Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/10290
DOI URL: http://dx.doi.org/10.21474/IJAR01/10290

RESEARCH ARTICLE

A RARE OFUJI PAPULO-ERYTHRODERMA REVEALING A LANGERHANS CELL HISTIOCYTOSIS INVOLVING ONLY LYMPH NODES

Nafissa Farid, Said Amal and Ouafa Hocar
Department of Dermatology, Mohammed VI University Hospital Center, Cadi Ayyad University, CHU Mohammed VI BP 2360 Principal Avenue Ibn Sina, Marrakech, Morocco.

Manuscript Info

Manuscript History
Received: 23 November 2019
Final Accepted: 25 December 2019
Published: January 2020

Key words:-
Langerhans Cell Histiocytosis – Lymphnodes - Ofuji papuloerythroderma

Abstract

Ofuji Papulo-erythroderma is a rare entity. Its origin is most often paraneoplasic: visceral neoplasias and blood diseases are the most frequently found. We report a case of Ofuji papuloerythroderma revealing langerhans cell histiocytosis involving only lymph nodes.We report the case of a 48-year-old man who presented a pruriginous papuloerythroderma, respecting skin folds, having progressed for 8 years in a context of deterioration of the general state, associated with bilateral inguinal lymphadenopathies. The patient had already had several skin biopsies that were inconclusive. The biological assessment revealed an inflammatory syndrome and a discreet hyper-eosinophilia. Lymph node biopsy confirmed the diagnosis of Langerhans cell histiocytosis. Clinical and imaging studies did not reveal abnormalities in other organs, concluding to isolated lymph node involvement by Langerhans cell histiocytosis. The patient received chemotherapy by vinblastine and corticosteroids with very good clinical progress. Lymph node involvement by Langerhans cell histiocytosis is rarely described in the literature, it is most often located in the drainage area of a cutaneous or bone lesion, and it is rarely isolated. Ofuji's papulo-erythroderma is a rare entity, most often paraneoplastic and exceptionally associated with Langerhans cell histiocytosis, which needs a very thorough etiological assessment to determine the underlying etiology.

Introduction:-

Ofuji papulo-erythroderma (OPE) was first described in 1984 by Ofuji and al. (Ofuji et al. 1984) They reported four cases of otherwise healthy elderly male patients affected by erythroderma which featured flat-topped papules and a characteristic sparing of the skin folds, laterly defined as 'deck-chair sign' (Torchia D et al. 2010). Initially it has been reported in Japan, but the literature has been enriched by punctual observations, mainly European ones. Nevertheless, Papuloerythroderma of Ofuji is probably still misinterpreted, underrecognized and therefore underdiagnose (Thomsen, 1998).

Langerhans cell histiocytosis (LCH) is a rare disease, more common among children, that can involve a number of anatomical locations especially bones, skin, neurohypophysis and others. Lymph node involvement by LCH without

Corresponding Author:- Nafissa Farid
Address:- Department of Dermatology, Mohammed VI University Hospital Center, Cadi Ayyad University, CHU Mohammed VI BP 2360 Principal Avenue Ibn Sina, Marrakech, Morocco.

other sites of disease is rarely described in the literature, and its association with Ofujipapulo-erythroderma is exceptional.

We report a case of Ofujipapulo-erythroderma revealing a Langerhans cell histiocytosis involving only lymph nodes without other location.

Case Report:-
A 48-year-old man presented a prurticpapulo-erythroderma that had been evolving for 8 years, with progressive worseningin a context of alteration of the general state. The patient had several skin biopsies before that were inconclusive.

Physical examination showed a dry erythroderma, sparing the skin folds, with a slightly lichenified aspect in the back and excoriated lesions of scratching. We noted also the coalescence of flat solid papules in the legs (fig. 1). This erythroderma was associated to a discreet palmar keratoderma and bilateral inguinal lymphadenopathy measuring 2cm of long axis for the largest one, without hepatomegaly or splenomegaly. The patient had also vitiligo lesions on both forefeets before the rash happened.

