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Sweet's syndrome

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Acute febrile neutrophilic dermatosis is an uncommon, dramatic skin disease, occurring predominantly in females aged 30 to 60 years[1,2]. The principal features of Sweet's syndrome are fever, peripheral blood leucocytosis, multiple raised asymmetric erythematous sharply marginated, painful cutaneous plaques, dense dermal Infiltrate consisting of mature neutrophils and rapid response to steroid treatment. In the majority of patients it is preceded by upper respiratory infection, tonsillitis or flu-like illness by two to three weeks[3], Fever and leucocytosis suggest sepsis but the appearance of rash after one to two days should make one think about this differential.

Case Report

A 45 year old female, known case of chronic myeloid leukemia for four years on hydroxyurea was admitted with fever and erythematous, tender swelling about 12x5 cm on left forearm. She also had mildly erythematosus papular lesions over lateral aspects of both knees and right arm. She was febrile 37.8°C and had hepatosplenomegaly. The initial Investigations revealed a haemoglobm of 12.0 gm/dl, total leucocyte count 12,600/cmm with normal differential and platelet count of 238,000/cmm. Serum electrolytes, PT, APTT, chest x-ray and urine analysis were normal. Peripheral blood smears were negative for malarial parasite on two occasions and four blood cultures did not grow any organism. She was initially diagnosed to have cellulitis and treated with high dose benzyl-penicillin. She continued to spike temperature upto 40°C and complained of nausea, vomiting and severe headache. Her skin lesion on left forearm started to improve but she developed a new erythematosus, markedly indurated tender patch 10x10 cm on right buttock. As the patient was not responding, a punch biopsy was taken from right buttock lesion and she was started on tablet prednisolone 30 mg per day as the lesion clinically looked like erythema nodosum and she did not respond to antibiotics over a week. At the same time her antibiotics were stopped. The lady showed a dramatic response, being afebrile within forty-eight hours and feeling much better. Her lesion resolved in four days time. The skin biopsy revealed features consistent with acute febrile neutrophilic dermatosis (Sweet's syndrome).

Discussion

In 1964, Sweet[1] first described acute febrile neutrophilic dermatosis and since then more than 130 cases have been reported[2]. It was initially grouped with erythema nodosum or erythema multiform. The rash appears as populonodular tender lesions which coalesce and evolve over days and weeks[3] and later resolve without scarring. They are mostly on face, neck and upper limbs but may also be seen on lower limbs where they resemble erythema nodosum. It is associated with headache, malaise and arthralgia. Inconsistent findings are conjunctivitis or episcleritis[2] but these have been noted in over 60 percent of patients in one large series[4]. An important aspect of Sweet's syndrome is its association with various systemic conditions In particular acute mycloid leukemia[5-9] where It may even precede the hematological malignancy. Our patient was a known case of chronic myeloid leukemia and in retrospect it is important to be aware of this clinical entity to suspect and hence diagnose It. It has also been reported In association with ulcerative colitis[1,10] benign monoclonal gammopathy[11] and internal malignancies[5,12,13] and rheumatoid arthritis[14]. Histopathologic features are edema of the papillary

body and a dense infiltrate of leukocytes (neutrophils) in the lower dermis[15]. Perivascular foci of leucocytóclasia are quite common and at low magnification may suggest vasculitis[15]. These patients show prompt response to prednisolone 30-60 mg/d tapered to 10 mg within 2-3 days. They also show dramatic response to potassium iodide 900 mg daily for two weeks. However, recurrence may be seen in upto 50% of patients. Coichicine has also been tried[13].

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