



PREGNANCY OUTCOME IN PATIENT WITH ADULT ONSET STILL'S DISEASE-A RARE CASE REPORT

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ABSTRACT

Still's disease is an arthritis of adult onset, rare in general population, even rarer in pregnancy, and often associated with flare-up and relapses during pregnancy and postpartum period. The following is a rare case of pregnancy in Still's disease with uneventfull outcome.

KEY WORDS: Still's disease, Pregnancy, Postpartum period.



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INTRODUCTION

Still's disease is a rare form of inflammatory arthritis. Its incidence is 1.5 cases per 100,000 – 1,000,000 population. The underlying cause is unknown. The disease presents with varied clinical features, including chances of abortion and preterm birth. Here is a case of ADULT ONSET STILL'S DISEASE, with no symptoms and a successful pregnancy outcome. We would like to report an uneventful pregnancy and delivery in a case of adult onset Still's disease in Sree Balaji Medical College, Chromepet, Chennai.

CASE REPORT

PREGNANCY AND UNEVENTFUL DELIVERY IN A CASE OF ADULT- ONSET STILL'S DISEASE:-

Mrs. Farzana, 24yrs, Gravida2, Abortion1, known case of Adult-Onset Still's Disease, diagnosed and treated one year ago by a Rheumatologist with steroids and Aspirin, which were stopped at the onset of the present pregnancy. The first pregnancy had been conceived spontaneously and went into spontaneous abortion at two months amenorrhoea-cause unknown, D&C not

done. The second pregnancy, being the present pregnancy, conceived with one cycle of ovulation induction, i.e., Tab. Letrozole by a private practitioner. She was booked at two months of amenorrhoea at our hospital. All three trimesters of pregnancy were uneventful. She had none of the symptoms or signs typical of Adult-Onset Still's Disease throughout the pregnancy, namely, Fever/ Arthralgia/ Arthritis/ Non-pruritic Salmon-coloured rash/ Sore throat/ Lymphadenopathy/ Hepatomegaly/ Splenomegaly/ Leukocytosis/ Abnormal Liver Function Tests. ANA, RA Factors were negative. She was admitted to hospital at 38 wks for institutional care. Labour was induced one day after her EDD by intracervical Prostaglandin E2 gel. Labour progressed well and she delivered within 6 hours by Labour natural, an alive term Male baby of 3.5kg with Apgar score of 8/10, 9/10, with no gross external anomalies. She had mild traumatic postpartum hemorrhage- the cause being a vaginal wall hematoma which was evacuated under anaesthesia and sutured. Postnatal period was uneventful. Patient was discharged on the third postnatal day. Review at 2nd and 6th postnatal week were uneventful.

DISCUSSION

Adult-onset Still's disease

is a rare form of inflammatory arthritis that was characterized by EG Bywaters in 1971.^[1] The underlying cause is unknown. It usually presents with high spiking fevers, joint and muscle pains, a salmon colored rash and other symptoms of systemic inflammation.

History

Still's disease is named after English physician Sir George Frederic Still

Epidemiology

Adult-onset Still's Disease is rare and has been described all over the world. Prevalence is estimated at 1.5 cases per 100,000-1,000,000 population. There is a bimodal age distribution with one peak incidence between ages 15–25 and a second peak between ages of 36–46 years

Pathophysiology

Its pathophysiology is cryptogenic: the underlying cause is unknown.

Signs and symptoms

The disease typically presents with arthralgia, fever, a 'salmon-pink' rash, and lymphadenopathy. Rheumatoid factor (RF) and anti-nuclear antibody (ANA) are classically negative and serum ferritin is elevated. Patients experiencing a flare-up from Adult-onset Still's disease usually report extreme fatigue, swelling of the lymph glands, and less commonly fluid accumulation in the lungs and heart.

Diagnosis

The diagnosis is clinical, not based upon serology-At least seven sets of diagnostic criteria have been devised, however the Yamaguchi criteria have the highest sensitivity. Diagnosis requires at least five features, with at least two of these being major diagnostic criteria.