Biologic investigations showed a discrete hyper-eosinophilia at 660 / mm3 associated with an inflammatory syndrome made of an accelerated VS at 42 mm in the first hour and 80 mm in the second hour, in addition to an inflamation profile in the electrophoresis of plasma proteins. Tumor markers were normal. The research of Sezary cells was negative. The rest of routine laboratory tests, HIV and hepatitis serologies were normal.

Radiological assessment showed normal chest x-ray and abdominopelvic ultrasound. The CT-scann revealed bilateral inguinal and external iliac lymphadenopathy, the largest measured 1,8 cm, without other abnormalities.

Skin biopsy was done at two different sites. In the back, it showed subacute and chronic nonspecific dermatitis. In infiltrated papular lesions of the leg, skin biopsy was also nonspecific, revealing subacute dermatitis with dense lymphoid infiltrate.

Since skin biopsies were inconclusive, we completed the investigations by a lymph node biopsy which objectified a histiocytic adenitis made of a pulp massively occupied by a proliferation of histiocytic cells. Immunohistochemical studies for CD1a and S100 protein were positive, concluding to a langerhans cell histiocytosis (fig.2).

X-ray of chest, long bones, skull, spine and a brain Magnetic resonance imaging were done in order to search other locations of LCH. All of the tests were normal, concluding to an isolated lymph node involvement by Langerhans cell histiocytosis.

The patient received chemotherapy combining vinblastine and corticosteroids according to a protocol of 42 weeks of treatment divided into attack phase and maintenance phase. The attack phase lasted six weeks and included six weekly cures of vinblastine at a dose of 6 mg / m² associated with oral corticosteroid therapy at a dose of 1 mg / kg / day. The maintenance phase lasted 36 weeks and included an injection of vinblastine followed by 5 days of steroids to be repeated every three weeks.

The clinical evolution was good since the first weeks of treatment. It was marked by a regression of erythroderma, a disinfiltration of papular cutaneous lesions and a regression of pruritus (fig.3).

Discussion:-

In our case, Ofujipapulo-erythroderma (OPE) revealed a LCH involving only lymph nodes wich is very rare.

OPE is defined by association of clinical, biological and histological signs. Initially, it was reported in Japan, but the literature has been enriched by punctual observations, mainly european ones. Nevertheless, it's probably under-estimated and underdiagnosed because of its recent identification and the poor awareness of OPE among clinicians. [4] It is mainly observed in patients over 55years, and more often in males (Thomsen, 1998).

The lesions are typically papular, brown-red, very pruriginous and localizated mainly on the limbs and trunk. They spare the bottom of the big folds (inguinal, axillary ...) as well as the abdominal and dorsal skin folds, giving a very characteristic appearance called « deck-chair sign » (Farthing et al.,1986)

The eruption evolves gradually over a few months to end by coalescence of the papules to an extensive eruption, but it's rarely a typical erythroderma. OPE Evolution is chronic, and the face is often respected. Palmo plantar keratoderma can be associated with OPE in only 20% of cases.

In general, there is no hyperthermia and the deterioration of the general state, when it exists, is the consequence of chronic pruritus, sometimes insomniac and rebellious to symptomatic treatments. Peripheral eosinophilia is found in more than 85% of cases. Lymphocytopenia and increased serum IgE are less frequent.

