An Atypical Pediatric Case Presentation of Erythema Elevatum Diutinum

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ABSTRACT

Erythema elevatum diutinum (EED) is a rare, chronic dermatosis that presents with red violet to red brown papules, plaques, and nodules on the exterior surfaces. Circulating immune complexes with repeated deposition and associated inflammation is thought to play a role in its pathogenesis and it is associated with various systemic diseases including infectious, autoimmune, and hematologic disorders. We report an interesting case of EED in a 6-year old patient who presented with an itchy rash and abdominal pain for 2 weeks and was later found to have Crohn’s disease.

CASE PRESENTATION

- A 6 year old Caucasian male presented with his mother for a 2-week history of a pruritic rash on his left elbow, lower legs, hands, and buttocks along with intermittent abdominal pain.
- PMH: None
- PSH: None
- Meds: None
- Allergies: NKDA

Clinical Course: The patient had complained of pruritis and his mother had been applying over the counter hydrocortisone cream with no improvement. He also experienced bouts of abdominal pain with diarrhea and constipation over the last two weeks and denied fever, chills or night sweats. The patient presented to our dermatology clinic and a shave biopsy was taken of a lesion on the right lower leg.

Biopsy Results: The biopsy results showed a mild hyperkeratotic epidermis, a grenz zone noted in the dermis, a perivascular and diffuse infiltrate composed of neutrophils and eosinophils.

The biopsy diagnosis was a neutrophilic dermatitis with a differential diagnosis including EED, a lesion related to Crohn’s disease, Behcet’s disease, or urticaria.

DISCUSSION

Variable | EED
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Epidemiology | Very rare, several hundred cases have been reported. Equally affects males and females most commonly middle or older age. No racial predilection is seen
Skin findings | Early lesions - erythematous or red/violet colored papules or purpuric which progress to become firm lesions
| Various presentations including: annular plaques with raised border, verrucous plaques on the soles, skin colored nodules usually seen on palmoplantar region usually in HIV infected patients
| Often asymptomatic but can be pruritic or burning in nature
Extra-cutaneous findings | Arthropalagia over affected joints, peripheral keratitis, nodular sciartis, panuveiitis and blindness
Distribution | Symmetrical over the extensor surfaces of the elbows, knees, hands, and feet. Additional sites may include the face, trunk, axillae, buttocks, genitals and Achilles tendon
Associated conditions: Infectious: Hep B, HIV, TB, Syphilis and beta-hemolytic streptococcal infections
| Autoimmune: Wegner’s granulomatosis, inflammatory bowel disease, celiac disease, relapsing polychondritis, SLE, RA and dermatitis herpetiformis
| Hematological: plasma cell dyscrasias, myelodysplasia, myeloproliferative disorder and hairy cell leukemia
Pathogenesis: Unknown but likely due to immune complex deposition resulting in complement activation, neutrophil recruitment and the release of destructive enzymes resulting in fibrin deposits around dermal vessels
Histologic features | Nodular and diffuse mixed infiltrates of neutrophils and eosinophils, plasma cells that extend to subcutaneous fat. Has a classic onion skin like perivascular fibrosis. Mixture of plasma cells and lymphocytes are the hallmark
Treatment | Dapsone, tetracycline, nicotinamide, sulfapyridine, cholchicine, intralesional / systemic corticosteroids
Clinical course | Relapsing and remitting. Majority resolve within 5 years

CONCLUSION

EED is a rare, chronic fibrosing leukocytoclastic vasculitis that presents with strange to yellow papules and plaques over the joints and the external surfaces of the elbows, knees, hands and feet. Multiple infectious, hematological and autoimmune conditions are associated with EED likely secondary to the presence of persistent circulating immune complexes leading to fibrin deposition around dermal vessels. The exact pathogenesis is unknown but thought to be associated with circulating immune complexes. It tends to have a relapsing and remitting course which can last multiple years and the mainstay of treatment is dapsone.

REFERENCES

CLINICAL PICTURE 1

• The clinical and histological differential diagnosis of EED depends on the stage of the lesion.
  - Clinically, early stage lesions resemble neutrophilic dermatoses, such as Sweet’s syndrome, purpuric vasculitis of the dorsal hands, and rheumatoid neutrophilic dermatitis. Late stage lesions resemble granuloma annulare, tuberous xanthomas, rheumatoid nodules, fibrod nodules of Borrelia, and multifibric reticulohistiocytosis.
  - Histologically, the differential diagnosis for early stage lesions resemble leukocytoclastic vasculitis with prominent interstitial neutrophils. Late stage lesions resemble lusious xanthomas, fibrotic disorders or tumors such as DFSP, fibrotic nodules of Borrelia, Kaposi sarcoma, and bacillary angiomatosis.

CLINICAL PICTURE 2