Case report
Sweet Syndrome Associated With Myelodysplastic Syndrome: Report of a Case. Review of the Literature

Delia Reina, a,∗ Dacia Cerdà, a Daniel Roig, a Ramon Fíguls, a M. Luz Villegas, b Hèctor Corominas a

a Unidad de Reumatología, Hospital Sant Joan Despí Moisés Broggi, Sant Joan Despí, Barcelona, Spain
b Servicio de Medicina Interna, Hospital General de l’Hospitalet, Barcelona, Spain

A R T I C L E   I N F O
Article history:
Received 21 October 2011
Accepted 25 January 2012
Available online 1 September 2012

Keywords:
Sweet syndrome
Myelodysplastic syndrome
Arthritis

A B S T R A C T
Sweet’s syndrome or acute neutrophilic febrile dermatosis is a systemic disease of unknown etiology characterized by the appearance of skin lesions produced by a neutrophilic dermal infiltrate, fever, and peripheral leukocytosis. It may be associated with hematologic diseases, including leukemia, with immune diseases as rheumatoid arthritis, or can occur in isolation. The myelodysplasias are hematological disorders characterized by one or more cytopenias secondary to bone marrow dysfunction. We present the case of a patient with Sweet’s syndrome associated with myelodysplastic syndrome and treated with glucocorticoids who did not present a good clinical outcome. We discuss the different treatment of these diseases because in most cases glucocorticoids, which are the treatment of choice in Sweet’s syndrome, may be insufficient.

© 2011 Elsevier España, S.L. All rights reserved.

Síndrome de Sweet asociado a síndrome mielodisplásico: a propósito de un caso. Revisión de la literatura

R E S U M E N
El síndrome de Sweet o dermatosis neutrófilica febril aguda es una enfermedad sistémica de etiología desconocida, caracterizada por la aparición de lesiones cutáneas producidas por un infiltrado dérmico neutrófilico, fiebre y leucocitosis periférica. Puede estar asociado a enfermedades hematológicas, incluida la leucemia, inmunológicas como la artritis reumatoide o presentarse de forma aislada. Las mielodisplasias son trastornos hematológicos caracterizados por una o más citopenias secundarias a la disfunción de la médula ósea. Se presenta el caso de un paciente con síndrome de Sweet asociado a un síndrome mielodisplásico que ha seguido tratamiento con glucocorticoides y no ha presentado una buena evolución clínica. Se discuten los diferentes tratamientos de estas enfermedades porque en la mayoría de las ocasiones los glucocorticoides, que son el tratamiento de elección en el síndrome de Sweet, pueden ser insuficientes.

© 2011 Elsevier España, S.L. Todos los derechos reservados.

Introduction

Sweet’s syndrome (SS) is a skin disease of unknown etiology1,2 which is characterized by fever, leukocytosis, and painful erythematous skin plaques. Histology shows dense neutrophilic infiltrates. The presence of anemia and thrombocytopenia may be associated with an underlying neoplastic.3

We present the case of a patient with SS and anemia diagnosed as a myelodysplastic syndrome (MDS), worsening the prognosis. MDS are hematologic diseases characterized by cytopenias that result in dysmorphic states of the cells of the bone marrow. One type of MDS is refractory anemia with blast excess (RABE), with the number of blasts being greater than 5% and less than 10% in type 1, and between 10% and 20% in type 2. These diseases are refractory to chemotherapy and stem cell transplant offers a cure. In RABE, supportive treatment is performed with red blood cell transfusions.

Case Report

The patient is a 70 years old male with no history of interest, who had recurrent episodes of 2 weeks with high fever, leukocytosis, and
The disease is more common in women in the fifth decade of life. The ESR is elevated and anemia present. It can last around 15%. The disease becomes chronic and recurrent. Indomethacin was prescribed a dose of 150 mg/day and potassium iodide was added, without improvement. In parallel, the patient was diagnosed with a type 1 RABE after the study of persistent neutrophilic infiltrate of the reticular dermis.

Systemic glucocorticoids are the treatment of choice. Indomethacin is effective. Colchicine, potassium iodide, dapsone, cyclosporine, interferon alpha or etretinate are therapeutic alternatives. There have been reports of SS and MDS successfully treated with intravenous immunoglobulin, glucocorticoids, and doxycycline or stem cell transplantation.

**Conclusion**

SS is a rare dermatological entity, which when accompanied by an MDS may worsen the patient’s prognosis. The treatment of choice for SS is the use of glucocorticoids, although in most cases this may be insufficient and require alternative therapies.

**References**