Histopathology of skin lesions is not very specific. It showes most often features of chronic dermatitis ; variable amounts of epidermal hyperplasia with occasional spongiosis and exocytosis and a mixed inflammatory dermal infiltrate mainly composed by memory T cells, macrophages, eosinophils and Langerhans cells, without atypical cells(Thomsen, 1998).Immunohistochemistryshows an important infiltrate of Langerhans cells and CD4 + T lymphocytes. Histopathology of lymph node biopsy, when it's realized, shows an aspect of dermopathic lymphadenopathy. The pathophysiology remains unknown. Several authors have been confused whether Ofuji'spapulo-erythroderma is a separate nosological entity or a clinical manifestation of an underlying pathology (Torchia D et al., 2010; Bagot and Revuz, 1992). The pathologies most commonly associated with papulo-erythroderma are neoplasias including solid cancers (stomach, lung, colon…) and blood diseases (lymphoma, leukemia, hodgkin'sdisease ..). It can also have an infectious origin, mainly secondary to HIV or viral hepatitis. A few cases of drug-induced papulo-erythroderma have been reported. In some cases no etiology will be identified, and it will be said idiopathic, requiring regular monitoring.

Given the rarity of OPE, no studies with a high level of evidence are available for treatment modalities of PEO. The management of this chronic and resistant rash should bebased principally on the search and treatment of any underlying cause, and on the regimens reported in the literature(Thomsen, 1998).

Among the treatments that have been effective in Ofujipapulo-erythroderma are found dermocorticoides, general corticotherapy, and finally the PUVAtherapy, prescribed alone or in association with one of the two above-mentioned treatment, can also be effective (Mutluer et al.,2004).

After a review of the literature, no cases of Langerhans cell histiocytosis were revealed by Ofujipapulo-erythroderma.

LCH is a rare disease, more common among children and adolescents. It is a pathology with multivisceral development and very diverse clinical manifestation. The diagnosis is histopathological. It results from the infiltration of one or more organs by dendritic langerhans cells, most often organized into granulomas. It can involve a number of anatomical locations especially bones, skin, neurohypophysis, lungs, lymphoid organs and more rarely other sites like liver and digestive system (Neel et al., 2015).The cutaneous involvement is frequent, and consists of a red macular or papular rash of orange-yellow or brown color, comonly on the trunk and large. Lesions could be extensive.

Lymphnode involvement by LCH is another site that has also been described in the literature (Morgenfeld and Schajowiicz,1971 ; Williams and Dorfman,1979). LCH involving lymph nodesusually occurs in patients in the pediatric age group with known systemic disease, most often in the drainage area of a skin or bone lesion. However, rarely, LCH can primarily involve lymph nodes without other sites of disease. M. Edelweiss and al. presented 20 cases of LCH restricted to lymph node, with no other sites of disease (Edelweiss et al., 2007)

R. Ruiz-Villaverde and al.(2014) described a case of LCH restricted to lymph nodes, revealed by erythroderma, initially taken for drug-induced rash. The diagnosis was corrected after lymph node biopsy concluding to LCH with positive immunohistochemistry of S100 and CD1a protein, like the case of our patient.

When LCH is diagnosed, an assessment of extension has to be done in order to search other localisations of the disease. Treatments are as varied as clinical forms and indications depend on sites and number of locations.

Conclusion:-

Ofuji'spapulo-erythroderma is a very rare entity that requires a very thorough etiological assessment to determine the underlying etiology that is more frequently neoplasias. Langerhans cell histiocytosis is heterogeneous in its clinical presentation and often capricious in its evolution. It can be revealed by erythroderma, or rarely by papulo-erytherma. Lymph node involvement is rarely isolated and must seek a secondary cause first.

