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RESEARCH ARTICLE

UNUSUAL PATTERN OF LIMB EDEMA: THE SCHULMAN'S DISEASE. CONTRIBUTION OF IMAGERY

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Abstract

Schulman's disease is a uncommon pathology described by Shulman in 1974. It is an attack of the connective tissue characterized by a symmetrical, indurated, painful edema associating a thickening of the skin and soft tissues which usually sits on the limbs with possibility of migrating to the trunk. Eosinophilic fasciitis or Shulman syndrome presents two peaks of incidence, between 20 and 30 years of age and between 45 and 60 years of age. Some pediatric and geriatric cases are described in the literature. It occurs sporadically and there do not seem to be, with a few exceptions, family cases. The place of imaging is not well defined but seems to be useful for diagnosis (especially MRI).

The diagnosis of certainty is brought by the biopsy of the fascia which finds a thickening of the fascia's with infiltration of lymphocytes and macrophages with or without eosinophils. Essentially medical treatment with corticosteroids or immunosuppressants is enough in most cases. The prognosis of Schulman disease is generally good. The aim of this work is to present and recall the imaging characteristics of Schulman syndrome from an illustration of a case and reviewed from the literature.

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Introduction:-

Necrotizing Fasciitis or Schulman's Disease was first described in 1974 by Dr. Schulman. It is a rare pathological entity, of poorly known etiology, the exact incidence of which is not well established, with only 300 cases reported in the literature since its first description. Its present two peaks off incidence between 20-30 and 45-60 years old. The demonstrations clinics are dominated by edema symmetrical painful, associated with induration and to a progressive thickening of the skin and soft tissues underlying dominate the ends.

The histology confirms the diagnosis by the demonstration of an infiltrate of lymphocytes and eosinophils in the fascia, which must be coraller to clinical, biological and the imagery.

Treatment with corticosteroid or immunosuppressants is generally sufficient. It's a good prognosis of disease with regression of symptoms in most situations. The case and literature review that we describe allows us to review the clinical and radiological characteristics of this condition.

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Observation:-

This is a patient of 63 years, in good condition overall, without ATCD, who consults for edema originally isolated, rapidly progressive members from six (6) months. The clinical examination noted a hardening of the skin, an orange peel appearance on the limbs, relative reduction in mobility and some joint pain. The biology shows a hyper eosinophilia (22%) and moderate inflammatory syndrome. Ultrasonography legs shows a hypoechoic hypodermis (edematous) and a fascia thick moderately higher than the normal range so bilateral. MRI of the lower limbs is in favor of a discrete signal abnormality thickening with fasciae of the two legs, one on T2 STIR, iso T1 ESF signal, a Weakness e Color Enhance after injection of gadolinium, hyperintense muscle of the leg on the sequences with suppression of Fat in T2 F S. The positive diagnosis is made on the histological data revealing a thickening of the superficial fascia site of an inflammatory infiltrate. The patient was taken care of dermatology department and received corticosteroid treatment. The evolution has been marked by a reduction of edema of the lower limbs and near complete remission of the edema of the members above.

Discussion: -

Eosinophilic fasciitis (EF), is a rare disorder of connective tissue, first reported by Lawrence Schulman in 1974, of poorly known etiology, the exact incidence of which is not well established, with 300 cases only reported in the literature since it was first described^[1-3]. The average age of onset has occurred is between 40-50 years.

The clinic directs, but there are no real published diagnostic criteria for this disease. These are cutaneous-phaneric manifestations such as painful symmetric (figure 1) edema, associated with progressive induration, thickening of the skin, and underlying soft tissues. This subcutaneous induration of the scleroderma type (thickened, cardboard skin, and impossible to pinch), predominate at the ends, can spread to other parts of the body while respecting the face, gives an appearance of orange peel members reached and venous paths dug embossed or "canyon sign" of pathognomonic of Schulman disease. The musculoskeletal manifestations are frequent in the opening phase (arthritis mono, oligo or polyarticular hands, wrists and knees, early myalgia sometimes severe). Generally it doesn't associate pulmonary, cardiac, neurological or renal disorder with a few exceptions^[2-3].

