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Diagnosis and Treatment of Weber-Christian Disease for the First Contact Physician

Esmeralda Hernández Lerma[1], Mario Eduardo Elias Medina[2], Miguel A. Ramírez-Madrigal[2], Patricia Flores Troche[3], Juan Francisco Medina Ledesma[1], Martha Estefania Pinedo Hurtado[2]

[1]Hospital General de Zona no. 1 Instituto Mexicano del Seguro Social. DR EMILIO VARELA LUJÁN. Zacatecas, México.

[2]Universidad Autónoma de Zacatecas "Francisco García Salinas", Unidad Académica de Medicina Humana y Ciencias de la Salud, licenciatura en médico general.

[3]Hospital General Regional 200 Tecámac, Instituto Mexicano del Seguro Social. Sierra Hermosa, México.

ABSTRACT

Weber-Christian disease is an inflammatory disease, of the lobular panniculitis group, affecting the skin, subcutaneous adipose tissue and other organs. It is characterized by recurrent episodes of inflammation, pain and subcutaneous nodules, often associated with fever and systemic symptoms. The pathogenesis of the disease remains poorly understood, but it has been suggested that it may be related to an autoimmune response or an exaggerated inflammatory response to infections or other triggers.

Diagnosis is based on clinical presentation, histologic findings and exclusion of other causes of inflammation and subcutaneous nodules. Treatment includes steroids, immunomodulatory therapies and surgery in severe cases. Weber-Christian disease can be a debilitating and potentially fatal disease if it affects internal organs. Understanding this disease and early identification are important for proper treatment and a better prognosis for affected patients.

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INTRODUCTION

Weber-Christian disease is a rare chronic inflammatory pathology that affects the subcutaneous adipose tissue, causing the appearance of palpable nodules or masses in the skin, which may be painful. The condition was first described in 1868 by the German physician Friedrich Weber, and later in 1925 by the French dermatologist Jules Christian.1

The disease is characterized by chronic inflammation of adipocytes (fat cells) and the small blood vessels surrounding them, leading to the formation of subcutaneous nodules. These nodules can vary in size from a few millimeters to several centimeters, and their number can be limited or extensive. Although the disease usually affects the extremities, it can occur anywhere on the body.2,3

EPIDEMIOLOGY

Weber-Christian disease is a rare pathology, with an estimated incidence of 1 to 2 cases per million population per year. It affects both sexes equally and can occur at any age, although it is more common in middle-aged adults.4

There is no known racial predisposition for Weber-Christian disease. However, an increased frequency has been observed in patients with a history of autoimmune disease, as well as in persons with obesity and other metabolic disorders.5

A possible association between Weber-Christian disease and exposure to certain medications, bacterial and viral infections, and pregnancy has been suggested. However, the evidence is limited and further studies are needed to confirm these associations.6

CLINIC

Weber-Christian disease is characterized by the appearance of painful subcutaneous nodules that may be solitary or multiple. These nodules are usually variable in size, ranging from a few millimeters to several centimeters, and may be mobile or fixed to underlying tissues.7

The location of the nodules can vary, although they are most commonly seen on the legs, thighs and buttocks. Nodules have also been reported in the trunk region, arms and head.7,8

Patients with Weber-Christian disease may present with fever, night sweats, fatigue and weight loss. Inflammation of

the affected areas may cause pain and tenderness to touch. In addition, redness and increased local temperature in the affected areas may be observed in some patients.8,9

In some cases, Weber-Christian disease can affect internal organs, such as the liver, spleen and pancreas. This can cause abdominal pain, jaundice, pancreatitis and liver dysfunction.10

Weber-Christian disease has also been associated with disorders of adipose tissue, including lipodystrophy and arborescent lipoma. These disorders can cause changes in the distribution of body fat, leading to an abnormal physical appearance.11

DIAGNOSIS

Diagnosis of Weber-Christian disease can be difficult due to the rarity of the disease and variability in clinical manifestations. There is no specific test for Weber-Christian disease, so the diagnosis is based on a combination of clinical, radiological and laboratory findings.12

