IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 22, Issue 8 Ser.3 (August. 2023), PP 56-59
www.iosrjournals.org

Pulmonary Involvement Revealing Weber Christian Disease

F.AitAhmed, M.A.Messaoudi ,N.Sarghini, I.Oudergea, H.Charaf, R.Zahraoui, M.Soualhi, J.E.Bourkadi

Pneumology department of Moulay Youssef Hospital, Ibn Sina University Hospital, Rabat, Morocco

Date of Submission: 02-08-2023 Date of Acceptance: 12-08-2023

I. Introduction

Weber Christian disease (WCD) or idiopathic lobular panniculitis is a rare condition of unknown etiology, characterized by recurrent inflammation of subcutaneous adipose tissue, it mainly affects adults with female predominance [1]. WCD is a nonspecific clinical entity that remains a subject of debate and its evolution is unpredictable, it is a diagnosis of exclusion. It manifests clinically with recurrent fever outbreaks and painful subcutaneous nodules, systemic manifestations of WCD frequently affects the liver, bone, kidneys, sporadically serous, spleen and lungs.

It could be life-threatening and it is considered a therapeutic challenge for clinicians.

We report a case of Weber Christian disease diagnose at the pulmonology department during the exploration of respiratory pathology

II. Case report

A 50-year-old man, former weaned smoker, type II diabetic under insulin, having as antecedents an acute pancreas stage E and a superficial venous thrombosis, which goes back to 4 months. The patient reports one week before his hospitalization a productive cough producing muco-purulent sputum associated with thoracic pains, a dyspnea of effort stage II mMRC and an intermittent fever quantified to 39°C.

The thoracic CT scan showed sub pleural nodules of the ventral segment of the right upper lobar and atelectasis in the middle lobe and dorsal segment of the left lower lobe associated to septal and non septal thickening, multiple hilo-mediastinal nodes and homogeneous hepatomegaly without suspicious lesions. (Figure 1-2-3)

The biological assessment showed an inflammatory anemia at 8g/dL, the erythrocyte sedimentation rate was 100mm with a C-reactive protein (CRP) at 400mg/L. The immunological assessment and viral serologies were negative. The hepatic, renal, lipid test, 24-hour proteinuria and lipasemia tests were normal. Pulmonary tuberculosis screening was negative. Echocardiography and abdominal ultrasound were normal.

During his hospitalization, the patient presented erythematous swelling on the back of both hands (figure 4), and painful subcutaneous nodules on these limbs. One week later, the patient developed a sudden scrotal edema, which the scrotal ultrasound showed to be bilateral epididimo-orchitis. The evolution was marked by installation 4 days later, of a painful left palpebral edema associated with a ptosis (figure 5-6). The orbital CT scan with contrast revealed a diffuse thickening of the preseptal soft tissues at the level of the medial canthus and an infiltration of intra and extra conical fat in both eyes (figure 7 A, B).

The biopsy of the subcutaneous nodule of the right leg showed lesions of lobular panniculitis made of a polymorphic inflammatory infiltrate of low density, with the presence of polynuclear neutrophils evoking the panniculitis of Weber Christian (fig 8 A, B).

The diagnosis of Weber-Christian disease with multisystem involvement was made.

The patient had ceftriaxone (2g/jour) and Ciprofloxacin (500 mg*2/jour) during 15 days, with methylprednisolone during 3 days.

The evolution was marked by apyrexia, disinfiltration of lesions, regression of ocular and scrotal edema

III. Discussion

Weber-Christian disease (WCD) as originally defined by Christian in 1928 (relapsing febrile nodular non suppurative panniculitis) is a rare inflammatory illness of unknown etiology, this disorder was characterized by recurrent attacks of fever associated with the development of subcutaneous tender nodules (particularly over the extremities). The diagnosis of Weber-Christian panniculitis is confirmed by histological examination of a subcutaneous nodule, that shows pleomorphic inflammatory cellular infiltration of the subcutaneous adipose

tissue and interlobular septa[2] .Systemic WCD frequently affects the liver(hepatomegaly), kidneys(hematuria , proteinuria) ,the bone marrow, and sporadically the lungs,heart,serosa,spleen,and intestines[3].Systemic manifestations such as recurrent fever episodes, fatigue, myalgia and polyarthralgia are also commonly encountered[4-5].

Respiratory involvement may be clinically manifested by cough, dyspnea or chest pain. Mediastinitis, pleuritis, and pleural effusion were rarely described [6-7-8].

Pulmonary nodules associated with WCD are also described whose histological examination is characterized by foam cell infiltration in the alveolar spaces and lymphocytes in the endobronchitis [9].

