

Case Letter

Sweet Syndrome Preceding a Carcinoid Lung Tumor and Multiple Myeloma

To the Editor:

Sweet syndrome (SS) is a rare neutrophilic dermatosis that is recognized as a paraneoplastic phenomenon, particularly in association with multiple myeloma (MM).^{1,2} We report a unique case in which SS preceded the diagnosis of a carcinoid lung tumor and MM by 11 years.

A 62-year-old woman with an otherwise unremarkable medical history presented to the dermatology department in 1996 with an intermittent fever and widespread painful plaques and nodules affecting the arms and legs of 1 year's duration. Initial blood tests showed only an elevated erythrocyte sedimentation rate. Histology revealed a diffuse inflammatory infiltrate with neutrophils in the dermis as well as marked edema and leukocytoclasia in the dermis. There was IgG paraproteinemia at 5 g/L but no urinary Bence Jones protein. Bone marrow protein aspirate was hypercellular, but the trephine was normal. A diagnosis of monoclonal gammopathy of undetermined significance was made and outpatient follow-up commenced. Because of coexistent iron deficiency, she was started on oral iron replacement. These findings were considered to be secondary reactive changes due to iron deficiency, and the patient was started on iron replacement therapy. A diagnosis of SS was made, which eventually was controlled with oral sulfamethoxypyridazine. The IgG paraprotein uneventfully persisted at 7 g/L. In early 2007, the patient developed hemoptysis and general malaise. She was diagnosed with a carcinoid lung tumor, which was treated via resection of the left lower lobe. The IgG paraprotein remained stable at 7 g/L. In December 2007, the patient presented with a painful thickened area on the right parietal eminence of 2 months' duration. Cranial computed tomography showed multiple osteolytic skull lesions. She had normocytic normochromic anemia (hemoglobin, 10.3 g/dL [reference range, 14.0–17.5 g/dL]), a normal white blood cell count, and a platelet count of 120 g/dL (reference range, 150–400 g/dL). The IgG paraprotein level was 20 g/L. Bone marrow aspirate and trephine demonstrated 20% and 15% plasma cell infiltration, respectively. A diagnosis of MM was established and the patient started therapy with lenalidomide, dexamethasone, and granulocyte colony-stimulating factor (G-CSF). There has been no further reoccurrence of the carcinoid tumor.

From the Department of Dermatology, Chelsea and Westminster Hospital, London, England.

The authors report no conflict of interest.

Sweet syndrome originally was described as acute febrile neutrophilic dermatosis by Sweet¹ in 1964. It can present as the following clinical entities: classic or idiopathic SS with a constellation of features that have no other identifiable cause; malignancy-associated SS with SS and a synchronous or metachronous malignancy³; and drug-induced SS with the most frequently implicated drug being G-CSF.²

In patients with MM, the incidence of SS is 0.25%.³ Our patient had persistent IgG paraproteinemia concurrent with SS. Elevated levels of paraproteins in cases of MM have not been directly incriminated in the pathogenesis of SS, as it is believed that elevated levels of G-CSF or granulocyte-macrophage colony-stimulating factor are more likely to be the cause.² Our patient did not receive G-CSF until the onset of MM.

Most cases of SS occur almost immediately preceding or synchronous with a diagnosis of MM, thus acting as a paraneoplastic marker. Most neoplasms associated with SS also tend to be of plasma cell, lymphoid, or other bone marrow-related origin; the association with acute myeloid leukemia is the most common.⁴

Our case is unique in that SS preceded the carcinoid tumor, which most likely is a coincidence, but this finding warrants consideration in the broader context of SS as a paraneoplastic phenomenon. It also is rare for patients with SS to develop 2 neoplasms. The carcinoid tumor and MM developed 11 years after SS was diagnosed, which also makes this case unique.

Angela Tewari, MBBS, BSc, MRCP
Arani Chandrakumar, MBBS, MRCP
Donald Macdonald, MBBS, MRCP, FRCPath
Richard Staughton, MA, MB BChir, FRCP
Christopher B. Bunker, MD, MBBS, FRCP

REFERENCES

1. Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermatol.* 1964;76:349-356.
2. Su WP, Liu HN. Diagnostic criteria for Sweet's syndrome. *Cutis.* 1986;37:167-174.
3. Bayer-Garner IB, Cottler-Fox M, Smoller BR. Sweet syndrome in multiple myeloma: a series of six cases. *J Cutan Pathol.* 2003;30:261-264.
4. Buck T, González LM, Lambert WC, et al. Sweet's syndrome with hematologic disorders: a review and reappraisal. *Int J Dermatol.* 2008;47:775-782.