Pfeifer-Weber-Christian Disease during Pregnancy Successfully Treated with Corticosteroids

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Sir,

Pfeifer–Weber–Christian disease (PWCD), also known as idiopathic relapsing febrile lobular nonsuppurative panniculitis, is a rare inflammatory disorder of subcutaneous adipose tissue.[1] There is no specific test for diagnosis and extensive investigations are required to exclude systemic causes of panniculitis. No uniform effective therapy is available and various drugs have been used in the past.[2] It has been reported most frequently in people in the fourth to seventh decades of life, and 75% of cases occur in women after the second decade of life.[3] We present a case of PWCD during pregnancy successfully treated with corticosteroids. To the best of our knowledge, this is the first case of PWCD during pregnancy, described in the published literature.

A 31-year-old female patient during the 13th week of pregnancy presented with high fever, lasting for several days and 10 days history of two lesions at the inner side of the right thigh and lower leg. Both lesions were reddish blue in color, slightly indurated, and painful that had led to a complete immobilization of the patient due to pain [Figure 1]. The patient further complained of malaise, nausea, bone pain, myalgia, and arthralgia. On presentation, patient’s temperature was just above 38°C. Palpable left axillary lymphadenopathy was noted. No hepatomegaly or splenomegaly was present. Laboratory investigations showed high inflammatory parameters (leukocytosis, elevated erythrocyte sedimentation rate, and C-reactive protein). Antinuclear antibody test was negative and cryoglobulins were not found. A1-antitrypsin, immunoglobulins, angiotensin-converting enzyme, and amylase were within the reference range. Computerized tomography of the chest and abdomen which was performed 7 months ago while investigating pyrexia of unknown origin that was finally diagnosed as drug-induced, were normal. Skin biopsy was obtained and showed an intact epidermis which however was infiltrated by small number of lymphocytes. No vasculitis was present. In the subcutaneous tissue, an infiltration of histiocytes and lymphocytes was noted [Figure 2]. Together with the clinical findings with a strong systemic inflammatory response without any indication of underlying infectious or malignant cause, we established the diagnosis of a PWCD. Due to the pregnancy, treatment options were limited. She was treated with prednisolone 35 mg once a day (0.5 mg/kg). The patient responded well to this treatment and temperature as well as inflammatory markers returned to normal levels rapidly. The initial dose of prednisolone was gradually reduced starting a week later. It was discontinued after a total course of 5 weeks and while patient was still going through the second trimester. On a routine check-up after 10 days on treatment, the patient presented in very good general condition with no more signs of inflammation. The skin lesions were still present but clearly reduced in size and painless [Figure 3]. The patient continued her pregnancy normally, without any complications. No steroids related side effects were noted in the mother. At 37th week of
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gestation, patient went into labor. Good neonatal and maternal outcomes were achieved.

PWCD is characterized by recurrent subcutaneous inflammatory painful nodules. Due to its rarity it is still under debate, if PWCD is a unique disease entity, or if it is just a substitute for every panniculitis that does not fit a common diagnosis.[4] PWCD has been classified as lobular panniculitis with mixed cell infiltrate. Lobular panniculitis can also occur in infections, malignancies, alpha-1-antitrypsin deficiency, pancreatitis, systemic lupus erythematosus and cytophagic histiocytic panniculitis, rendering PWCD an exclusion diagnosis following careful evaluation of the patient.[1] The lesions found in PWCD are often symmetric in distribution. Thighs and legs are involved more commonly. Occasionally, the epidermis overlying the nodule breaks down, discharging an oily liquid, called liquefying panniculitis. In most cases, it is associated with fever, malaise, myalgia, arthralgia and abdominal pain.[1] On histological examination, fat lobules are replaced by neutrophils, lymphocytes, and histiocytes in the early stages and by macrophages and fibrotic tissue later on. There is no septal involvement or vasculitis.[5] No uniformly effective therapy is known. Treatment of this condition during the second trimester of pregnancy can be challenging. This case reports that oral corticosteroids can be a safe and effective treatment option other than immunosuppressive agents, such as cyclosporine, in suppressing acute exacerbations of the disease.

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Conflicts of interest
There are no conflicts of interest.

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