Our Dermatology Online

Continuing Medical Education

Dermatologic Emergencies CME Part V: Abuse-Related skin manifestations, dermatologic surgical emergency and other rare miscellaneous emergencies

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ABSTRACT

The last part of this continual medical education series reviews the cutaneous manifestations of physical and sexual abuse and their differential diagnosis, dermatologic surgical emergencies and miscellaneous dermatologic emergencies such as acute graft-versus-host disease, hypereosinophilic syndromes and vascular Ehlers-Danlos syndrome

Key words:*Physical abuse; Sexual abuse; Dermatologic surgeries; Acute graft-versus-host disease; Hypereosinophilic Syndromes*

Abuse can take many different forms, including physical and emotional abuse. Child abuse, elder abuse, and domestic violence are common and affect patients of all socioeconomic classes and races. The diagnosis of physical abuse requires a high level of suspicion. A thorough history and physical examination are the cornerstones of diagnosis.

ABUSE-RELATED SKIN MANIFESTATIONS

Over 55 million children in the WHO European Region are affected by child maltreatment. A high prevalence of child abuse was documented in the 2013 European report on the prevention of child abuse, from 9.6 percent for sexual abuse, 16.3% for physical neglect, 18.4% for emotional neglect, 22.9% for physical abuse, and 29.6% for emotional abuse [1].

Cutaneous manifestations of physical abuse may include unexplained bruises, lacerations and abrasions, bite marks, curvilinear marks, burns and traumatic alopecia [2]. Signs of sexual abuse may include attenuation, fresh tears, or scars of the hymen and the anal margin extending out onto the perianal skin (Fig. 1).

Treatment

The diagnosis of physical abuse requires a high index of suspicion. A thorough history and physical examination are the mainstays of diagnosis [3]. Dermatologists may be the first physicians to encounter the abused patient, so it is important to be alert for signs of abuse, perform the appropriate documentation and testing, and report it to the proper authorities [4].

DERMATOLOGIC SURGICAL EMERGENCY

Even for healthy patients, surgical procedures include some risk and can result in unpredictable complications. Although uncommon, recognition of dermatologic surgical emergencies while operating in outpatient setting is vital to avoid undesirable outcomes (Table 1).

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Practical pearl

- Early diagnosis of abuse is crucial; as there is a high chance of recurrent abuse resulting in serious injury or even death.
- It is essential to speak with the child away from the caregivers if the child is verbal using simple, understandable, open-ended questions as appropriate history taking is the gold standard for diagnosis besides the physical signs and social risk factors.
- In abused children, bruises are the most common form of injury. Bruises at the protected areas of the body that are away from bony prominences and in different healing stages should raise the suspicion of child abuse.
- Although it usually raises the possibility of sexual abuse, the presence of anogenital warts in children alone is not diagnostic of sexual abuse without supporting clinical and social history.
- The importance of psychological support to abuse victims cannot be overemphasized. Physical scars of abuse can ultimately heal, but the emotional and psychological sequelae can persist and become more disastrous.

OTHER RARE MISCELLANEOUS EMERGENCIES

• Acute Graft-versus-host disease

Acute Graft-versus-host disease (GvHD) is caused by an immunological reaction between **donor** T lymphoid cells and the host tissue. Around 35 to 50 percent of recipients of hematopoietic stem cell transplant (HSCT) will experience acute GVHD typically before day 100 after the HSCT. The risk of aGvHD depends on the patient's age, source of the stem cell, prophylaxis and conditioning methods [24].

Skin involvement is often the first indicator of acute GVHD (81%), followed by gastrointestinal (54%) and liver disease (50%). Skin manifestations range from a mild, asymptomatic morbilliform eruption to erythroderma with TEN-like bullae and desquamation (Table 2). Gastrointestinal involvement is usually the most severe and challenging to treat. It manifests as abdominal pain, nausea/vomiting, and secretory diarrhea. Hepatic disease is characterized by jaundice, elevated total bilirubin, and alkaline phosphatase levels [25]. Diagnosis is usually based on clinical features after the exclusion of similar conditions [26]. Skin biopsy, upper and lower GI endoscopy, and cholestatic pattern liver function derangement may aid in the diagnosis [27-29].