Corticosteroids are the mainstay in the treatment of WCD, although other immunosuppressive agents such as cyclosporineA ,cyclophospamide, and mycophenolate mofetil have been tried with variable results[10,11-12]. Recently, infliximab has been used to treat idiopathic lobular panniculitis and satisfactory results were achieved.

In the case of our patient, the expression of the disease was multisystemic involving, inflammation of lungs, skin, inflammatory orbitopathy, pancreatitis, coagulopathy and anemia, he was put on corticosteroids at a dose of 0,75 mg/kg/day with good clinical progression.

IV. Conclusion

Weber Christian disease is a rare inflammatory disease of unknown etiology. The diagnosis can only be made after eliminating other causes of lobular panniculitis. The prognosis is determined by visceral lesions. More studies are needed for a better understanding of this multisystemic disease.

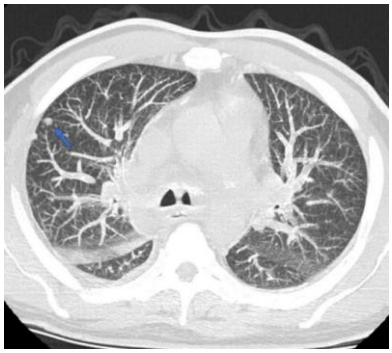

Figure1: Axial section of a parenchymal windowed chest CT with MIP reconstruction, showing subpleural nodules of the ventral segment of the LSD

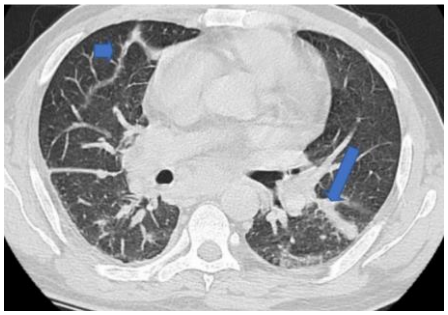

Figure2: Axial parenchymal window CT section, showing bands of atelectasis in the middle lobe and dorsal segment of the left lower lobe with associated septal and nonseptal thickening

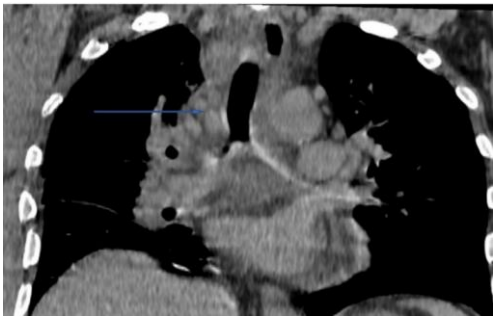

Figure3: Chest CT in coronal section, mediastinal window, showing multiple hilo-mediastinal nodes

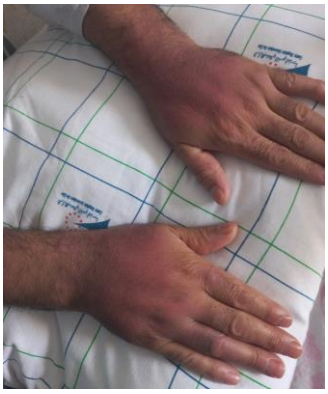

Figure 4 : picture showing tender erythematous swellings on the back of both hands

Figure 5 : picture showing left upper eyelid swelling and erythema

Figure 6 picture showing right upper eyelid swelling

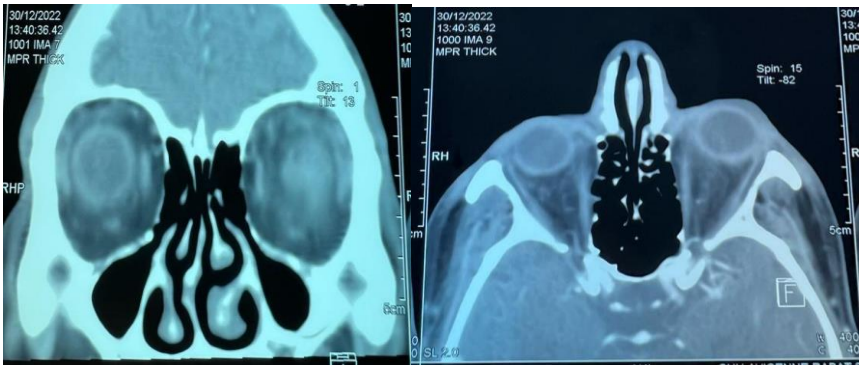

Figure 7 A **Figure 7 B**

Figure 7 A – B : Orbital CT scan revealed diffuse thickening of the preseptal soft parts at the level of the inner canthus and infiltration of intra and extra conical grease in both eyes

