WEBER-CHRISTIAN DISEASE IN A 9 MONTH OLD BOY: REPORT OF A CASE

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Abstract: ***Idiopathic lobular panniculitis, sometimes called relapsing febrile nodular panniculitis or Weber Christian disease first described in 1892 by Pfeifer, is the term used to describe a group of diseases that present as subcutaneous inflammatory nodules and histologically display an inflammatory focus primarily within the fat lobules. Weber-Christian disease is the term applied to idiopathic cases of lobular panniculitis associated with systemic symptoms. The disease occurs predominantly in middle aged females although it can be seen in both sexes and at all ages. Hereby, we represent a case of diagnosed Weber-Christian disease in a 9-month-old boy. The diagnosis was approved by skin biopsy and exclusion of other differential diagnosis of lobular panniculitis. Weber-Christian disease is a rare disease especially in children and can be misdiagnosed, so we found it interesting to report this case.***

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INTRODUCTION

Systemic idiopathic lobular panniculitis or Weber-Christian disease was described in 1892 by Pfeifer and further delineated by Weber (1925) and christain (1-2).

The diagnosis of Weber-Christian disease is a diagnosis of exclusion (3-4). As a clearer understanding of the etiology of the panniculitis evolved, it was possible to separate lupus, factitious, pancreatic associated, hostiocytic cytophagic and alpha 1 antitrypsin deficiency panniculitis as diseases distinct from Weber-Christian disease (1).

The disease occurs predominantly in middle aged women (2-3), but cases have been reported in both cases and ages including the neonatal period (3-5).

The etiology of disease remains unknown. It has been suggested that it results from an immunologically mediated reaction to diverse antigenic stimuli because of an association in some patients with elevated levels of circulating immune complexes (1-2). Clinically, the disease is characterized by the appearance of crops of erythematous, edematous, sometimes-tender nodules and plaques in the subcutaneous fat, usually in association with mild fever (1-3). New waves of lesions appear at intervals from weeks to months (1). The nodules are usually 1 to 2 cm in size but may be much larger (1).

Lesions are usually symmetrical in distribution and occur most often on the thighs and lower legs but involvement of arms, trunk and face is sometimes reported (1-3). Occasionally due to fat necrosis, an oily brown liquid is discharged from the lesions (liquefying panniculitis) (1-3). Individual nodules involute over the course of few weeks leaving a depressed atrophic, sometimes hyperpigmented scar (3).

The appearance of cutaneous lesions is generally accompanied by symptoms of malaise, fever, arthralgia and fatigue (1). Involvement of intravisceral fat (of liver, spleen, myocardium or adrenals) and perivisceral fat (pericardium, pleura, mesenteric and omental fat) is also common. Bone marrow, lungs, intestines and kidneys may be involved (1). Finally reports of eye involvement (proptosis), cardiac manifestations (congestive heart failure), and skeletal muscle are present in literature (6-7-8).

Histologically, lobular panniculitis evolves through three phases (3):

The first phase occurs when there is only erythema and induration clinically. There is infiltration of neutrophils, lymphocytes and macrophages with degeneration of fat cells in lobules.

The second phase is recognized by an infiltrate mainly composed of macrophages (many with vacuolated cytoplasms due to ingestion of fat called foam cells), with few lymphocytes and plasma cells.

The third phase found in clinically depressed lesions is recognized by the presence of many fibroblasts, scattered lymphocytes and few plasma cells. Collagen deposits result in dense fibrosis.

Vascular changes are usually mild with only endothelial proliferation and edema, and thickening of the vessel walls (1).

No uniform effective therapy for idiopathic lobular panniculitis is recognized (1). Therapeutic

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responses to systemic steroids, NSAIDs, azathioprine, thalidomide and cyclosporine and some other drugs have been reported (1-2). Recently successful treatment with cyclosporine in some patients were reported (9,10).

The prognosis is extremely variable (1). In patients with only cutaneous involvement the prognosis is good (1). On the other hand a clinical course characterized by frequent remissions and prominent visceral involvement may eventually lead to death. Exacerbation of cutaneous lesions with eventual permanent remissions after several years is common (1).

