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E-CASE REPORT

Intralymphatic Histiocytosis: A Report of 2 Cases☆

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KEYWORDS
Intralymphatic histiocytosis;
Reactive cutaneous angiomatosis;
Rheumatoid arthritis;
Treatment

Abstract Intralymphatic histiocytosis is a benign condition characterized by poorly defined erythematous plaques (sometimes forming a reticular pattern) as well as the presence of nodules and vesicles. Its etiology and pathogenesis appear to be related to chronic inflammation in the affected area, prior surgery, or systemic disease, particularly rheumatoid arthritis. We report on 2 new cases, both associated with joint surgery in the affected area and osteoarticular disease (primary synovial osteochondromatosis and rheumatoid arthritis). This is a chronic disease and there is no specific treatment. Different treatment options were chosen in the 2 cases described. A spectacular response to treatment with oral pentoxifylline and topical tacrolimus was observed in 1 of the patients.
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PALABRAS CLAVE
Histiocitosis intralinfática;
Angiomatosis cutánea reactiva;
Artritis reumatoide;
Tratamiento

Histiocitosis intralinfática, a propósito de 2 casos

Resumen La histiocitosis intralinfática es un proceso benigno caracterizado por placas eritematosas mal delimitadas, a veces reticuladas, otras con nódulos o vesículas en su interior. Su etiopatogenia parece estar relacionada con procesos inflamatorios crónicos en el área afectada, cirugía previa o con enfermedades sistémicas, sobre todo con la artritis reumatoide. Presentamos 2 nuevos casos, ambos asociados a cirugía articular del área afecta y enfermedad osteoarticular (osteocondromatosis sinovial primaria y artritis reumatoide). Esta entidad tiene un comportamiento crónico y no existe un tratamiento específico. En los 2 casos presentados se plantearon distintas opciones terapéuticas, mostrando uno de ellos una respuesta espectacular al tratamiento con pentoxifilina oral junto con tacrolimus tópico.
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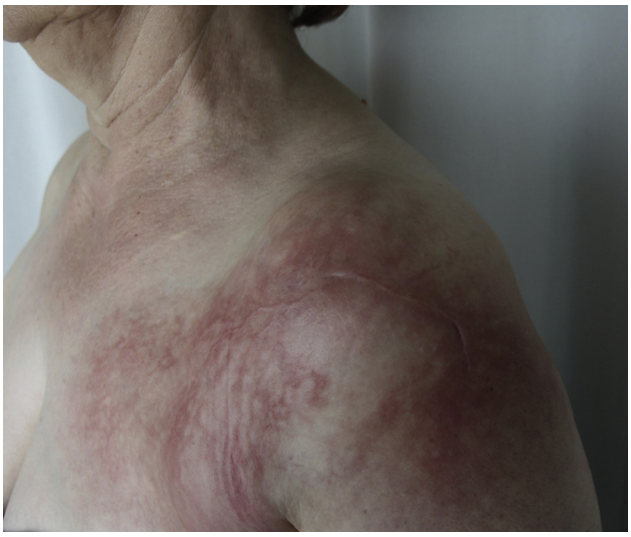

Figure 1 Case 1. Edematous plaque with a livedoid appearance and poorly delimited borders covering the entire shoulder region, extending to the proximal third of the deltoids.

Introduction

Intralymphatic histiocytosis is a rare entity that was first described by O'Grady et al[1] in 1994. Some 54 cases have since been reported, most in association with rheumatoid arthritis and in patients with prosthetic joint replacements.[2,3] Because the clinical presentation is nonspecific, diagnosis is fundamentally based on histologic findings. Immunohistochemistry demonstrates the presence of dilated lymphatic vessels (positive for D2-40, CD34, and CD31) in the reticular dermis and aggregates of (CD68-positive) intraluminal histiocytes.[2] It has been hypothesized that this dilation of the lymphatic vessels is caused by an obstruction of lymphatic drainage arising from either a congenital abnormality or acquired damage in the vessels.[2]

