THE BRUISE THAT DOES NOT DISAPPEAR

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Background: Localized scleroderma is a rare musculoskeletal disease that can potentially severely affect a child's growth and function. The early lesions often do not strike physicians as worrisome as they can appear as demarcated erythema or a bruise like lesion. Like other rheumatologic diseases, history is key in distinguishing these worrisome lesions from similar benign lesions. It is important to recognize the lesions early as they are not reversible when they have reached the fibrotic stage. A localized scleroderma lesion can also be a sign of a more wide-spread disease such as eosinophilic fasciitis (EF).

Materials/Methods: We describe a 10 year old Hispanic female who presented with 6 months of progressive bruise-like lesion on her right leg. On presentation the lesions extended from above the knee to below the ankle. The lesion had not responded to topical treatment. The family also reported right knee swelling and difficultly using her hands.

Results: Initial exam was concerning for localized scleroderma lesion crossing two joints characterized by a hyperpigmented, waxy, indurated center with border of erythema. She also had skin changes of the left leg with prominence of veins notable at the medial knee. She was also noted to have diffuse swelling of her hands and flexion contractures of all of her fingers. Her labs were significant for an eosinophilia of 17% and hypergammagloblinemia. An MRI scan of her hips and pelvis was performed which was notable for striking edema along the fascial planes involving hip adductor and extensor compartments and the femoral extensor and flexor compartment. An MRI of her right hand which revealed extensive tendonitis and also marked superficial fascial edema. Biopsy confirmed the diagnosis of EF and demonstrated a mixed inflammation of the fascia consisting of lymphocytes, plasma cells, histiocytes and eosinophils.

Conclusions: Localized scleroderma is especially worrisome when it crosses joint lines as it can irreversibly affect joint function and cause limb length discrepancies. In this case the scleroderma lesion was actually the most visible manifestation of EF, which is a disease related to scleroderma that preferentially targets the fascia of the extremities in a symmetric fashion. Like scleroderma, the disease is difficult to treat in the inflammatory stage and once it has progressed to the fibrotic stage the changes can be permanent. Fibrosis of the tissues surrounding the growing skeleton of a child can lead to significant growth deformities.