Case report:

A 9 months old boy with a history of recurrent fever started 20 days after birth, sometimes with eruption of tender erythematous nodules arms, legs, thighs, trunk and face was referred at Children Medical Center. He was hospitalized several times at different hospitals without definite diagnosis (work-up of infectious and malignant causes were all negative) and received different medications including anti - TB drugs (despite a negative BK smear and cultures and negative P.P.D.) without good response. He was the fourth child of a father with lung cancer and his mother was receiving anti-TB drugs due to strongly positive PPD test.

Clinical examination at the time of admission revealed: fever (38.5^{o} C), tender cutaneous nodules in face, trunk and extremities, hepatomegaly (liver span: 9 cm), splenomegaly, insignificant cervical lymphadenopathy and pallor in face and conjunctivae. Laboratory tests and other diagnostic investigations revealed: leukocytosis (WBC= 27400) and polynucleosis (60%), anemia (Hg= 7.2), elevated ESR (ESR= 110), normal biochemical and liver function tests, negative infectious disease work up, hyperplastic bone barrow (increased myeloid series), normal chest x-ray and echocardiography.

At last, skin biopsy was performed and pathologic report was as follows (Fig. 1 and 2). " Sections show skin with focal heavy infiltrate of neutrophils, lymphocytes and plasma cells mainly in fat lobules resulting in destruction of fat cells (Fig 1, Fig 2). Vascular changes (endothelial proliferation and wall thickening) are present. The overall picture is in concordance with stage one in Weber-Christian disease".

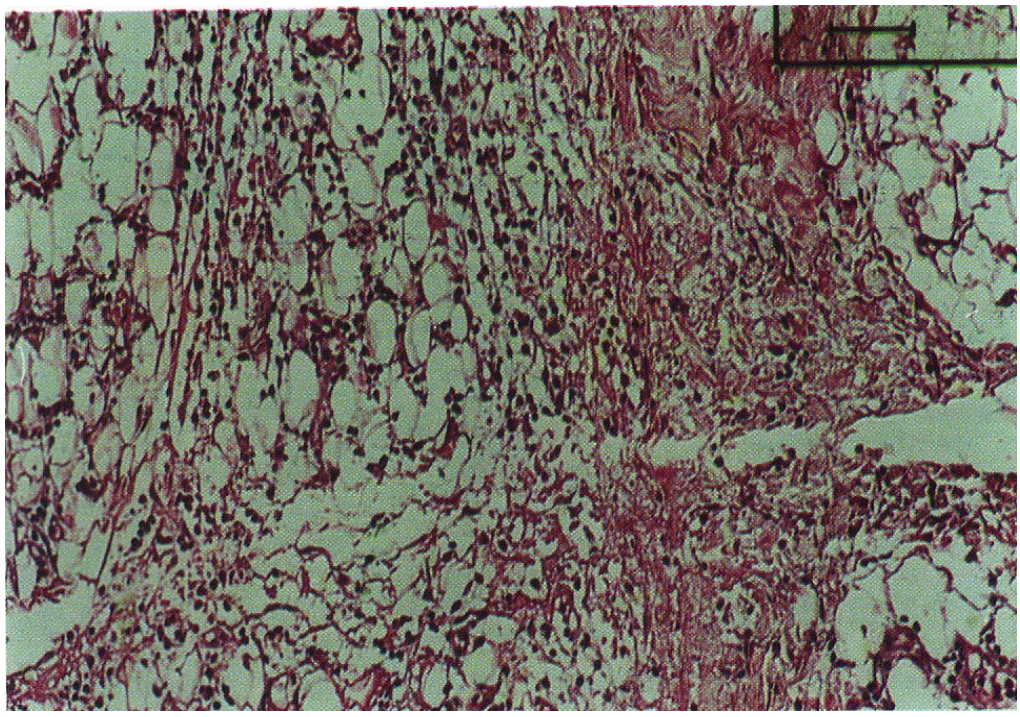

Fig. 1. Skin biopsy, heavy infiltrate of neutrophils, lymphocytes and plasma cells mainly in fat lobules