The pathophysiology of this pathology remains largely unknown, even if certain promoting factors participate in fibrogenesis by a decrease in serum collagenase levels, in particular matrix metalloproteinases-13 (MMP-13)^[4], an increase in inhibitor rates metalloproteinases in tissues^[5] and serum levels of TGF- β , a pro-fibrotic cytokine^[6]. Biology may show hyper eosinophilia in 60 to 90% [1], leukocytosis, inflammatory syndrome, hypogammaglobulinemia and rarely an elevation in serum creatinine phosphokinase (4-10%) most often reflecting myopathymoderate. A rise in aldolase is positive for antinuclear antibodies in 15 to 20%, but without anti-DNA antibody or anti-nuclear soluble antigen antibody^[7-8]. The assay of anti-cytoplasm antibodies of neutrophils is negative and makes it possible to distinguish it from eosinophilic granulomatosis with poly angiitis^[1-2]. Any other abnormality in the hemogram (anemia, thrombocytopenia, pancytopenia) must have an underlying hemopathy (aplastic anemia) eliminated.

The conventional radiology may be normal (no reference in the literature), ultrasound (figure 2) shows a hypoechoic hypodermis (edematous) and a fascia hypoechoic thickness greater than the normal^[3-9-10]. The sonographic appearance had to be characteristic enough to decide to conduct a biopsy including micro-histological analysis to confirm the diagnosis. Also, some studies suggest the use of this method for monitoring patients under treatment and more recently as a measure of skin elastometry^[11]. The CT scan with injection can detect a thickening of the muscular fascia and is particularly interesting for the evaluation of the extension of lesions. MRI (figure 3) has been studied in a few series and seems to be useful for diagnosis and pre-biopsy identification. The minimal acquisition protocol consists of a T1SE sequence and a STIR sequence in the axial plane. The typical appearance before injection of contrast medium is thickening of the fascia's (deep peripheral fascia and less frequently intermuscular fascia) on the T1-weighted sequences, in relative hypersignal compared to the muscles on the T2-weighted sequences with suppression (T2 STIR) or saturation (T2 FS) of the fat signal^[3-7]. The injection of contrast product is not systematic but when it is carried out, it is followed by a T1SE sequence with saturation of the fat signal (T1SE FS) in the axial plane. The achievement is typically bilateral, the two segments members must be explored comparatively^[10-12].

Due to the rarity of this disease, we could not find in the literature a study which describes all the imaging modalities (ultrasound, CT and MRI), However, there are cases mainly focused on resonance imaging magnetic.

FDG positron emission tomography typically shows hyper fixation or hypermetabolism of the fascia, respecting muscle mass and subcutaneous fat. It has the advantage of carrying out in a single examination an exhaustive assessment of the fascia's, of seeking differential diagnoses, a neoplastic cause and of being able to be used in the event of a contraindication to MRI ^[3-13-14]. The skin sample must be deep and involve the skin, the fascia and a little of the underlying muscle. Histological confirmation is always recommended for the definitive diagnosis and to rule out other differential diagnoses (Systemic Scleroderma, Myalgia-eosinophilia Syndrome, Spanish Oil Syndrome, and Churg-Strauss Disease) ^[12]. Typically, pathology examination reveals a thickened fascia with inflammatory infiltrates composed mainly of CD8 + T lymphocytes (CD4 / CD8 ratio <1) ^[15] or in 69 to 75% of cases of eosinophils whose presence is not necessarily essential for diagnosis. Because, they can be absent in the late phase of the disease or shortly after the start of corticosteroid treatment ^[14-16].

