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Case report

Adult-onset Still's disease associated with gastric cancer☆

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ABSTRACT

The diagnosis of Adult-onset Still's disease (AOSD) requires the exclusion of infectious, malignant, and autoimmune diseases. However, a poorly symptomatic neoplastic process can easily be overlooked, or even onset later during the course of the disease. Therefore, numerous cases of Adult-onset Still's disease associated with malignancy have been reported. The case is reported of an 84-year old woman with previous diagnosis of AOSD who developed a gastric tumor with fatal outcome 2 years after the diagnosis of her rheumatic disease.

Enfermedad de Still del adulto asociada a neoplasia gástrica

Palabras clave:
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RESUMEN

El diagnóstico de enfermedad de Still del adulto requiere la exclusión de cuadros infecciosos, tumorales y autoinmunes. Sin embargo, un proceso neoplásico poco expresivo clínicamente y en las pruebas complementarias puede pasar desapercibido al diagnóstico o comenzar con posterioridad, habiéndose descrito numerosos casos de enfermedad de Still asociada a tumores. Presentamos el caso de una paciente de 84 años con diagnóstico previo de enfermedad de Still del adulto, que desarrolló un tumor gástrico de evolución fatal 2 años después del diagnóstico del cuadro reumatológico.

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Introduction

Adult-onset Still's disease is a rare multisystemic condition characterized by the presence of intermittent fever in peaks generally in the evening, arthritis or arthralgias and macular or maculopapular salmon-colored evanescent rash, typically coincident with the fever. Other frequent characteristics are lymphadenopathies, hepatosplenomegaly, odynophagia, myalgias, serositis, leukocytosis and neutrophilia.[1,2] There are different classification criteria for adult-onset Still's disease that require the exclusion of neoplastic, infectious and autoimmune processes. However, sometimes a paucisymptomatic tumor may go unnoticed or even begin months after the adult-onset Still's disease, hence the importance of maintaining a close follow-up of the patient and a high index of suspicion in the presence of any data suggestive of possible neoplastic disease throughout it.[3,4]

Clinical observation

An 84-year-old woman with a history of arterial hypertension and dyslipidemia, diagnosed at 82 years of age with adult-onset Still's disease due to a clinical picture of deterioration of the general condition, with daily spiking fever, orange-pink macular rash in lower limbs coinciding with fever, arthromyalgias, odynophagia and chest pain with pleuropericardial characteristics. Analytically, leukocytosis with neutrophilia, elevated CRP and ESR, hypertransaminasemia, marked hyperferritinemia (16,400 ng/ml) and elevation of soluble CD25 stood out, and the autoimmunity study showed negative ANA and RF and normal complementemia. The thoracoabdominopelvic CT scan showed bilateral pleural and mild pericardial effusion, as well as an image of diffuse increase in the density of rhe mesenteric fat of the small intestine, suggestive of mesenteric panniculitis. As part of the study to rule out underlying neoplasia, a PET/CT scan and a bone marrow biopsy were performed, which did not showed findings of interest. Once an infectious etiology was reasonably ruled out by means of a microbiological study, as well as a possible tumor cause, treatment was initiated with boluses of 1 g of methylprednisolone IV for 3 days and subsequently oral prednisone at a dose of 1 mg/kg/day, presenting partial improvement and developing diabetes and steroid myopathy as complications. For this reason, it was decided to start treatment with anakinra 100 mg/day SC (taking into account the advanced age of the patient, it was considered as a safer option and with less risk of complications than other cytostatic agents), with rapid clinical and analytical response, which allowed to decrease and even discontinue the steroids, and it was maintained until October 2015. Ten months later, the patient developed a picture of odynophagia, abdominal pain and nausea with elevation of acute phase reactants, without improvement with steroids at medium doses, so in August 2016 the anakinra was reintroduced, with little response. It was performed an abdominal ultrasound, that was normal, and given the clinical worsening, the patient was admitted to the hospital to complete the study. Analytically, in addition to the elevation of acute phase reactants, there was an alteration of the mixed hepatic profile and a slight deterioration of kidney function. The CT scan showed a large abdominal tumor, of probable gastric origin, with infiltration by contiguity of the left hepatic lobe and pancreatic tail, presence of hepatic nodules suggestive of metastasis and peritoneal carcinomatosis. Given the tumor extension and the poor general condition of the patient, she was not considered a candidate for chemotherapeutic treatment by the medical oncology service or to performing invasive endoscopic procedures for biopsy, and palliative management was decided. Finally, the patient died a few days after admission.

Discussion

Although the diagnosis of adult-onset Still's disease is a diagnosis of exclusion that requires a thorough analytical and imaging study to rule out the presence of possible tumor processes that explain the patient's symptoms, numerous cases in which there is a time relationship between Still's disease and the development of neoplasia have been described in the literature, and it may happen that an early stage tumor process goes unnoticed in the initial study or that it begins after the diagnosis of adult's Still disease (often months after it). These neoplasms can be solid, and the most frequently described associations are ductal breast carcinoma and non-small cell lung cancer, or hematological, mainly diffuse large B-cell lymphomas and T-cell lymphomas.[3]

Some differential characteristics of adult-onset Still's disease associated with neoplasia with respect to the primary forms of the disease are: more advanced age of onset, poor initial response to steroid or immunosuppressive treatment and presence of atypical clinical, biological and immunological characteristics of the disease[3,4] (for example, an atypical non-macular or maculopapular rash, or not coincident with fever[5]; presence of atypical leukocytes in peripheral blood or marked elevation of LDH in the case of lymphoproliferative processes). In our case, the characteristic that attracts attention is the very advanced age of the patient: although there are other cases described in the literature of patients in the seventh decade of life at the time of the diagnosis of Still's disease,[6] 75% of these cases occur between 16 and 35 years of age,[7] while in the cases of Still's disease associated with neoplasia, 71% of the cases occur in patients over 40 years of age.[4]

In all patients with adult-onset Still's disease and especially in those with any of the above mentioned characteristics, it is important to keep a close follow-up and a high index of suspicion in the presence of any clinical data suggestive of possible underlying neoplastic disease. While there are currently no recommendations in the literature on the systematic implementation of complementary tests in such follow-up, it seems reasonable to conduct an analytical and imaging study aimed at the findings suspicious in the anamnesis and physical examination (e.g., abdominal CT scan and endoscopies if there are digestive symptoms.)

Conflict of interest

The authors declare that they do not have any conflict of interest.

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