Treatment

patients with mild (grade I) skin involvement without significant hepatic or GI symptoms usually respond to high-potency topical steroids. More severe disease (stage II-IV) is treated with systemic steroids (Fig. 2). Other modalities for steroid-unresponsive aGvHD include mycophenolate mofetil, extracorporeal photopheresis, biologics, pentostatin, methotrexate, and mesenchymal stem cells- [30].

Practical pearls

- Viral exanthems, engraftment syndrome and chemotherapy-related skin eruptions should be considered in the differential diagnosis of aGvHD
- Although it can involve any part of the body, the aGvHD rash usually starts on palms and soles and involves the facial area.
- Diarrhea associated with hyperbilirubinemia and skin rash involving the face, palms, and soles should raise the possibility of aGvHD.
- Intestinal involvement is usually patchy and normal endoscopy does not exclude the diagnosis of aGvHD [31]
- Plasma levels of elafin may have useful diagnostic and prognostic values [32]

• Hypereosinophilic Syndromes (HES)

Hypereosinophilic Syndrome is a rare heterogeneous group of disorders characterized by significant eosinophilia (>1500 eosinophils/mm^3 on at least two separate determinations or evidence of prominent tissue eosinophilia) associated with signs and symptoms of eosinophil-related end-organ damage after exclusion of secondary causes of eosinophilia, such as parasitic infections, allergic reactions, drug or chemical-induced eosinophilia, and neoplasms [34].

Common skin manifestations of HES include pruritic eczematous lesions, urticaria with or without angioedema, mucosal (oral and genital) ulcers, vasculitis, and erythroderma [35,36]. The two major clinical subtypes of HES are myeloproliferative and lymphocytic-variant HES. Myeloproliferative HES

Table 1: The most common encountered emergencies while performing dermatologic surgeries

Complication	Presentation	Management
Hemorrhage	Even the most qualified surgeons will experience bleeding at some point in their practice. Postoperative bleeding can result in hematoma formation with subsequent infection or wound dehiscence.	Measures that can help decrease the risk of intraoperative hemorrhage include: proper positioning of the patient, addition of epinephrine to local anesthetics, undermining of the surgical defect. Management of undergoing bleeding includes manual pressure, topical styptics/hemostatic agents, electro-cautery/surgery [6]. The previous approach to stop anti-coagulant/platelets before minor operations is no longer supported; as the risk of life-threatening thrombosis exceeds the minor risk of bleeding [7,8].
Air embolism	Air embolism can present as a complication of foam sclerotherapy and presents with clinical manifestations ranging from asymptomatic to pulmonary or neurologic events. Factors that may create an increased risk for the symptomatic event include right-to-left cardiac shunt like a patent foramen ovale, sclerotherapy foam characteristics, in addition to the sitting position, and Low central venous pressure [9]. Despite being typically safe, air embolism has been described after Mohs surgical technique on the scalp [10].	Regardless of air embolism etiology, the treatment aims to seal the potential site of air entry into the systemic circulation, supportive care, rapid restore flow to the cardiopulmonary circulation, and to promote the reabsorption of intravascular air [11].
Hypertensive crisis	Acute hypertensive crisis is usually defined as systolic blood pressure <180 mm Hg and/or diastolic blood pressure <120 mm Hg or a sudden increase in systolic blood pressure of 30% or more, with impending or progressive organ damage [12,13]. The clinical manifestations of hypertensive emergency are directly related to the particular end-organ dysfunction, including encephalopathy, aortic dissection, myocardial infarction, pulmonary edema, eclampsia, renal failure, ischemic stroke, or bleeding [14].	Although the dermatologic surgeons will not treat the hypertension crisis, awareness of the symptoms is essential to quickly mobilize the patient to the local emergency medical services to avoid further end- organ damage and control the blood pressure as soon as possible [15]. Notably, the amount of epinephrine used in local anesthetics during dermatologic surgery is too little to cause a significant elevation of blood pressure [16].
Vascular occlusion	Vascular occlusion can occur during dermatologic surgical procedures including, tamponade caused by soft tissue augmentation material, inadvertent injection of injectable filler or autologous fat directly into a vessel lumen, or intraarterial injection of sclerosant during sclerotherapy. Typical Symptoms of impending occlusion and necrosis include blanching, white or bluish discoloration, livedo reticularis, slow capillary refill with or without pain that manifests seconds to hours after injection [17,18].	The injection should be stopped if the vascular occlusion is identified during the injection session, and every single attempt should be made to aspirate or extract the product. Aggressive massage and warm compresses will help to improve blood flow and dissipate the agent [19]. Due to its vasodilator effect, the nitroglycerin paste application may help improve the flow on small-caliber arterioles [20]. Perilesional hyaluronidase injection is a highly effective option to alleviate the problem if the filler is HA derivate [21]. Other strategies that may be beneficial include a course of aspirin or heparin and hyperbaric oxygen [22,23].

