Dermatomyositis Developing in the First Trimester of Pregnancy

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Abstract: Dermatomyositis in pregnancy is rare. Pregnancy may be a precipitating factor at the onset or many develop during the course of dermatomyositis which could exacerbate the disease activity. In this study we report a female patient who developed skin rash over face & both arms & hands & progressive proximal muscle weakness with cramps in the legs in 12th week of pregnancy. She was diagnosed with dermatomyositis and underwent therapeutic abortion and treated with methyl prednisolone.

Keywords: Dermatomyositis, pregnancy

1. Introduction

Dermatomyositis is a rare auto-immune disease with an incidence rate of one per million population. Only 14% of cases during child-bearing age group and only a few cases of dermatomyositis associated to pregnancy complications have been reported. We report the patient diagnosed with dermatomyositis first time in the first trimester of pregnancy. This is to our knowledge a patient with dermatomyositis treated with methyl prednisolone after therapeutic abortion. She still comes for follow-up.

2. Case Report

A 28 year-old female 3rd gravida with 2 children alive was admitted to our hospital on 13th January 2012 with history of 3 months amenorrhoea, major joint pain, stiffness of shoulder, elbow, hip joint and knee joint lasting for half hour, more in the morning with inability to walk due to weakness since two months, associated with muscle cramps & no swelling of joints.

She was not able to squat or sit up from lying down position without support & hyperpigmented rash over face, extremities. No previous history of similar episode in the past. No history of drugs or urinary complaints or no petechiae or purpurae or Raynod’s phenomenon. No history of joint swelling or dysphagia or oral ulceration. No history of fever or abortion.

On examination - Patient’s vital were normal. No evidence of joint swelling or effusion or synovial thickening or no joint deformity. Restricted shoulder, elbow, knee, hip, joint movements due to pain. Violaceous maculopapular rash over face, both extremities, non-blanching type. No petechiae or purpurae.

CNS-Higher functions And cranial nerves normal. Motor system-, power grade III, III. Reflexes depressed, no spinal tenderness, no cerebellar signs. Investigation: Hb-8.7 gm%, WBC- 13900, N-81%, L-118 ESR- 62mm/hr, CPK total 11,000 IU/L, RA factor negative.ANA-Positive, HIV And VDRL-Negative. Anti-SM and Anti-JO, Anti-DsDNA Negative, X-Ray LS Spine normal, USG Abdomen Gravid Uterus-12 Weeks. CT Chest, CT Spine normal. Sacroiliac Jt. normal. CT Abdomen Normal. The X-Rays and CT were done after MTP. EMG Myopathic pattern. NCV- Normal.

3. Skin and Muscle Biopsy

There is moderately dense inflammatory infiltrate composed of lymphocytes and plasmacells seen within the endomysial connective tissue. There is necrosis of muscle fibres along with phagocytosis. Perifascicular fibres show involvement with vocal perifascicular atrophy. Perivascular inflammatory infiltrates are seen. There is no vasculitis. There is increase in endo and perimysial connective tissue. No granulomas or organisms seen. Screening with CD 20, CD 3 reveals a mixed infiltrate of B & T Lymphocytes with CD 4 lymphocytes predominating. CD 138 highlights the plasma cells. The skin biopsy shows epidermal atrophy, loss of rete ridges and collagenized dermis. There is scant perivascular lymphocytic infiltrate in superficial dermis. One focus shows intra-epidermal lymphocytes. This is depleted on subsequent immunohistochemistry sections.

4. Impression

4.1 Muscle & Skin Biopsy

Muscle biopsy shows inflammatory myopathy. Taken in conjunction with the skin biopsy, the histology favours a diagnosis of Dermatomyositis. So the patient was diagnosed as dermatomyositis after thorough clinical and available diagnostic positive tests. She had undergone medical termination of pregnancy and then treated with IV methyl prednisolone followed by oral prednisolone and NSAIDS. She was observed for three weeks and discharged. Her power improved to grade IV. Now she comes for follow up regularly and she is in remission.
5. Discussion

Dermatomyositis is classified as idiopathic inflammatory myopathy. Women are affected twice as often as men and in adults the peak incidence occurs in fifth decade. Although all age groups are affected, only a few cases of dermatomyositis evolve complications in pregnancy. Very little information is available on this. Foetal morbidity and mortality is substantial and seem to parallel maternal disease activity. Still birth and neonatal death, complicated pregnancy is seen in 7/15 patients with active disease. Even in patients who have disease under control with treatment in pregnancy may result in foetal death and prematurity. When disease onset is in the first trimester foetal mortality is very high (83%). Even in second or third trimester the risk for premature delivery remains high. No foetal death occurred in such instance. 50% of previous reported cases of dermatomyositis began during first trimester of pregnancy and also in our present case study there was flareup and exacerbation of dermatomyositis first time diagnosed in pregnancy first trimester treated effectively and pt was discharged after pulse therapy of methyl prednisolone. Her symptoms were improved with MTP and steroids. On follow up she is in remission.

References