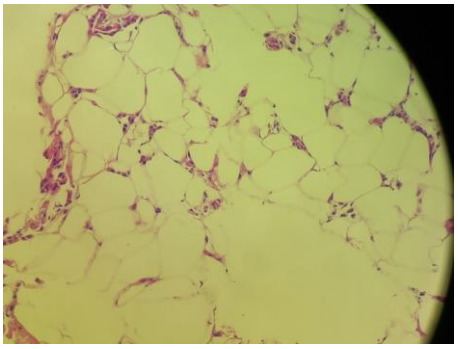

Figure 8 A

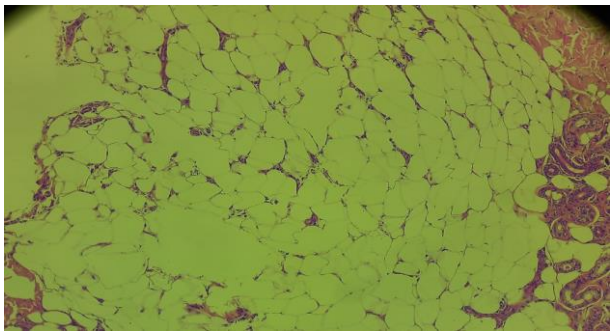

Figure 8 B

Figure 8 A et B : subcutaneous nodule of the right leg showed lesions of lobular panniculitis made of a polymorphic inflammatory infiltrate of light density , with neutrophil polynuclear.

Reference

[1]. Mathai R. IADVL Textbook And Atlas Of Dermatology. 2nd Ed. 2001. Disorders Of Subcutaneous Fat; Pp. 944–5. [Google Scholar]
[2]. Ungar, H. (1946). J. Path. Bact., 58, 175.
[3]. Miranda-Bautista J, Fernandez-Simon A, Perez-Sanchez I, Menchen L. Weber- Christian Disease With Ileocolonic Involvement Successfully Treated With Inflix- Imab. World J Gastroenterol. 2015;21:5417-5420
[4]. Yoshimura A, Itoyama S, Mori S, Mori W, Inoue G. An Autopsy Case Of Weber- Christian With Immune Complex Glomerulonephritis. Nihon Jinzo Gakkai Shi. 1988;30:291-296
[5]. Başkan EB, Sarıcaoğlu H, Tunalı Tolunay ŞŞ. Effective Treatment Of Relapsing Idiopathic Nodular Mycophenolate Mofetil Effective Treatment Of Relapsing Idio- Pathic Nodular Panniculitis (Pfeifer—Weber—Christian Disease) With Mycophe- Nolate Mofetil. J Dermatolog Treat. 2003;14:57-60.
[6]. Hirasaki S, Murakami K, Kanamori T, Mizushima T, Hanayama YKN. Weber- Christian Disease Developing Into Mediastinitis And Pleuritis With Massive Pleural Effusion. Intern Med. 2015;51:943-947
[7]. Wang Y, Zhao J, Ji LI, Zhang SZZ. Weber-Christian Disease Present With Lung Nodules Dramatically Improved With Corticosteroid Therapy: One Case Report And Literature Review. Int J Rheum Dis. 2018;21:573-578.
[8]. Achten G, Moriame-Roussel N, Wanet J, De Coninck J, Vertommen J. Idio- Pathic Liquefying Panniculitis With Fatal Outcome. Ann Dermatol Venereol. 1977;104:693-696.
[9]. Pinals RS. Nodular Panniculitis Associated With An Inflammatory Bone Lesion. Arch Dermatol. 1970;101:359-363.
[10]. Baskan EB, Saricaoglu H, Tunali S, Et Al. Effective Treatment Of Relapsing Idiopathic Nodular Panniculitis (Pfeifer-Weber-Christian Disease) With Mycophenolate Mofetil. J Dermatolog Treat. 2003;14(1):57—60
[11]. Wasaki T, Hamano T, Ogata A, Et Al. Successful Treatment Of A Patient With Febrile, Lobular Panniculitis (Weber-Christian Disease) With Oral Cyclosporin A: Implications For Pathogenesis And Therapy. Intern Med. 1999;38(7):612—4
[12]. Kirch W, Duhrsen U, Hoensch H, Et Al. Cyclophosphamide-Induced Remission In Weber-Christian Panniculitis. Rheumatol Int. 1985;5(5):239--40