

Unexplained Facial Scar: Child Abuse or Ehlers-Danlos Syndrome?

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Abstract

Context: Child abuse is a serious problem, and its physical manifestations can be mimicked by certain diseases and conditions. These conditions can include genetic, congenital and other disorders that may result in poor weight gain, bone fractures or skin lesions that look like bruises or burns. **Case Report:** This paper reports the case of a seven-year-old girl with Ehlers-Danlos Syndrome (EDS), which was misdiagnosed as child abuse. This child was referred to us for treatment of an unexplained facial scar that was alleged to be the result of child abuse. **Conclusion:** When unusual skin presentations are observed, dermatologists should consider the possibility of child abuse to protect the child. Furthermore, they should be aware of the cutaneous abnormalities that mimic injuries associated with abuse to avoid the unnecessary reporting of child abuse.

Keywords: Child abuse, Ehlers-danlos syndrome, Facial scar

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Introduction

Child abuse is a public health issue that has significant social and family impacts.^[1] Although child abuse is a real problem, many physical illnesses cause symptoms that may have the appearance of child abuse. There are several medical conditions that mimic the physical manifestations of child abuse. These conditions include genetic, congenital and other disorders that may result in the bone fractures or skin lesions that look like bruises or burns.^[2] Although child abuse must always be suspected when the child has unusual marks or injuries that do not match the given history,^[3] but it is necessary to distinguish such abuse from the results of certain diseases.^[4]

The skin is the first organ that is affected by physical aggression, and it shows the most frequently recognizable forms of abuse.^[5-7]

About 90% of the victims of physical abuse show skin lesions on examination.^[8] These lesions include bruises, lacerations, abrasions, human bites, scars and burns, which mimic abusive injuries.^[6]

This paper reports the case of a seven-year-old girl with Ehlers-Danlos Syndrome (EDS), which was mistaken as a case of the child abuse. There are no high-quality studies that have examined the differences between underlying skin diseases and child abuse. We discuss briefly how to distinguish between the physical results of actual child abuse and the results of diseases that mimic the results of child abuse.

Case Presentation

A seven-year-old girl, who had bruises and scars predominantly all over her face [Figure 1], lower extremities and on her body was referred by a forensic expert. She lived with her mother, and her father had

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10.4103/1947-2714.145482

passed away. The child was evaluated in the pediatric clinic and child abuse was suspected as the cause of the bruises and scars, and hence the child was admitted to the pediatric ward for evaluation.

The mother was significantly upset by the suggestion that child abuse may have occurred and refused to allow her daughter to remain in the clinic for observation. The child was born by normal vaginal delivery and there were no complications associated with her birth. On examination, the child appeared normal.

Dermatologic examinations revealed that the child had very smooth, lax skin, but she had wide, atrophic, cigarette paper-like scars on both shins. Other clinical findings included hyperextensible skin [Figure 2], increased range of motion of the joints, long fingers, toes that bruised easily and fragile skin that had a brownish discoloration [Figure 3]. There were hyper mobile joints with the score of 6/9 on the Beighton scale.

There were no joint dislocations. She had no major health concerns. Other investigations, such as complete blood count and routine examination of the stool proved normal, and the urine and serum levels of sodium, potassium, urea and creatinine were within the normal limits. Reviewing her family history indicated hyperextensible joints and skin in her father's family. Based on the characteristics of the clinical findings and the positive family history, she was thought to have classic EDS, type I or II. Thus, the EDS were included in the differential diagnosis to avoid possible harm to the family that could result from an allegation of abuse. The child was returned to her mother after the correct diagnosis.

Discussion

This paper reported the case of a seven-year-old girl with EDS, which was misdiagnosed as child abuse. Violence against children tends to be an uncomfortable topic for many doctors, in part due to the lack of training in recognizing and dealing with the problem. In general, the reasons for this uncomfortable topic are:

1. Little guidance or no training in recognizing the problem and
2. Not understanding its true dimensions.^[4]

Although, the injury patterns in an abusive situation against the accidental trauma have been described in the literature, there is a paucity of information that differentiates the effects of EDS from child abuse.^[9,10] When a common illness mimics child abuse, it is typically diagnosed, but this often is not the case for rare diseases. The physicians often misdiagnose child abuse with other rare diseases. The morbidity and mortality of a missed diagnosis of child abuse have been well documented.^[11]



Figure 1: Typical features of Ehlers-Danlos Syndrome (wide atrophic facial scars)



Figure 2: Cutaneous hyperextensibility: Cutaneous hyperextensibility is one of the cardinal features of Ehlers-Danlos Syndrome