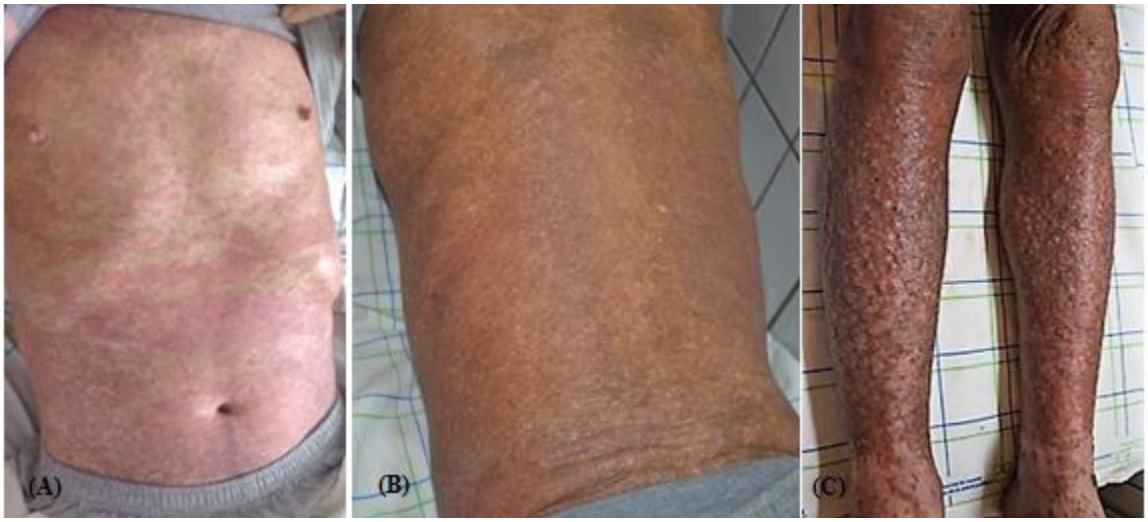

Fig. 1:- Dry erythrodermasparing the skin folds (A), with a slightlylichenified aspect in the back (B), and coalescence of flat solid papules in the legs (C) .

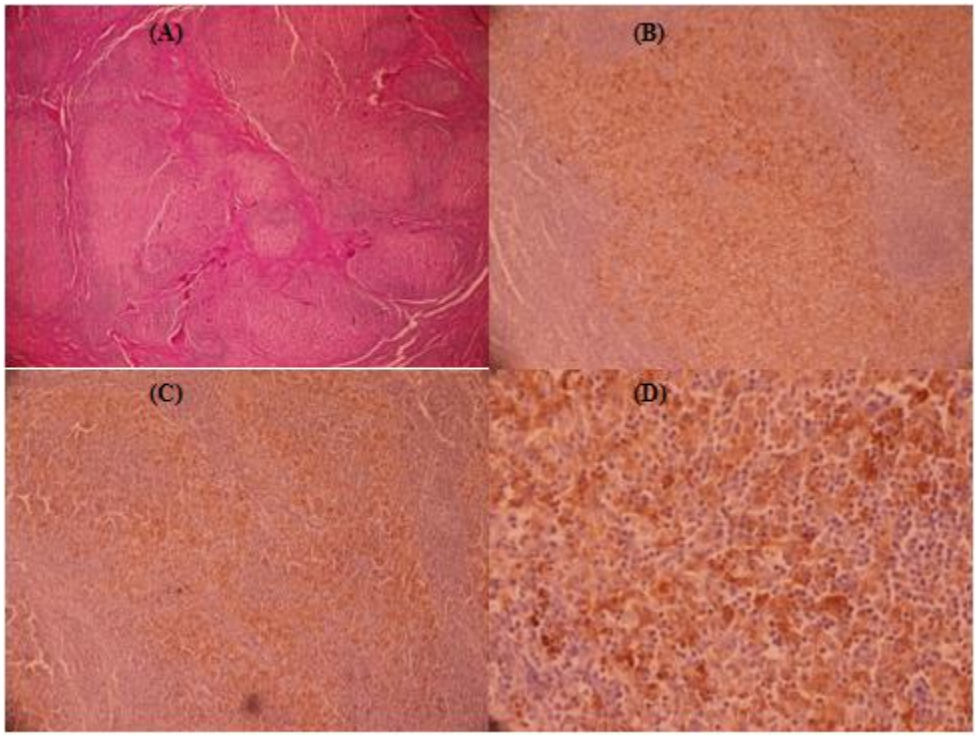

Fig. 2:-Histopathology and immunohistochemistryexamination of the lymphnoderevealing a Langerhans cellhistiocytosis. A : histiocyticadenitis made of a pulpmassivelyoccupied by a proliferation of histiocyticcells, B :important expressionof anti-protéin S100, C et D :important expressionof anti CD1a.

