Case report

Subcutaneous sarcoidosis: a rare involvement of the paralateral nasal region

Sarcoidosi sottocutanea: un raro caso di coinvolgimento della regione paralatero nasale

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SUMMARY
A case of subcutaneous sarcoidosis involving the paralateral nasal region is described and a brief review of the literature is made. Subcutaneous sarcoidosis without systemic disease is a rare entity and has seldom been reported on the trunk and face. Diagnosis is always difficult as it can only be confirmed by histological studies.

KEY WORDS: Head & neck • Paralateral nasal region • Subcutaneous sarcoidosis

RIASSUNTO
In questo articolo viene descritto un raro caso di sarcoidosi sottocutanea con coinvolgimento della regione paralatero-nasale e viene presentata una revisione della letteratura sull’argomento. La sarcoidosi sottocutanea senza interessamento sistemico è una patologia inusuale, e ancor più raramente è riportato il coinvolgimento del distretto cervico-facciale. La diagnosi di tale affezione può essere difficoltosa: necessita sempre della conferma istologica.

PAROLE CHIAVE: Testa e collo • Regione para-latero nasale • Sarcoidosi sottocutanea

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Introduction

Subcutaneous sarcoidosis is a rare cutaneous expression of systemic sarcoidosis.
Sarcoidosis is a multisystem disorder characterized by the formation of non-caseating granulomas. It is reported to be caused by an oligoclonal response of CD4-positive T cells to an unknown antigen. The disorder may be manifested in numerous ways. In 90% of the patients, intra-thoracic involvement is revealed on chest radiography. Other frequently involved sites are the reticulo-endothelial system, the skin, and the eye. Sarcoidosis can also affect joints, muscles, exocrine and endocrine glands, the heart, the central nervous system, the upper respiratory tract, and the kidney.

Subcutaneous sarcoidosis is characterized by non-infectious sarcoideal or epithelioid granulomas with minimal lymphocytic inflammation involving predominantly the panniculus. Subcutaneous involvement, over the extremities, occurs primarily in 20-35% of patients with systemic sarcoidosis, but may also occur without systemic disease. Histological assessment is mandatory for a definitive diagnosis. The rare case of subcutaneous sarcoidosis with paralateral nasal involvement is presented.

Case report

A 38-year-old female was addressed to the ENT Department of the University Hospital of Ferrara for the assessment of a subcutaneous right paralateral nasal mass, also involving the suborbital region.
The patient had been complaining of a swelling in her right paralateral nasal region, associated with recurrent frontal headache, for 3 months. She had already been treated with systemic steroids (prednisone 25 mg/die for 3 weeks) and antibiotics (ciprofloxacin 500 mg/die for 10 days), which were unsuccessful. Computed tomography (CT) scans and magnetic resonance imaging (MRI) of the paranasal sinuses were performed (Fig. 1), disclosing a subcutaneous mass, about 1 cm in diameter, at the inferior edge of the right nasal bone.

Her past medical history revealed histological diagnosis of erythema nodosum (EN) (right leg), when she was aged 21 years. She underwent an open septo/rhinoplasty when she was 33 (in another hospital).

At the examination, the lesion appeared nodular, non-tender, firm and painful, in the right paralateral nasal region. Serological testing for syphilis, anti-neutrophil cytoplasmatic antibodies, and antinuclear antibodies all resulted negative. Serum angiotensin-converting enzyme (ACE)
Subcutaneous sarcoidosis of the paralateral nasal region

Fig. 1. The paranasal sinuses CT (a) and MRI (b) scans revealed a subcutaneous mass about 1 cm in diameter, located at the inferior edge of the right nasal bone. Right nasal bone erosion can be detected at CT.

The patient was addressed to the Rheumatology Unit where she was treated with prednisone 12.5 mg/die and this treatment has been maintained till now. After 30 months’ of follow-up, no signs of local recurrence have been observed (Fig. 3) and, so far, no other localizations of disease have been reported.

Discussion and conclusions

Sarcoidosis is a multisystem disease defined by the formation of non-caseating granulomas in various organs. The diagnosis is well established when clinical and radiological findings are supported by histological evidence of non-caseating granulomas in one or more tissues. Subcutaneous involvement occurs in 20-35% of patients with systemic sarcoidosis but may also occur alone without systemic disease. Subcutaneous sarcoidosis alone is defined as a rare manifestation of sarcoidosis. Darier and Roussy reported the first case of subcutaneous sarcoidosis in 1904. Vainsencher and Winkelmann, in 1984, reported...
4 cases of subcutaneous sarcoidosis, 3 of which showed systemic involvement. It is reported to occur mainly in women, most often in their fifth or sixth decades.

The diagnosis of subcutaneous sarcoidosis is difficult to achieve and can only be confirmed by histological studies. Various non-specific laboratory abnormalities may be seen, such as an elevated erythrocyte sedimentation rate and hyper-gamma-globulinaemia. An elevated serum angiotensin-converting enzyme (ACE) level is reported to have a sensitivity of 40-90% and a positive predictive value of up to 90% but may be falsely positive in several other granulomatous diseases, hyperthyroidism, liver cirrhosis, diabetes mellitus, and Gaucher’s disease. Furthermore, it may be falsely negative when ACE inhibitors are used. Other reported laboratory abnormalities include hypercalcaemia and hypercalciuria (2-63%)..

A specific diagnostic test is the Kveim-Siltzbach skin test that has a reported sensitivity of 30-96% and is not widely used. The histological assessment is mandatory for a definitive diagnosis, as in the case presented here.

The association between subcutaneous sarcoidosis and EN, as in this case, is not frequently reported and some Authors describe the occurrence of EN as a first manifestation of immunologic disease.

Moreover, the present case report illustrates a rare localization of subcutaneous sarcoidosis, as the nodules are mainly distributed over the extremities (upper extremities, mainly in the forearms, and usually are bilateral and asymmetric) and rarely on the trunk and face. Differential diagnosis includes lipoma, lupus vulgaris, leprosy, rosacea, granuloma, reaction to foreign bodies.

At present, there is no curative treatment for sarcoidosis.

References


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