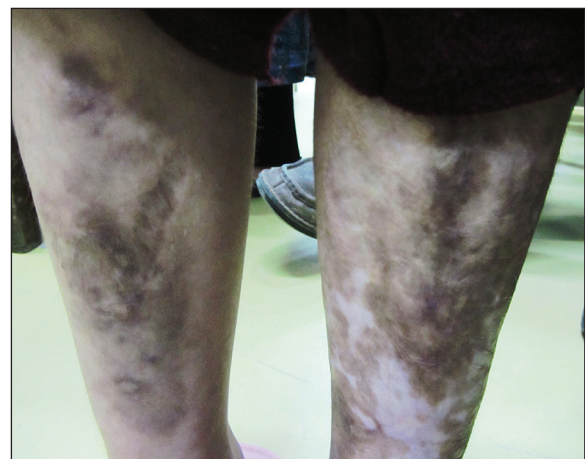


Figure 3: Easy bruising, characteristic brownish discoloration of the skin, especially in exposed areas, such as the shins

When child abuse is misdiagnosed, there are emotional, psychological, and financial burdens on both the family and the child. Hence, knowing of the

Table 1: Conditions that mimic child abuse

Condition	Comments
Bleeding into the skin (ecchymoses, purpura)	Areas most commonly traumatized by children (e.g. shins, knees) Common sites of trauma, especially shins Primarily children from SEAsia Favors lower extremities as well as the buttocks in HSP and the face in acute hemorrhagic edema; associated edema and arthritis
Multiple bruises due to platelet or clotting disorders	
Ehlers-Danlossyndrome	
Bruising due to "cupping" or caogio (coin rubbing)	
Vasculitis, particularly Henoch-Schbnleinpurpura (HSP) and acute hemorrhagic edema of infancy	Lack of progression of color to green or yellow
Blue discoloration of skin mistaken as bruising	
Dermal melanocytosis	
Infantile hemangioma (deep) or vascular malformation	

cutaneous manifestations of child abuse is critical to the accurate identification and reporting of injuries that result from abuse.^[6] Physicians, especially dermatologists and pediatrician, should know the signs of the physical abuse and the differential diagnoses (skin and/or systemic diseases) that mimic or simulate the main skin lesions resulting from physical violence to avoid mistaken diagnoses. This is vitally important because mistakenly accusing parents of child abuse is devastating, and it is highly unlikely that an atmosphere of trust can ever be re-established. It is difficult to diagnose conditions that have been mistaken for the parental maltreatment, and when the diagnosis is incorrect, there are devastating consequences for the families.^[12]

Table 1 shows some of the signs and symptoms of the medical conditions that have been mistaken for the abuse [Table 1].^[13] EDS is one of the conditions that may be mistaken for child abuse due to the ecchymosis and scars.^[14]

EDS belongs to a heterogeneous group of collagen disorders that are characterized by the hyperextensibility and fragility of the skin, with easy bruising and hypermobility of the joints.^[14,15]

It is a rare genetic disorder that affects only 1 in 5000 people, and it is recognized frequently by pediatricians and dermatologists.^[14] This illness can easily be misdiagnosed because the cutaneous findings mimic traumatic injuries.^[6]

The additional findings in EDS, such as fragile skin that bruises or tears easily, poor and slow healing of wounds, and widened atrophic scars with the formation of cigarette paper-like scars (papyraceous) healing, easily can lead to a diagnosis of child abuse.^[13] In EDS, connective tissue disorders and the weakness of blood vessels lead to bleeding. An example in which an erroneous diagnosis of Shaken Baby Syndrome was made initially was described by Drs. S. M. Omen and D. L. Roberts.^[9,10]

The fragile skin is manifested by the splitting of the dermis following relatively minor trauma, especially over pressure points (knees, elbows) and areas prone to trauma (shins, forehead, and chin). The fragility of the skin may cause dehiscence of the sutured incisions in the skin or mucosa.^[16]

Easy bruising is a common finding and manifests as spontaneous ecchymoses, frequently recurring in the same areas and causing a characteristic brownish discoloration of the skin, especially in the exposed areas such as the shins and knees. There is a tendency toward prolonged bleeding (e.g., after brushing of the teeth) in spite of a normal coagulation status.^[16] Thus, EDS should be included in the list of childhood diseases that can be misinterpreted as the child abuse. The physician should consider that error or omission may cost the lives of children and undue child abuse charges could cost the reputation of an innocent adult.^[17]

Conclusion

Dermatologist should consider the diagnosis of child abuse in the presence of unusual skin presentations to protect the child. Furthermore, they should be aware of the cutaneous abnormalities that mimic abusive injuries to avoid the unnecessary reporting of child abuse.

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How to cite this article: Abtahi-Naeini B, Shapouri J, Masjedi M, Saffaei A, Pourazizi M. Unexplained facial scar: Child abuse or Ehlers-Danlos syndrome?. *North Am J Med Sci* 2014;6:595-8.

Source of Support: Department of Dermatology, Skin Diseases and Leishmaniasis Research Center, Isfahan University of Medical Sciences, Isfahan, Iran. **Conflict of Interest:** None declared.