Table 2: Grading of acute graft versus host disease [33]

Stage	Skin	Liver	Gut	Grade	Histologic
1	Erythematous macules and papules, <25% BSA	Bilirubin 2 to <3 mg/dl	Diarrhea, 500–1000 ml/day, or persistent nausea	I	Focal vacuolar change of basal keratinocytes
2	Erythematous macules and papules, 25–50% BSA	Bilirubin 3-6 mg/dl	Diarrhea, 1000–1500 ml/day	II	Grade 1 plus necrotic keratinocytes in the epidermis and/or hair follicle and a dermal lymphocytic infiltrate
3	Erythematous macules and papules (>50% BSA) to generalized erythroderma	Bilirubin 6-15 mg/dl	Diarrhea, >1500 ml/day	III	Grade 2 plus fusion of basilar vacuoles to form clefts and microvesicles
4	Generalized erythroderma with bulla formation	Bilirubin>15 mg/dl	Severe abdominal pain with or without ileus	IV	Grade 3 plus large areas of separation of epidermis from dermis

(M-HES) is usually associated with the tyrosine kinase fusion gene FIP1-like 1/Platelet-derived growth factor receptor (PDGFR) or other molecular mutations associated with eosinophil clonality. It is characterized by splenomegaly, thrombocytopenia, anemia, severely debilitating mucosal ulcers, and endomyocardial disease [37,38].

In lymphocytic-variant HES (L-HES), eosinophilia results from T-cell clones producing eosinophilopoietic cytokines such as IL-5. Compared to M-HES, this variant is more organ-restricted with a more protracted course and lower tissue fibrosis incidence [39].

The diagnosis of HES is based mainly on clinical features with the exclusion of other causes of eosinophilia.

Treatment

Corticosteroids are the first-line therapy for most patients with HES [40]. a tyrosine kinase inhibitor,