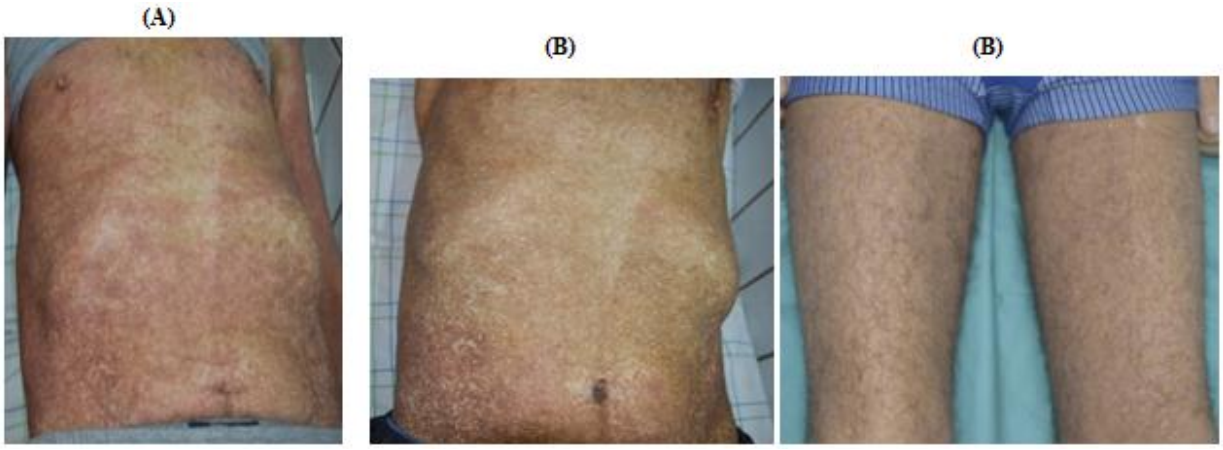

Fig. 3:- Clinical evolution under treatment. A : after 8 weeks of treatment, B : after 16 weeks of treatment.

References:-

1. Bagot and Revuz (1992). Faut-il individualiser la papulo-érythrodermie d'Ofuji?Ann Dermatol Venerol, 119: 628.
2. Farthing et al. (1986). Papuloerythroderma– a further case with the 'deck chair sign'. Dermatologica, 172:65-66.
3. Morgenfeld and Schajowicz (1971). Solitary eosinophilic granuloma of lymphnode: five-year follow-up. Pediatrics, 48:301- 5.
4. Mutluer et al.(2004). Treatment of papuloerythroderma of Ofuji with Re-PUVA : a case report and reviewtherapy. J EurAcadDermatolVenereol, 18:480-483.
5. Néel et al. (2015). Histiocytose langerhansienne de l'adulte. La Revue de médecine interne, 36:658–667.
6. Ofuji et al. (1984). Papuloerythroderma. Dermatologica, 1984, 169:125–130.
7. Thomsen (1998). Ofuji'spapulo erythroderma: a study of 17 cases. Clin Exp Dermatol, 23: 79–83.
8. Torchia et al. (2010). Papuloerythroderma 2009 : two new cases and systemic review of the world wide literature 25 years after its identification by Ofuji et al. Dermatology, 220:311-320.
9. Williams and Dorfman (1979). Lymphadenopathy as the initial manifestation of histiocytosis X. Am J Surg Pathol, 3:405-21.
10. Edelweiss et al. (2007). Lymph node involvement by Langerhans cell histiocytosis: a clinico pathologic and immunohistochemical study of 20 cases. Human Pathology, 38:1463– 1469.
11. Ruiz-Villaverde et al. (2014). Erythroderma as an Initial Presentation of Langerhans Cell Histiocytosis Involving the Sinus. Actas Dermo-Sifiliográficas (English Edition), 105(Issue 6):630-2.