The therapeutic management of eosinophilic fasciitis is not well codified, however the standard treatment remains general high-dose corticosteroid therapy (0.5 to 1 mg / kg) for a duration greater than 6 months and close to from 24 to 31 months on average ^[16-18] in the first line or secondarily in associated action to an immunosuppressive (methotrexate, azathioprine, cyclophosphamide, cyclosporine and more recently biotherapy) in case of an elevated cortico-dependence or initial corticosteroid resistance. A physiotherapy with flexible mobilization and soft skin is recommended to reduce stiffness and joint limitations. The interest of other therapeutic do to be evaluated case by case according to the related pathologies, in particular hematological ^[18]. The prognosis is good in general, is the disease is not associated a severe blood disease, one we observe at least partial regression of all symptoms in most cases ^[19-20].



Figure 1:- Front and profile photograph of the two legs (a and b), moderately erythematous with appearance of orange peel of the 60-year-old patient followed for Schulman's disease.

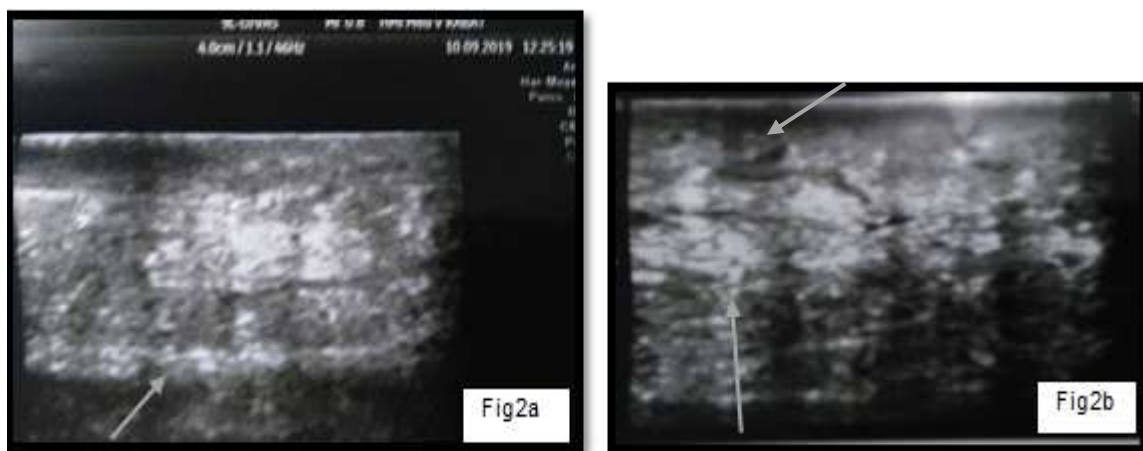


Figure 2:- Same patient, longitudinal ultrasound section of the legs, in favor of a hypoechoic hypodermis (edematous) and increase in the thickness of the subcutaneous tissues.