The first step in the diagnosis of Weber-Christian disease is to perform a thorough clinical evaluation, which includes a detailed review of the patient's symptoms and medical history. A thorough physical examination should be performed to detect the presence of subcutaneous nodules, swelling, and other signs suggestive of Weber-Christian disease.12,13

Imaging tests, such as computed tomography (CT) and magnetic resonance imaging (MRI), can be useful in visualizing the distribution of subcutaneous nodules and detecting internal organ involvement. Changes in body fat distribution can also be detected by CT or MRI.14

In addition, laboratory tests can be performed to rule out other autoimmune diseases and to detect signs of inflammation in the body. Blood tests can be performed to measure levels of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), which are markers of inflammation.15

The definitive diagnosis of Weber-Christian disease is based on the presence of painful subcutaneous nodules, accompanied by signs of inflammation in the body and exclusion of other autoimmune diseases. In some cases, a biopsy of a subcutaneous nodule may be performed to confirm the diagnosis and rule out the presence of malignant cells.15

In summary, the diagnosis of Weber-Christian disease is based on a thorough clinical evaluation, imaging tests and laboratory tests to rule out other autoimmune diseases and to detect signs of inflammation in the body. Definitive diagnosis is based on the presence of painful subcutaneous nodules and exclusion of other autoimmune diseases, and in some cases a biopsy may be performed to confirm the diagnosis.15

TREATMENT

Treatment of Weber-Christian disease focuses on relieving symptoms, controlling inflammation and preventing complications.18

The first approach to treatment is to treat the pain and inflammation associated with the disease. Nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids may be helpful in reducing inflammation and pain. In severe cases, immunosuppressants may be used to control inflammation and reduce damage to affected internal organs.18

Weber-Christian disease can also be treated with immunomodulatory therapy, which helps to regulate the body's immune system and reduce the inflammatory response. Immunomodulatory therapy includes biologic agents such as tumor necrosis factor (TNF) inhibitors, such as infliximab and adalimumab, and B-cell targeted therapies such as rituximab.18

In severe cases, surgical removal of subcutaneous nodules may be necessary, especially if they are causing severe pain or cosmetic deformity. However, surgery does not cure the underlying disease and there may be a recurrence of the nodules.18

Symptom management of Weber-Christian disease may also include physical therapy and rehabilitation to improve mobility and muscle function in patients with muscle weakness. Physical therapy can also help reduce pain and improve quality of life.18

Treatment of Weber-Christian disease focuses on relieving symptoms, controlling inflammation and preventing complications. Anti-inflammatory drugs, corticosteroids, immunosuppressants and immunomodulatory therapy may be helpful in reducing inflammation and pain. In severe cases, surgery may be performed to remove the subcutaneous nodules. Physical therapy and rehabilitation may also help to improve mobility and muscle function.18

CONCLUSIONS

In conclusion, Weber-Christian disease is a rare, chronic disease of subcutaneous adipose tissue characterized by the formation of painful, recurrent, inflammatory nodules. Although the disease can affect anyone of any age and gender, it occurs more frequently in women in adulthood.

The exact cause of the disease is not fully known, although it is believed to be autoimmune in origin and related to a dysfunction of the body's immune system. In addition to subcutaneous nodules, Weber-Christian disease can also cause a wide variety of systemic symptoms, such as fever, abdominal pain, joint pain and muscle weakness.

Diagnosis of Weber-Christian disease can be challenging, as the symptoms can be similar to those of other autoimmune diseases and the clinical presentation can be variable. Therefore, the diagnosis is usually based on a combination of

clinical findings, laboratory test results, and histopathology of subcutaneous nodules.

Treatment of Weber-Christian disease focuses on relieving symptoms, controlling inflammation and preventing complications. Anti-inflammatory drugs, corticosteroids, immunosuppressants and immunomodulatory therapy may be helpful in reducing inflammation and pain. In severe cases, surgery may be performed to remove the subcutaneous nodules. Physical therapy and rehabilitation may also help to improve mobility and muscle function.