Major criteria	Minor criteria
Fever of at least 39C for at least one week	Sore throat
Arthralgias or arthritis for at least two weeks	Lymphadenopathy
Nonpruritic salmon colored rash (usually over trunk or extremities while febrile)	Hepatomegaly or splenomegaly
Leukocytosis (10,000/microL or greater), with granulocyte predominance	Abnormal liver function tests
	Negative tests for antinuclear antibody and rheumatoid factor

Treatment

A 1987 article published in Arthritis & Rheumatism suggested that patients with Adult-onset Still's Disease can be differentiated into four types¹. Some patients experience monocyclic systemic disease, some experience polycyclic systemic disease, some experience chronic articular monocyclic systemic disease and others experience chronic articular polycyclic systemic disease. Treatment efficacy may depend on the nature of the patient's disease. Adult-onset Still's disease is treated with anti-inflammatory drugs. Steroids such as prednisone are used to treat severe symptoms of Still's. Other commonly-used medications

include hydroxychloroquine, penicillamine, azathioprine, methotrexate, etanercept, anakinra, cyclophosphamide, adalimumab, rituximab, and infliximab. A growing number of case studies have focused on the efficacy of Anakinra (or Kineret) in the treatment of Adult-onset Still's Disease. A 2005 article published in Arthritis & Rheumatism reported rapid improvement in four patients with Adult-onset Still's Disease who were treated with Anakinra. A randomized, multicenter study published in the Journal of Rheumatology in 2012 reported better outcomes in a group of 12 patients treated with Anakinra than in a group of 10 patients taking other disease-modifying antirheumatic drugs. Authors of a 2012 review article published in the International Journal of Inflammation concluded that "a growing number

of evidences supports the the utilisation of Anakinra in Adult-onset Still's Disease". The condition "juvenile-onset Still's disease" is now usually grouped under juvenile rheumatoid arthritis. However, there is some evidence that the two conditions are closely related.

REVIEW OF LITERATURE

The role of hormonal factors was first postulated to influence disease susceptibility in AOSD when Bywaters¹³ noted a slight female predominance. Indeed, our literature review identified 10 patients with first manifestations and 7 exacerbations in 12 pregnancies in patients with known AOSD². Although only 3 pregnancies were reported to be uneventful, it is likely that good pregnancy outcomes from patients with established AOSD are under-reported. First manifestations mostly occurred in the 5th to 6th gestational months . Three patients manifested AOSD in the first trimester , one in the post-partum period , and the timing of one was not mentioned . Exacerbations occurred despite inactive disease at conception, mostly in the post-partum period from 3 days to 5 months after delivery or even following spontaneous abortion . Two other exacerbations developed in the 4th to 5th gestational months^{3,4,5,6} . All episodes were classical acute systemic manifestations, except for one with an insidious onset of rash and arthritis. Elective termination of the pregnancy at the time of relapse did not result in ablation of disease activity. Transient articular symptoms of

mild severity were also frequent complaints . Multiple exacerbations in the same pregnancy were not common. Rather, most symptoms resulted from incomplete treatment of an earlier exacerbation. Salicylates and nonsteroidal antiinflammatory drugs (NSAID) showed initial response, but there was a subsequent resurgence of symptoms within weeks . One patient with no systemic complaint fared well with low dose prednisolone. Complete and partial responses were achieved with moderate doses (≤ 0.5 mg/kg/day) of prednisolone. High dose prednisolone (1 mg/kg/day) was successful and without adverse event. Pulse methylprednisolone, intravenous immunoglobulin (IVIG; and pulse cyclophosphamide (given after elective termination, were also reported to be efficacious⁶. Disease modifying agents including hydroxychloroquine, gold salts, MTX, and azathioprine were used as maintenance therapy after delivery, with variable response. Unfavorable pregnancy outcomes including

spontaneous abortion, prematurity and IUGR were occasionally observed⁸. It is likely that prematurity and IUGR occur more commonly in pregnancies complicated by disease exacerbation. Maternal morbidities were not common. Impaired glucose tolerance and preeclampsia were among those reported. Patients with AOSD may experience exacerbation of disease in pregnancy^{9,10}. Corticosteroid therapy is generally safe and can achieve satisfactory response and perhaps better fetal outcome. Long term data are needed for management of these patients.

CONCLUSION

Adult-onset Still disease in pregnancy can be confused with many other diseases, but its diagnosis, after exclusion of other infectious, malignant, and rheumatic conditions, can portend good maternal and fetal outcomes.

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