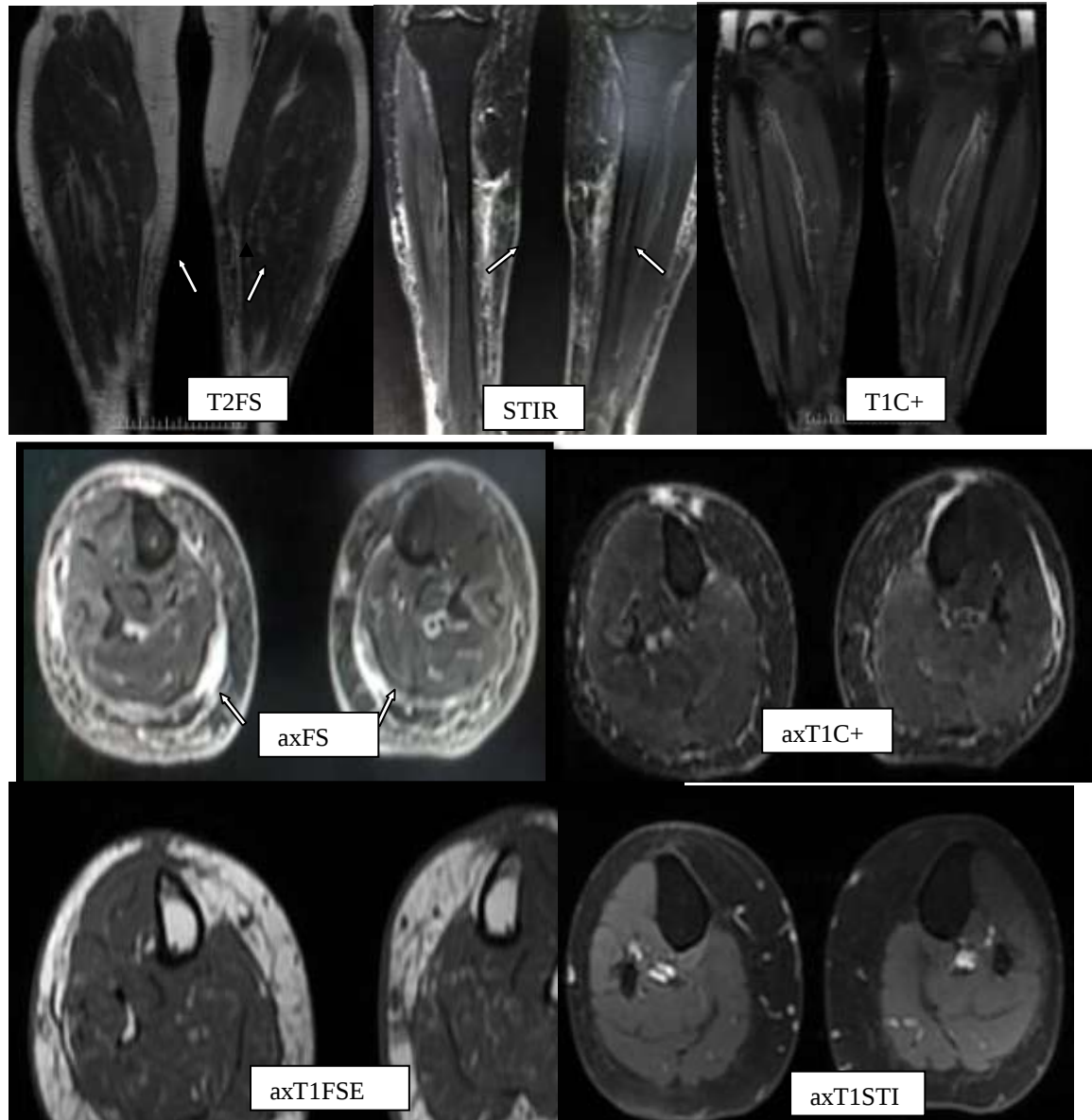


Figure 3:- MRI of the lower limbs coronal and axial slices, in favor of a discreet thickening of the fascia's of the two legs with signal abnormality in T2 STIR in iso signal T1FSE, a slight enhancement at an early time after injection of Gadolinium associating a relative hypersignal of the muscles of leg on sequences with removal in FS.

Conclusion:-

Schulman's disease is a rare entity that can be disabling. The diagnosis is discussed on arguments clinical, biological and the picture series by a thickening of the fascia with a contrast enhancement in MRI. It is certified by the presence of an inflammatory infiltrate of the fascia on the analysis histological. In principle, this syndrome has a good prognosis and responds well to treatment with steroids alone or in combination with immunosuppressants

Conflicts of interest:

The authors declare no conflict of interest

Authors contributions:

Aaron ICKAMBA HOUCI: design of the initial manuscript and writing of the final study manuscript
Wilson BIZIMANA: data collection and bibliographic references of the project

Raïssa KAUKONE: image formatting

Abdellatif DARBI: team supervision and project monitoring

Bouchaib RADOUANE: validation, proofreading and revisions of the final manuscript

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