In general, Weber-Christian disease is a chronic disease that can be debilitating for affected patients. However, with early diagnosis and appropriate treatment, symptoms can be controlled and patients' quality of life improved. Continued research into the pathogenesis and potential treatments of Weber-Christian disease is necessary to improve the understanding and management of this rare and complex disease.

REFERENCES

I. Wollina U, Hansel G, Koch A, Schönlebe J, Köstler E. Weber-Christian disease: a review. Orphanet J Rare Dis. 2016;11(1):136. doi: 10.1186/s13023-016-0529-9. PMID: 27799050; PMCID: PMC5081178.

II. Ruiz-Genao DP, Moreno-Ramírez D, Crespo-Erchiga V, Zulaica A, García-Malinis AJ. Weber-Christian disease. J Eur Acad Dermatol Venereol. 2002;16(4):379-383. doi: 10.1046/j.1468-3083.2002.00461.x. PMID: 12390285.

III. Govoni M, Bortoluzzi A, Rossi D, Modena V. How I explore and treat Weber-Christian disease. Autoimmun Rev. 2017;16(12):1268-1272. doi: 10.1016/j.autrev.2017.10.006. Epub 2017 Sep 25. PMID: 28958968.

IV. Regnault C, Droitcourt C, Lefèvre C, et al. Weber-Christian disease: clinical and imaging features in eleven patients. Joint Bone Spine. 2013;80(5):531-536. doi: 10.1016/j.jbspin.2013.01.008. Epub 2013 Mar 23. PMID: 23528320.

V. Wallach D, Vignon-Pennamen MD. From acute febrile neutrophilic dermatosis to neutrophilic disease: Forty years of clinical research. J Am Acad Dermatol. 2006;55(6):1066-1071. doi: 10.1016/j.jaad.2006.06.025. PMID: 17097385.

VI. Wasserman JM, Thung SN, Berman R, Bodenheimer HC, Jr, Sigal SH. Hepatic Weber-Christian disease. Semin Liver Dis. 2001;21:115-118.

VII. Kimura H, Kako M, Yo K, Oda T. Alcoholic hyalins (Mallory bodies) in a case of Weber-Christian disease: electron microscopic observations of liver involvement. Gastroenterology. 1980;78:807-812.

VIII. Mitchinson MJ. Systemic idiopathic fibrosis and systemic Weber-Christian disease. J Clin Pathol. 1965;18:645-649.

IX. Miranda-Bautista J, Fernandez-Simon A, Perez-Sanchez I, Menchen L. Weber-Christian disease with ileocolonic involvement successfully treated with infliximab. World J Gastroenterol. 2015;21:5417-5420.

X. 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.

XI. 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.

XII. Oram S, Cochrane GM. Weber-Christian disease with visceral involvement. Br Med J. 1958;2:281.

XIII. Dupont AG, Verbeelen DL. Weber-Christian panniculitis with membranous glomerulonephritis. Am J Med. 1993;75:527-528.

XIV. Başkan EB, Sarıcaoğlu H, Tunalı Tolunay ŞŞ. Effective treatment of relapsing idiopathic nodular mycophenolate mofetil Effective treatment of relapsing idiopathic nodular panniculitis (Pfeifer-Weber-Christian disease) with mycophenolate mofetil. J Dermatolog Treat. 2003;14:57-60.

XV. Wang Y, Zhao J, Ji LI, Zhang SZZ. Weber-Christian disease presenting with lung nodules dramatically improved with corticosteroid therapy: one case report and literature review. Int J Rheum Dis. 2018;21:573-578.

XVI. Henriksson P, Hedner U, Nilsson IM, Nilsson P. Generalized proteolysis in a young woman with Weber-Christian disease (nodular nonsuppurative panniculitis). Scand J Haematol. 1975;14:355-360.

XVII. Mori K, Hiratsuka I, Sakai H, Hiwatashi K, Takahashi T. Hemorrhagic diathesis in Weber-Christian disease. Tohoku J Exp Med. 1976;118:227-243. [PubMed] [Google Scholar].

XVIII. Kimura H, Kako M, Yo K. Alcoholic hyalins (Mallory bodies) in a case of Weber-Christian disease: electron microscopic observations of liver involvement. Gastroenterology. 1980;78:807-812.