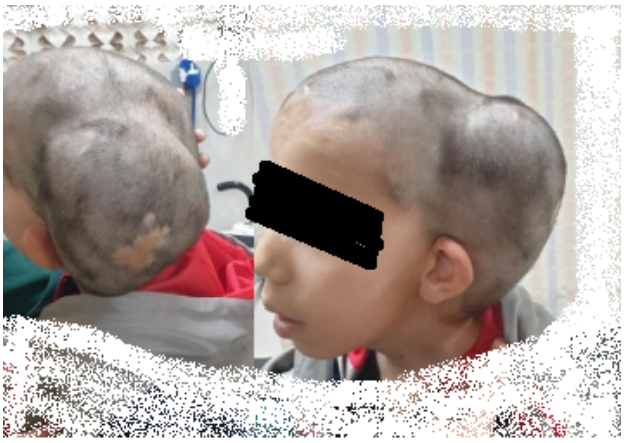

Fig. 1. Huge subgaleal hematoma after trivial head trauma

Vital signs were normal (BP: 110/60 mmHg, HR: 96 beat /min, RR: 16 /min). The laboratory tests of hemoglobin (HB), platelets, prothrombin time, and partial thromboplastin time were 11.9 g/dl, 623 *109/l, 13.4 seconds, and 24.6 seconds; respectively. A Brain CT scan showed a huge scalp hematoma measuring about (11 x 5) cm with an underlying linear skull fracture (**Fig. 2**).

Upon admission, the hematoma rapidly enlarged in size with extension to the left postauricular area, and the patient

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became lethargic with a blood pressure of 110/70 mmHg and a pulse rate of 100 beats/min. Follow-up HB after three hours showed a drop to 9.3 g/dl.

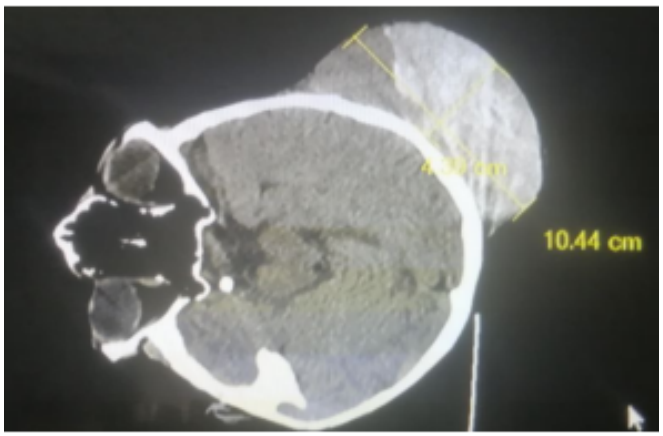

Fig. 2. Initial brain CT scan showed huge subgaleal hematoma

We compressed the hematoma using a crepe bandage after marking the hematoma. Blood transfusion started with 150cc Packed red blood cells (PRBCs), with strict monitoring for vitals and urine output in a high-dependency unit. Unfortunately, the hematoma continued to enlarge and extended downward to the whole left side of the neck and upper part of the chest, and the skin appeared ecchymotic (**Fig. 3**).

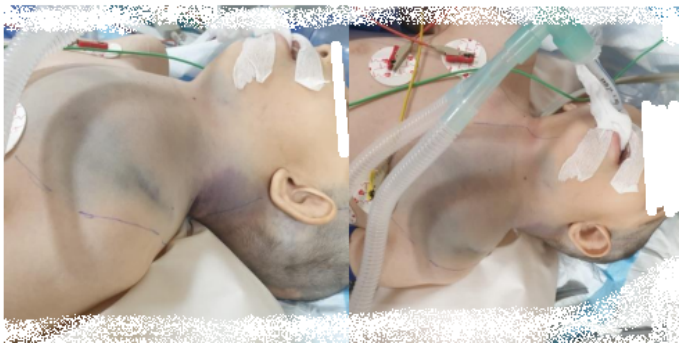

Fig. 3. extension of the hematoma to the neck and upper chest

Hb dropped to 6.2 g/dl within two hours, and the transfusion rate increased with another 250cc PRBCs and two fresh frozen plasma units.

The neurosurgeon, otolaryngologist, anesthetist, pediatric hematologist, and general surgeon on-call teams decided to electively intubate the patient for the risk of airway compression and prepare him for hematoma evacuation with hemostasis.

Unfortunately, the patient developed sudden bradycardia and hypotension; CPR started according to the protocol and was led by an anesthetist, with a simultaneous aspirating of more than 400cc from the hematoma existing the neck using two 18G cannulas. Because of the development of sudden hypotension and bradycardia, the clinical diagnosis of cardiac tamponade was considered and pericardiocentesis was done with an aspiration of 40cc of blood. The CPR lasted about 45 minutes with no response, and the patient was deceased six hours later.

3. Clinical discussion

Ehler-Danlos syndrome is a rare hereditary connective tissue disease caused by abnormal collagen products. Ecchymosis and hematoma formation following minor trauma is frequently seen in these patients but some of these hematomas can develop into life-threatening conditions as in the case we are presenting. In our case, the cephalohematoma rapidly enlarged and then extended to the neck and into the chest of the patient within a few hours causing fatal results

Initially, the decision was to manage this hematoma conservatively because of the patient's good and stable clinical condition, and all the previous traumatic hematomas in the patient's clinical history were treated conservatively without complications. Unfortunately, the rapid deterioration in the clinical scenario of this case did not allow us to interfere with any surgical procedure to evacuate the hematoma and secure the patient especially when the hematoma extended to the neck and upper chest. As we did not expect this rapid deterioration and expansion of the hematoma we were late for the surgical intervention that might result in a better outcome.

After reviewing the literature, we found three similar cases with good prognoses unlike ours. The summary of these cases is presented in **Table 1**.

Table 1. Summary of the studies that described EDS patients presented with sublegal hematoma

Study ID	Age (years)	Gender	Medical treatment	Previous related history	Management	Prognosis
Gurses et al. (2022), (4)	10	Male	NR	NR	1. Blood evacuation with needle aspiration 2. Compress a skull using a cap-like compressive bandage 3. The patient was given intravenous immunoglobulin and Vitamin K	Alive
Felton et al. (2013), (3)	41	Female	NR	Hematoma formation in chest wall and extremities and easy bruising	1. Factor IX complex was given to the patient 2. Compress directly on the scalp and face 3. Hematoma was evacuated	Alive
Morel et al. (1998), (5)	17	Male	NR	The patient was admitted with similar symptoms one year ago	1. Blood evacuation had been done 2. Pressure was applied on the skull	Alive

4. Conclusion

We conclude that physicians should be aware of the importance of aggressive treatment and intervention of these hematomas as they can rapidly enlarge causing fatal results. Hematoma compression, massive blood transfusion, intubation, and early surgical intervention should be one of the treatment modalities considered first in rapidly progressive hematomas in EDS.

Informed Consent

A written informed consent was obtained from the family for publication of this case report and accompanying images

Conflict of interest

All authors have no conflicts of interest to declare.

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Authors' contributions

Concept: E.B., M.A., S.A., Design: E.B., M.A., S.A., Data Collection or Processing: E.B., M.A., S.A., Analysis or Interpretation: E.B., M.A., S.A., Literature Review: E.B., M.A., S.A., Drafting: E.B., M.A., S.A.

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