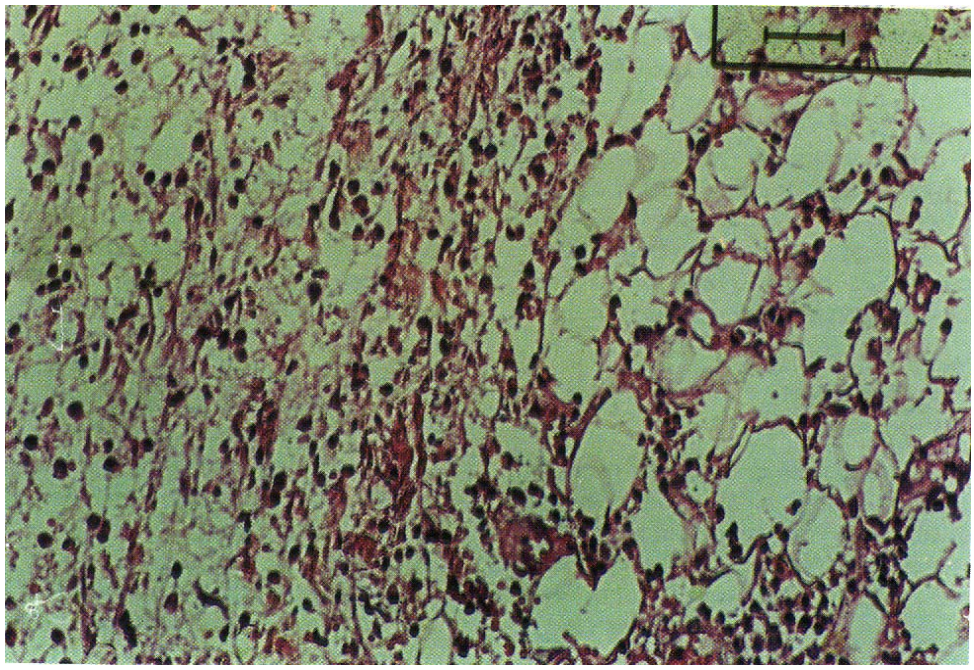

Fig. 2. Skin biopsy, closer view

Diagnosis of Weber-Christian disease was established and treatment with corticosteroid and Ibuprofen instituted for the patient. He had an initial good response. He is now under close follow up.

DISCUSSION

Although Weber-Christian disease is a rare disorder and is not as yet reported in Iran (particularly in children), but should be considered in confrontation with such cases in order to prevent the long delay between disease flare up and final diagnosis and to avoid unnecessary medications such as anti TB drugs as were used in this case.

As mentioned earlier, Weber-Christian disease is a diagnosis of exclusion and through work up of patients in order to exclude other causes of lobular panniculitis are highly recommended.

We could not find any cases of Weber-Christian disease with similar onset age in other case reports.

REFERENCES

1. Bondi EE, Margolis DJ, Lazarus GS. Panniculitis in: Fifth edition. The united state of America: Mc Graw-Hill, 1999; p. 1275-88.

2. Daniel WP. Disease of the subcutaneous Tissue. In: Moschella, S. and Hurley HJ. Editors. Dermatology 3 rd ed. Philadelphia: Saunders company, 1992; p. 1312-32.

3. Winkelmann RK, McEvoy MT, Peters MS. Lipophagic panniculitis of childhood. J AM Acad. Dermatol. 1989; 21(5): 971-8.

4. White JW, Winklmann RK. Weber-Christian panniculitis, a review of 30 cases with this diagnosis. J AM Acad. Dermatol; 1998, 39(1): 56-62.

5. Hendricks WM. Ahmad M. Weber-Christian syndrome in infancy. Br J Dermatol; 1978, 98: 175-89.

6. Cook JN, Kikkawa DO. Proptosis as the manifesting sign of Weber-Christian disease. American. J Ophtalmol; 1997, 124(1): 125-6.

7. Lemley DE, Ferrans VJ. Cardiac manifestations of Weber-Christian disease. J Rheumatol; 1991, 18(5): 750-60.

8. Ohara S et al. Myalgia as the major symptom in systemic panniculitis (Weber-Christian disease). Eur Neurol; 1992, 3(6): 321-3.

9. Viravan S, Wisuthsarewong W, Manonukul J, Tanphaichitr VS. Successful treatment of cytophagic histiocytic panniculitis by cyclosporine A: a case report. Asian. Pac. J. Allergy Immunol; 1997, 15(3): 161-6.

10. Iwasaki T et al. Kakishita E. Successful treatment of a patient with febrile, lobular panniculitis (Weber-Christian disease) with oral cyclosporine A: implications for pathogenesis and therapy. Intern Med; 1999, 38(7): 612-4.