Abruptio Placentae in Adult Still's Disease with Onset During Pregnancy: A Case Report

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Abstract

Background: We investigated a woman who was presented fever of unknown origin, rash, artralji due to adult-onset Still's disease who developed abruption placenta.

Case: The patient was admitted at 20 weeks' gestation with fever, malaise, sore throat, cervical lymphadenopathy and a diffuse erythematous maculopapular rash for 10 days. After exclusion of an infectious or malignant disease, adult onset Still's disease was diagnosed according to the Yamaguchi criteria. After the exclusion of other infectious, malignant, and inflammatory causes, a diagnosis of adult-onset Still disease meeting the Yamaguchi criteria was made. She was treated with prednisone and had immediate improvement. Despite this, the dead fetus is delivered at 22 weeks of gestation with placental abruption.

Conclusion: To the best of our knowledge, this is the first case of AOSD who presented with placental abruption. Patients with adult onset Still disease are usually subjected to multiple diagnostic procedures and laboratory tests as well as empiric treatment with antibiotics and other medications. Corticosteroid therapy can achieve satisfactory response and perhaps better fetal outcome.

Keywords: Still's disease, abruption placentae, hyperferritinemia.

Gebelikte başlayan erişkin tip still hastalığında dekolman plasenta: olgu sunumu

Amaç: Plasenta dekolmanı gelişen bir Still hastalığı olgusu incelendi. Erişkin tip Still hastalığı nedeni açıklanamayan ateş, eklem ağrısı ve döküntü özellikleri ile seyreden romatolojik bir rahatsızlıktır.

Olgu: Gebeliğin 20. haftasında olan hasta ateş, yorgunluk, boğaz ağrısı, servikal lenfadenomegali ve 10 gündür süren vücutta yaygın eritematö-makulopapüler döküntüyle başvurdu. Enfeksiyöz, malign ve enflamatuar nedenlerin araştırılması sonrasında, Yamaguchi kriterlerine dayanılarak erişkin tip Still hastalığı tanısı konuldu. Prednizolona cevap veren hastada iki hafta sonra gelişen dekolman plasentaya bağlı fetal ölüm oluştu.

Sonuç: Plasenta dekolmanı ile ortaya çıkan literatürdeki ilk erişkin tip Still hastalığı olgusunu sunduk. Erişkin tip Still hastaları tanı konulmadan önce genellikle birçok laboratuar ve klinik testlere maruz kalırlar ve sıklıkla antibiyotik ve bazı diğer ilaçlarla ampirik olarak tedavi edilmeye çalışılırlar. Kortikosteroid tedavisi bu hastalarda tatmin edici sonuçların alınmasını sağlayarak fetüsün iyilik durumuna katkıda bulunabilir.

Anahtar Sözcükler: Still hastalığı, plasenta dekolmanı, hiperferritinemi.

Backgrond

Adult-onset Still's disease (AOSD) is an inflammatory condition that can involve internal organs, joints and also sometimes other parts of the body. The cause of the disease and occurrence mechanism is not understood clearly. Clinical properties are intermittent raising body temperature, arthralgia, macular or maculo-papular skin eruption, sore

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throat, myalgia, lymphadenomegaly and splenomegaly. ETSH is rarely diagnosed during pregnancy. This disease has to be differentially diagnosed especially with infectious diseases with viral ecsantemas, malignencies (lymphomas) and some rheumatologic diseases. Mostly a lot of laboratory and radiologic tests are done and these cause a delaying in diagnosis and therapy. Elevated serum ferritine has a diagnostic value in acute progressing Adult type Still disease.[1]

Adult type Still disease presenting with placental at pregnancy is studied with clinical and laboratory characteristics and reported in this article.

