

Sweet's syndrome and pancreatic neoplasm: an atypical association

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RESUMEN

El síndrome de Sweet es una dermatosis inflamatoria poco común, que se ha asociado a tumores malignos, principalmente de tipo hematológico. Presentamos un caso clínico de síndrome de Sweet asociado con una rara neoplasia pancreática, siendo uno de los pocos casos reportados en la literatura médica acerca de esta asociación.

Palabras claves: Síndrome de Sweet; Dermatitis Neutrófilica; Neoplasia Pancreática; Enfermedades Paraneoplásicas.

SUMMARY

Sweet's syndrome is an uncommon inflammatory dermatosis, which has been associated with malignant tumors, mainly of hematological type. We report a clinical case of Sweet syndrome associated with a rare pancreatic neoplasm, which is one of the few cases reported in the medical literature about this association.

Key words: Sweet's Syndrome; Neutrophilic Dermatoses; Pancreatic Neoplasm; Paraneoplastic Diseases.

Sweet's syndrome (SS) or acute febrile neutrophilic dermatosis, is a rare inflammatory condition characterized by fever, neutrophilia, and abrupt onset of painful erythematous nodules or plaques,^{1,2,3} a dermal infiltrate of mature neutrophils on histology and a fast response to the steroidal treatment.^{4,5} It has an incidence of 3 cases per million persons-year⁶, and predominates in women (female-male ratio, 4:1).^{3,7} Malignancy is associated with SS in approximately 20% of cases,⁸ being the hematological tumors the most frequently associated (85%),⁷ and on the other hand, nearly two-third of the patients with an association with solid tumors have carcinoma of genitourinary organs.⁹ Intraductal papillary mucinous neoplasms (IPMN) is a cystic neoplasm from the epithelial cells lining the pancreatic ductal system, with an incidence of 1/281,000 patients per year,¹⁰ and it is the first case of SS associated to IPMN in medical literature.

CASE

An 80-year-old man, with compensated diabetes and treated with metformin, was referred to our hospital with fever, polyarthralgia and multiple nodular erythematous and painful lesions with pseudovesicles on his face, neck and extremities of a four-days evolution (Figure 1). There were no digestive symptoms, weight loss, muscle weakness, or anorexia. A full blood examination showed leukocytosis (10.300/mm³) with neutrophilia (84%), erythrocyte sedimentation rate (30 mm/hour) and C-reactive protein (32,2 mg/dL). Histologically, a skin biopsy from a lesion on the neck demonstrated pustules and edema of the upper dermis, accompanied by perivascular neutrophilic infiltration (Figure 2). These findings were consistent with SS. The patient was admitted to our hospital and received prednisone 60 mg/day by oral administration, with a fast improvement of fever and skin lesions. Searching

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Figure 1
Sweet's syndrome, clinical manifestation. Multiple nodular erythematous lesions on the face and the hands.

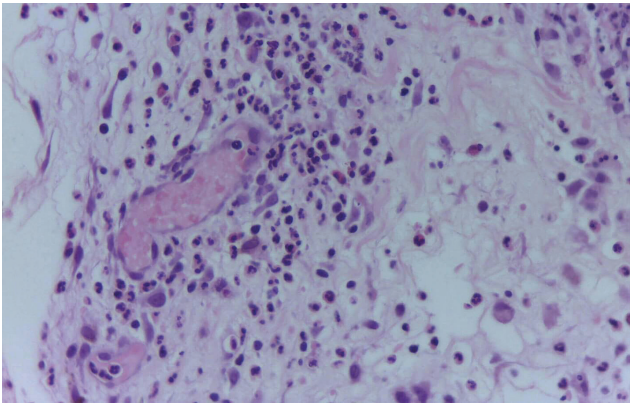


Figure 2
Sweet's syndrome, histopathology (40x, original magnification). Perivascular neutrophilic infiltration with no evidence of vasculitis.

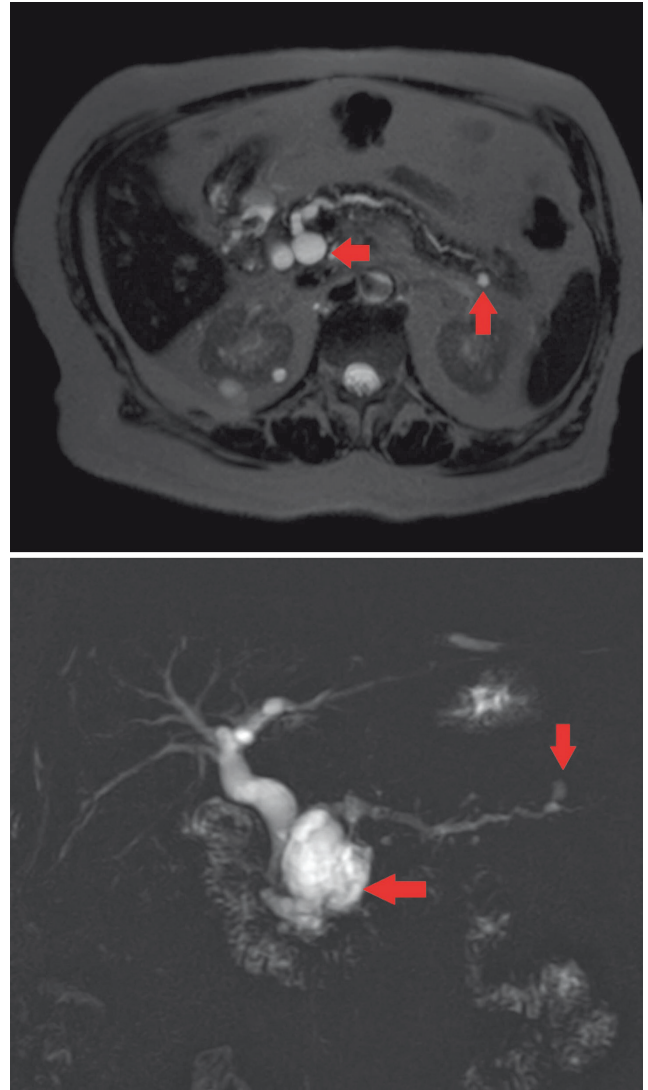


Figure 3
Intraductal papillary mucinous neoplasms, radiology. Tumoral lesions located in the pancreas (red arrows), with abdominal magnetic resonance (above), and cholangioresonance (below).

for indicators of malignancy, an abdominal magnetic resonance and a cholangioresonance showed two IPMN, located in the body and head of the pancreas (Figure 3). The location of the tumors determined that surgical treatment was not an option. A low-dose corticosteroid therapy was therefore considered to reduce the risk of recurrence. After 6 months, the patient didn't present new exacerbations.

DISCUSSION

SS is an inflammatory skin condition characterized by tender, erythematous and edematous skin lesions and systemic corticosteroids are the cornerstone of management for this illness. According to the patient's age, tests to rule out malignancies were performed, and a pancreatic neoplasm was diagnosed. Additionally, we should consider that the SS could reappear because the pancreatic lesions were not removed, and a strict follow-up becomes indispensable, because a recurrence of SS has been reported in up to two-thirds of the cases, when the neoplasm has not been removed.⁴ There have been reports of SS arising in association with other malignancies; however, it is the first reported association of SS with a pancreatic neoplasm in medical literature.

CONCLUSION

It is imperative that the dermatologist doesn't miss the sentinel nature of this clinical syndrome, because the SS could alert the physician to the diagnosis of cancer, especially in elderly people.

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