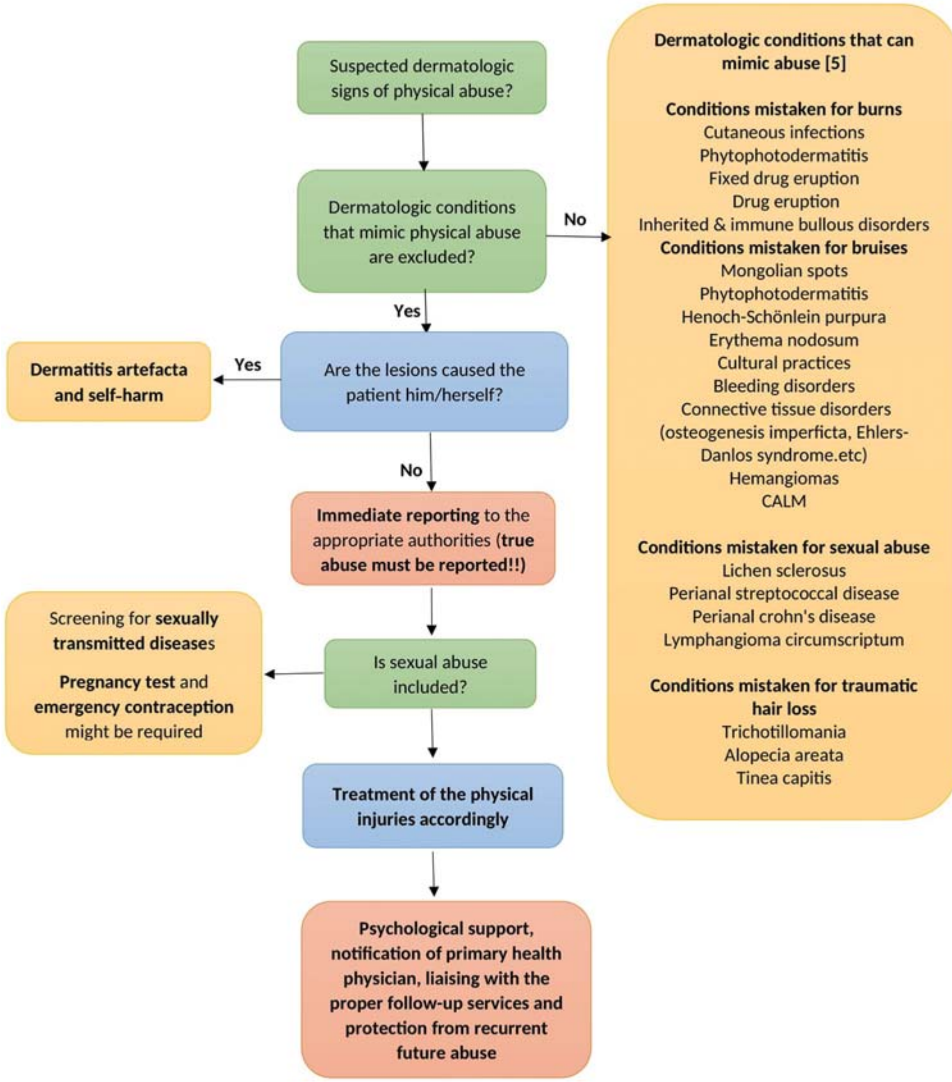

Figure 1: Abuse-related skin manifestations Dermatologic conditions that can mimic abuse should be excluded before making the diagnosis of physical/sexual abuse

Imatinib mesylate, is FDA approved for treatment of HES M-HES with a dramatic, quick response [41,42].

• Vascular Ehlers-Danlos syndrome (vEDS)

Vascular Ehlers-Danlos syndrome is a rare inherited connective tissue disorder caused by mutated type III collagen. The condition is characterized by thin translucent skin with extensive bruising and hypermobility of the small joints. Patients with vEDS usually have a characteristic facial appearance, including pinched nose, thin lips, micrognathia, hollow cheeks, and prominent eyes due to decreased periorbital adipose tissue [44,45]. vEDS is associated with an increased risk of spontaneous arterial aneurysms, dissection, or rupture involving the medium-sized arteries, intestinal and uterine perforations, pneumothorax, and sudden death [46,47].

Practical pearls

- unless the treatment is begun early before fibrosis, cardiac involvement is irreversible
- The endomyocardial disease may worsen during the first several days of imatinib treatment, so troponin levels should be monitored!!
- Steroids should be used with imatinib in M-HES patients with evidence of myocarditis to prevent myocardial necrosis [43]
- Despite being a steroid-responsive with less organ involvement, the L-HES can progress to lymphoma, especially those with CD4+CD3- lymphocytic Populations
- Strongloidosis parasitic infection should be excluded before initiating systemic steroid therapy