Case

A 25-year-old patient with 20 weeks pregnancy (G3, P2) is admitted with fever, sore throat, cervical lymphadenomegaly and diffuse erithematomaculopapular eruptions lasting 10 days. An anatomically normal fetus concordant with pregnancy week and normal placenta and amnion fluid is found in obstetrical ultrasonography . Her body temperature was 39.5°C when she admitted. Parenteral fluid therapy and third generation cephalosporin and erythromycin is started. There has been no difference in patient's condition during following three days. Fever of the patient was usually raising on mornings. Eruptions were rising while there was fever and then color of the eruptions were getting pale with decreasing of fever. In the patient's laboratory findings; anemia; Hb: 8.6 g/dl, Hct: % 23.6, ESR: 80 mm/hour, leukocytes: 19.500/mm3, platelets: 186,000/mm^3, ALT 32 U/L, AST 65 U/L and C-reactive protein (CRP) 10 mg/dl (normal value < 0.6 mg/dl) were found. ANA, ACA and rheumatoid factor and also blood, urine and throat cultures were found negative. After discarding infectious, malignant and inflammatory reasons that can cause this clinical disease, adult type Still's disease is diagnosed based upon Yamaguchi criteria.[2] Significantly elevated serum ferritin level in patient (> 13.000 mg/dl) is evaluated as a specific sign (normal values 15-300). Lymph node analysis from cervical region, reactive lymphocytosis is determined consistent with Still's disease. The patient's symptoms improved dramatically after 12 hours starting the treatment of 30 mg cortisone daily; fever decreased and did not increase again, myalgias and arthralgias disappeared, patient could wake up from her bed without help. After 10 days of this nice improvements patient had a vaginal bleeding and placental detachment is diagnosed in ultrasonography, the patient who intrauterine death occurred is delivered with induction (Figure 1).

Discussion

Adult type Still's disease is rarely seen and estimated incidence is reported as 0.34 in 100.000 women.[3] The disease which was first described at year of 1896 by George Still is being named as "juvenile idiopathic arthritis" in recent years.[4] The adult form of Still's disease which is generally seen in children is described by Bywaters.[5] Stein published first case appearing in pregnancy at 1980.[6] In a study that was made at year of 2004, 22 pregnancies were followed up in 17 patients with adult type Still's disease.[7] In ten patients Still's disease appeared in pregnancy for the first time, relapses occurred in seven of 12 pregnancies seen in previously diagnosed patients. Symptoms generally appeared in first trimester in the patients that were diagnosed in pregnancy for the first time. Although, it is reported that only three pregnancies resulted without problem, probably the other patients who were not developed any problem are not reported. Abortus, premature birth, retardation in intrauterine development and newborn death are reported related with pregnancy. Premature birth and retardation in intrauterine development are determined during relapses in patients previously diagnosed. Recurrences appeared most frequently in postpartum period. Impaired glucose tolerance and preeclampsia are reported for maternal morbidity. As understood from the study pregnancy condition does not affect the disease that is dragging on with recurrence and recovery periods. In another study made at 2003, 33 pregnancies occurring in 24 patients were examined.[8] According to this study there was not stated expressly a clear evidence about the effect of the pregnancy on adult type Still's disease or about the effect of adult type Still's disease on pregnancy. In the article, it is reported that beginning of the disease could be either in the antepartum or postpartum periods,

