A Rare Histologic Variant of a ‘Not So Sweet’ Rash

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Introduction

The neutrophilic dermatoses are a clinically diverse group of disorders with characteristic histology of intense epidermal and/or dermal inflammatory infiltrates composed primarily of neutrophils without evidence of infection.1

The neutrophilic dermatoses include Behcet’s disease, bowel (intestinal) bypass syndrome, erythema elevatum diutinum, neutrophilic dermatosis of the dorsal hand, neutrophilic eccrine hidradenitis, pyoderma gangrenosum, and Sweet’s syndrome (SS).7

The pathogenesis of neutrophilic dermatoses is unknown, however these disorders may represent a state of immunologic reactivity.4

SS (Acute febrile neutrophilic dermatosis) is an uncommon benign disease, with a worldwide distribution and female predominance except in the internal malignancy subgroup.4 SS is a reactive process that is characterized by fever, peripheral blood neutrophilia, and painful erythematous plaques that are occasionally bullous and favor the face, and upper extremities and contain dense neutrophilic dermal infiltrates histologically.4

SS can be subdivided into subcutaneous SS and histiocytoid SS. Histiocytoid SS is a rare variant of SS with a strong association with hematologic malignancies but rarely renal cell carcinoma.6 We present a case of a 65-year-old Caucasian male with an 8-year history of a diffuse unspecified rash previously managed by an outside provider. Subsequent punch biopsy of the left inferior postauricular skin confirmed the diagnosis of histiocytoid Sweet’s syndrome.

Case Presentation

A previously healthy, 65-year-old Caucasian male presented to the dermatology clinic with an 8-year history of a diffuse unspecified rash previously managed by an outside provider. Upon examination, multiple edematous red plaques were located on the neck, trunk and upper and lower extremities (Figure 1).

A 4 mm punch biopsy was obtained from the left inferior postauricular skin, which demonstrated mixed dermal infiltrate with karyorrhexis and a prominent histiocytoid cellular component (Figure 2). The biopsy was consistent with histiocytoid Sweet’s syndrome.

Our patient was initially treated with prednisone 20 mg daily and recommended to obtain a full malignancy workup. Upon completion of the malignancy workup, the patient was diagnosed with renal cell carcinoma of the right kidney and subsequently underwent nephrectomy.

Throughout the patient’s dermatologic management, the patient’s cutaneous findings were recalcitrant to prednisone alone, thus various doses of prednisone were tried along with indomethacin 150 mg daily in combination with multiple class I topical steroids which produced variable success. While on systemic steroids, daily calcium and vitamin D supplements were recommended. Steroid sparing agents such as dapsone were considered, but patient’s anemic state prevented safe and successful use. A trial of doxycycline and nitricamide was attempted with limited benefit.

Along with the recalcitrant cutaneous findings, the patient also experienced intermittent episodes of vision loss, palpitations, fatigue, anemia, dyspnea, atrial fibrillation, diffuse cutaneous fungal infections, and ultimately, metastatic renal cell carcinoma to the contralateral kidney and lung. Currently, the prednisone dose is being tapered in order to limit the systemic and immunomodulatory effect while the patient is being managed by oncology.

Discussion

SS was first described in 1964 by Dr. Robert Douglas Sweet as “acute febrile neutrophilic dermatosis”.2 In 1970 Whittle et al. reported a similar case and named it “Sweet’s syndrome”.7 SS is characterized by painful, erythematous, cutaneous plaques and nodules of rapid onset accompanied by fever, leukocytosis, and neutrophilia.7 The face, neck, and upper extremities are frequently involved.4 The characteristic histologic presentation consists of a diffuse dermal neutrophilic infiltrate with karyorrhexis and massive papillary dermal edema, which is responsible for the pseudovesicular clinical morphology.9 SS generally lacks leukocytoclastic vasculitis.8

SS can be further subdivided histologically into subcutaneous Sweet’s syndrome and histiocytoid Sweet’s syndrome. Histiocytoid Sweet’s syndrome is a rare variant of SS. Histologically it is characterized by dermal and/or subcutaneous infiltrate of neutrophils and “histiocytic” cells, which are immature myeloid cells that stain positively for myeloperoxidase.8 Recent studies suggest this variant may have a stronger association with hematologic malignancies but rarely with renal cell carcinoma.9 To the best of our knowledge this is the only reported case of renal cell carcinoma causing histiocytoid SS in the literature.

SS is benign and lesions typically involute in weeks to months without treatment, however recurrences occur in approximately 30% of cases. The most effective therapy for SS is oral prednisone (0.5–1.0 mg/kg/day) for 4–6 weeks. Other major alternative treatments for SS are potassium iodide (900 mg/day), dapsone (100-200 mg/day), and colchicine (1.5 mg/day).40 Nonsteroidal anti-inflammatory drugs, clofazimine, cyclosporine, thalidomide and interferon-a have also been reported to lead to improvement of Sweet’s syndrome.39

References