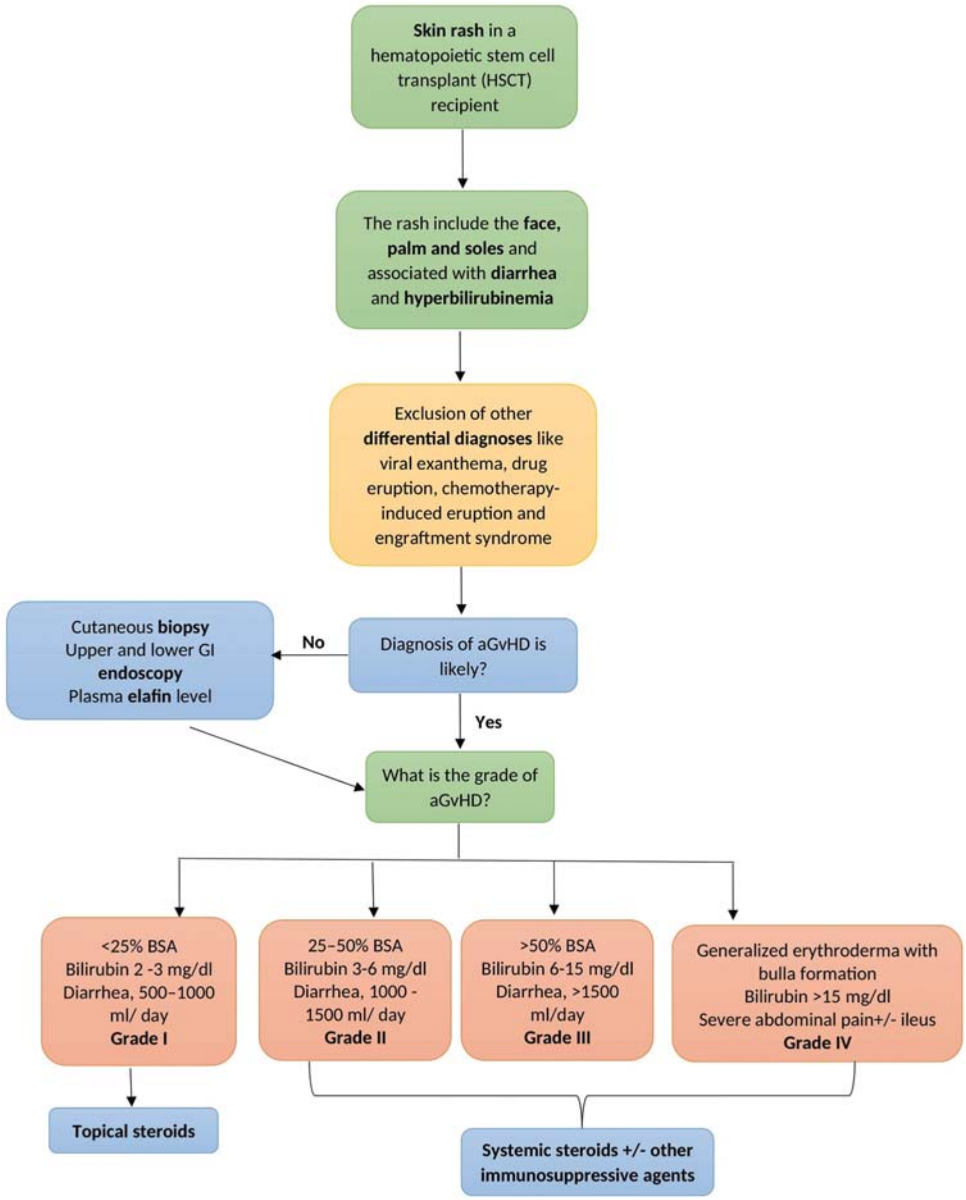

Figure 2: Graft-versus-host disease Management of acute Graft-versus-host disease according to its severity

Practical pearls

- vEDS is inherited in an autosomal dominant manner; hence genetic counseling is essential in most patients. De novo mutations and sporadic cases account for about half of the reported cases
- Patients with vEDS should avoid contact sports, medications that impair platelet function, and invasive procedures like arteriography and endoscopy unless necessary
- The patients should be provided with an emergency care card or medical alert document (a "vEDS passport") to explain the condition to the ER team

Diagnosis can be made using a combination of clinical features and molecular genetic testing to identify the mutated gene.

Treatment

There are currently no specific treatments for vEDS; the aim is to alleviate the symptoms, prevent fatal complications, and provide genetic counseling [48,49].

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