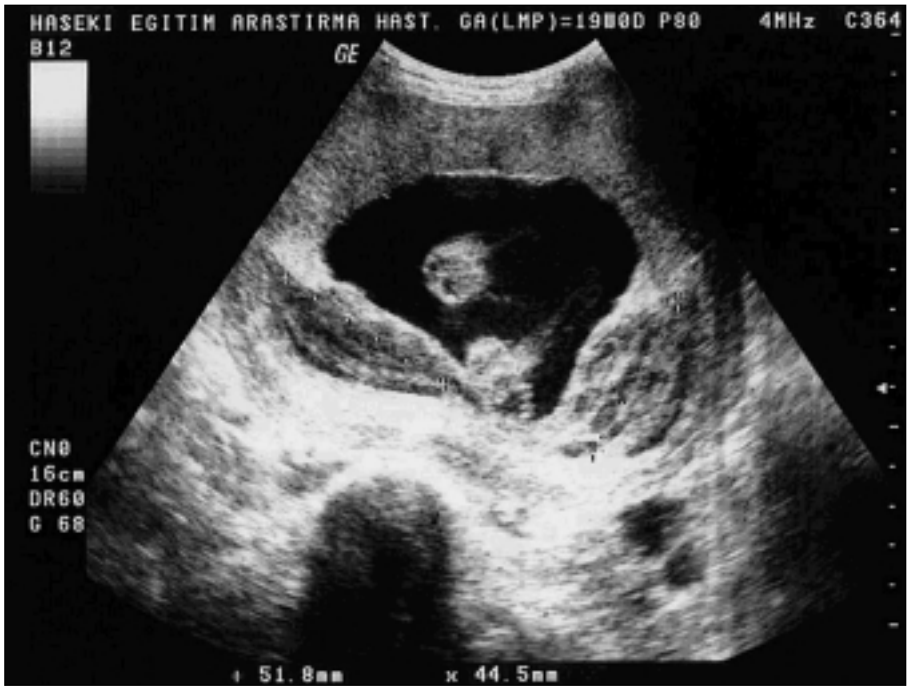

Figure 1. Retroplacental hematoma regions are seen at right and left.

Table 1. Major and minor criterion in adult type Still's disease.

Major criteria	Minor criteria
Fever > 39°C	Sore throat
Arthralgia > 2 weeks	Lymphadenomegaly or splenomegaly
Still eruptions	Elevation in liver enzymes
Neutrophilic leukocytosis	Negative rheumatoid and antinuclear factor

and also disease symptoms could get improved, did not change or recurrences were seen during pregnancy or in postpartum period. In long term follow-ups of children from mothers with adult type Still's disease there was no problem concerning education, job and social functions.[9] Clinical course is generally slight with recurrence and relapse periods. Life threatening internal organ involvement was not seen, however liver failure, pericarditis, acute respiratory distress syndrome, myocarditis causing heart failure, arrhythmias, pancytopenia, thrombotic thrombocytopenic purpurae-hemolytic uremic syndrome and diffuse intravessel coagulation were evident. In our case, retroplacental hematoma and placental detachment was observed. The observed detachment is interesting in the aspect that it shows a severe form of the disease. It may possibly be an indication that the disease affects the placental arteries. In recent studies, a relationship is found between the activation of the adult Still's disease and the increased ferrite serum level.[10] The increase of the serum ferrite concentration over 10000 ng/ml is considered to show the active period of the illness and for this reason, it is suggested to be used not only for the prediction of the illness but also the for monitoring the activity of the illness. Although many diagnosis criteria are suggested, it is accepted that criteria of Yamaguchi maintains the highest sensitivity (96.2%) and specificity (92.1%).[2] The adult type Still's disease is diagnosed with 10 or more points from 5 major criteria (2 points for each) or combination of major and minor criteria (one point each) (Table 1). Furthermore, there must be five or more criteria and at least two of them must be major criteria. The Adult Still's disease criteria predicted in our case are as follows: intermittent fever reaching peak points and then decreasing, continuing for two weeks together with sore throat, maculo-papular eruption seen with the increase of the fever and disappearing with the decrease of the fever, leukocytosis and arthralgia. In addition, increased level of ferrite for

the patient maintained the distinguishing from other rheumatologic diseases and is consistent with the laboratory results of this disease. As far as we can understand from studies which we could access, our case is the fist adult type Still's disease presented in the literature together with placental detachment. This case shows an instance of the adult type Still's disease presented with severe symptoms. ETSH should be considered in the diagnosis of patients administered to the hospital for fever with unknown reasons. Thus, the development of life threatening complications ETSH can be prevented with early diagnosis and remedy.

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