Case Descriptions

The first patient was a 65-year-old woman who had undergone surgery to repair the rotator cuff of the left shoulder 1 year earlier. She had no history of rheumatoid arthritis or other diseases. The patient sought care for an erythematous and slightly pruritic patch on the left shoulder that had appeared 2 months earlier and had not responded to various regimens of oral antibiotics prescribed to treat the initial suspicion of cellulitis. Physical examination revealed an erythematous, edematous plaque with a livedoid appearance and poorly defined borders, covering the entire shoulder region and extending to the proximal third of the deltoids (Fig. 1). A 4-mm punch biopsy was diagnostic, showing dilated vessels in the reticular dermis that were positive for CD31, CD34, and D2-40 as well as CD68-positive intraluminal cell aggregates (Fig. 2). The results of a full blood workup were normal, including levels for complement, immunoglobulins, rheumatoid factor, and C-reactive protein. Magnetic resonance imaging showed signs of advanced joint degeneration and the possible existence of evolved primary synovial osteochondromatosis, although no malignancy has been demonstrated to date.

The second patient was a 77-year-old woman who had been diagnosed with rheumatoid arthritis 20 years earlier and was receiving treatment with nonsteroidal antiinflammatory drugs. Two months after surgery on the left shoulder (repair of a partial biceps tendon rupture), she sought care for an asymptomatic patch that had gradually appeared in the weeks following the procedure (Fig. 3). During rehabilitation treatment, she was diagnosed with contact dermatitis, although no clear causative agent was identified. Topical corticosteroid treatment was prescribed, but no response was achieved. Physical examination revealed an erythematous, edematous plaque with imprecise borders on the anterior surface of the left shoulder, extending to the pectoral area. Skin biopsy, performed on the basis of clinical suspicion of intralymphatic histiocytosis, showed dilated vessels in the reticular dermis that were positive for CD31, CD34, and D2-40 (Fig. 4), CD68-positive cell aggregates inside lymphatic vessels of the reticular dermis, and perivascular lymphohistiocytic inflammatory cellularity, all confirming the diagnosis. Treatment was started with topical tacrolimus, applied once daily, and oral pentoxifylline (400 mg/d), achieving an excellent response and practically complete resolution after 4 months of treatment (Fig. 5).

Discussion

Intralymphatic histiocytosis is a rare entity that appears to be more frequent in older adults and in women.[1–5] The most common site is the limbs, including the shoulder and hip areas.[6] Intralymphatic histiocytosis has a nonspecific clinical presentation[5] and tends to manifest as a single erythematous plaque and occasionally in the form of papules or nodules.[3] Most of the fewer than 60 cases described in the literature[3] are associated with rheumatoid arthritis,[1–9] with skin alterations on the affected joints or, less frequently, in patients with prosthetic joint replacements.[1,2,5–7,10] There have also been anecdotal reports of intralymphatic histiocytosis in association with surgical scars in patients with breast cancer[1,5] and Merkel cell carcinoma,[1,5] in the pectoral region of a patient with cancer of the colon,[2,5] in the chest of a patient who underwent axillary lymphadenectomy to treat lymphatic tuberculosis,[2] and in a patient with Crohn disease with involvement of the suprapubic region.[2,5] Cases of primary intralymphatic histiocytosis, without association with any underlying disease, have also been reported.[2]

The etiology and pathogenesis of intralymphatic histiocytosis are not entirely understood, as the disease has been associated with pathologic processes of various kinds. Some authors classify it as a cutaneous reactive angiomatosis, a histopathologic pattern that arises in reaction to various inflammatory processes.[1,3,7] The most widely accepted hypothesis is that the lymphatic vessels dilate in response to obstruction of lymphatic drainage caused by congenital abnormalities of the vessels, or by acquired damage from trauma, surgery, radiotherapy, or chronic inflammatory processes such as rheumatoid arthritis.[1,7] Lymphostasis leads to poor antigen clearance, thus chronically stimulating the proliferation and aggregation of histiocytes.[1,2,7] Chronic

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