Case Report

Adult onset still's disease: Newly diagnosed in pregnancy with rare clinical presentation as pleural effusion

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ARTICLE INFO

Article history:
Received 27-05-2021
Accepted 02-09-2021
Available online 26-11-2021

Keywords:
Adult onset still’s disease
Pregnancy
Pleural effusion

ABSTRACT

Introduction: Adult onset Still’ disease (AOSD) is a rare inflammatory disease which involves multiple systems and is of unknown etiology first described in 1971. It has more prevalence among young females and few studies show the exacerbation of symptoms during pregnancy. Patients usually present with high grade fever, arthritis, arthralgia, salmon pink rash, organomegaly, lymphadenopathy but rarely can present with pleural symptoms, cardiac symptoms. Several criteria’s are used for diagnosis but Yamaguchi’s has the highest sensitivity of 93.5%. Yamaguchi’s classification criteria includes major and minor criteria’s and exclusion criteria’s. Although diagnostic criteria are there but AOSD till date is a diagnosis of exclusion. All the causes of clinical symptoms should be excluded which involves malignancies, infections, inflammatory conditions, autoimmune diseases before the diagnosis of AOSD is made.

Case Report: A 26-year-old female with G2P0010 and POG 6 weeks presented to medicine OPD with high grade fever associated with chills and rigors from 1 week. She was also complaining of stiffness and pain in multiple joints with chest pain, easy fatigability. She was having rashes on upper back below nape of neck. All the necessary investigations were carried out. It was found that she was also having right sided pleural effusion and severe microcytic hypochromic anemia. After batteries of investigations and consultation by obstetrician, rheumatologist, dermatologist diagnosis of Adult Onset Still’s disease was made. It was a diagnosis of exclusion. Patient was started on steroids and she responded well. All the symptoms including pleural effusion was subsided.

Discussion: Adult onset still’s disease is a form of still’s disease. It is a rare systemic auto-inflammatory disease. Yamaguchi is the most sensitive criteria but till date it is diagnosed after excluding possible causes of malignancies, infections and autoimmune conditions. Sometimes patients do present with rare symptoms like pleural effusion, pericarditis some may present as acute respiratory distress syndrome or mild symptoms like cough, pleuritic chest pain. In our case also all the possible causes were excluded as patient presented with pleural effusion for the first time during pregnancy, so the impact of the disease on the fetal outcome was also considered.

Conclusion: AOSD has always been a diagnosis of exclusion but as in our case and cases with similar presentation AOSD should be consider as a differential diagnosis. Although there are only few studies which shows that there is fatal outcome of pregnancy in AOSD however there are studies showing exacerbation of symptoms of AOSD during pregnancy. Its early diagnosis and prompt treatment can be helpful in the better outcome of pregnancy as well as better prognosis of disease.

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1. Introduction

Adult onset Still’ disease is a rare inflammatory disease which involves multiple systems and is of unknown
etiology first described in 1971.\(^1\) It is named after Sir George Frederic Still. It is also known as Wissler – Fanconi syndrome. There are several subdivisions of AOSD: 1. Monophasic 2. Polyphasic 3. Chronic. It has more prevalence among young females with few studies supporting the exacerbation of symptoms or first time manifestation during pregnancy. Patients usually presents with high grade fever, arthritis, arthralgia, salmon pink rash, organomegaly, lymphadenopathy but rarely can present with pleural symptoms, cardiac symptoms. Cardiac symptoms may involve myocarditis, pericarditis. Pleural symptoms may involve pleuritic pain, pleural effusion, cough and may even present as acute respiratory distress syndrome. For diagnosis several criteria are used but Yamaguchi’s has the highest sensitivity of 93.5%.\(^2\) Yamaguchi’s classification criteria includes major and minor criteria’s. Major criteria are high grade intermittent fever, arthralgia, salmon pink rash and leukocytosis. Minor criteria are sore throat, splenomegaly, lymphadenopathy, abnormal liver functions, negative RF and ANA. Although diagnostic criteria are there but AOSD till date is a diagnosis of exclusion. All the causes of all the clinical symptoms should be excluded which involves malignancies, infections and autoimmune diseases.

2. Case Report

A 26 year-old female with G2P0010 and POG 6 weeks presented to medicine OPD with high grade fever associated with chills and rigors for 1 week. She was also complaining of stiffness and pain in multiple joints with chest pain, easy fatigability. She was having rashes on upper back and below nape of neck. All the necessary investigations were carried out. Investigations showed Hemoglobin level of 7.4gm/dl. Peripheral smear showed sever microcytic hypochromic anemia. Total leucocyte count was 20,000 cells/ cumm. ESR 140mm 1\(^{st}\) hr, CRP positive, Ferritin and LDH raised. RA factor, anti CCP and ANA were negative. On chest X- Ray it was found that she was also having right sided pleural effusion and fluid analysis revealed an exudative fluid. Fluid appearance was turbid, fluid LDH 4200 U/L, fluid TLC 22160 cells /cumm with 98% neutrophils. Investigations for tuberculosis was also carried out as there was pleural effusion with fever but turned out to be negative. Blood and urine samples was also sent for culture and sensitivity. The cultures were sterile which ruled out the infective cause for all the manifestations. CT and X Rays were done to look for any mass, malignancies. Only positive finding on CT and X-Ray was the pleural effusion, this ruled out the malignant cause. After batteries of investigations and consultation by obstetrician, rheumatologist, dermatologist diagnosis of Adult onset Still’s disease was made. It was a diagnosis of exclusion. Patient started on steroids orally and she responded well. All the symptoms including pleural effusion was subsided. Pregnancy was continued. She was discharged on oral steroids and was advised to visit to medicine and OBG OPD for follow up.

3. Discussion

Adult onset Still’s disease is a form of Still’s disease. It is a rare systemic auto-inflammatory disease characterized by a clinical triad of fever, joint pain and salmon colored rashes. The disease is still diagnosed after excluding possible causes of malignancies, infections and autoimmune condition. In our case also all the possible causes were excluded and after conducting all the necessary investigations for malignancies, infections and autoimmune diseases the diagnosis of adult onset Still’s disease was made. In our case the patient presented with pleural effusion for the first time during pregnancy so the impact of the disease on the fetal outcome was also considered. The expression of AOSD in pregnancy was first reported in 1980 by Stein et al.\(^3\) Many studies are done in the past regarding correlation between AOSD and pregnancy. Le Loet et al found that pregnancy had no adverse effect on AOSD and AOSD had no influence on pregnancy outcome.\(^4\) First time expression of ASOD during pregnancy can be associated with sex hormones\(^5\) however we in our study cannot explain the relation between the sex hormones and AOSD expression but the patient did come with the symptoms for the first time during pregnancy. The rare symptoms of AOSD are pleural effusion, pericarditis some may present as acute respiratory distress syndrome. The mild rare symptoms may include cough, pleuritic chest pain with episode of shortness of breath.\(^6,7\)

4. Conclusion

Although AOSD has always been a diagnosis of exclusion but as in our case and cases presented with similar complaints, AOSD should be consider as a differential diagnosis. Although there are only few studies which shows that there is fatal outcome of pregnancy in AOSD however there are studies showing exacerbation of symptoms of AOSD during pregnancy\(^8\) as happened in our case also. Its early diagnosis and prompt treatment can be helpful in the better outcome of pregnancy and better prognosis in patients of AOSD, as in our case the patient was relieved of all the symptoms including pleural effusion on oral steroids only and the pregnancy was also continued with normal fetal development as shown by follow up ultrasounds.

5. Conflict of interest

None.

6. Source of Funding

None.
References

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