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EVALUATION OF SERUM FERRITIN LEVEL AS A DIAGNOSTIC MARKER IN STILL'S DISEASE: A CASE REPORT

Ramazan SARI, MD*
Alper SEVİNÇ, MD*
Cevher ÖZCAN, MD*
Süleyman BÜYÜKBERBER, MD*

* Inönü University, School of Medicine, Department of Internal Medicine,
MALATYA, TURKEY

Still's disease (SD) is an acute systemic inflammatory disorder characterized by a triad of spiking fever, skin rash and polyarthritis. Hyperferritinemia is one of the major laboratory findings. A markedly increased serum ferritin level can be used not only as an indicator of disease activity but also as a diagnostic marker of the disease. We report the case of a 15-year-old girl who was admitted to the hospital because of worsening joint pain and fever and diagnosed as SD.

***Key words**: Still's disease, ferritin, diagnosis*

Still hastalığı tanısında bir belirleyici olarak serum ferritin düzeyinin değerlendirilmesi: olgu sunumu

Ateş, cilt döküntüsü ve poliartrit ile seyreden Still hastalığı (SD), akut sistemik inflamatuar bir hastalıktır. Hiperferritinemi önemli laboratuvar bulgularından biridir. Serum ferritin düzeyinin aşırı artışı, hem hastalık aktivitesinin göstergesi hem de tanı kriteri olarak kullanılabilir. Bu vaka sunumunda eklem ağrısı ve yüksek ateş nedeniyle hastanemize başvuran ve SD tanısı konulan 15 yaşında bir olgu sunuldu.

***Anahtar kelimeler** : Still hastalığı, ferritin, tanı*

Correspondence address:
Dr Ramazan SARI
İnönü Üniversitesi Tıp Fakültesi, İç Hastalıkları Anabilim Dalı
MALATYA,TÜRKİYE

Still's Disease (SD) was first described by George Frederic Still in 1897. Classical SD is an acute systemic inflammatory disorder that has traditionally been considered as the juvenile form of rheumatoid arthritis. In the systemic form of the disease with rash and high fever, variable articular manifestations occur. A mild non-specific inflammation is also observed in the involved joints.[1]

SD is a rare syndrome of unknown origin consisting of recurrent fever, urticarial rash, arthralgias or arthritides, lymphadenopathy and splenomegaly. There is no pathognomonic symptom or specific laboratory abnormality. Increased erythrocyte sedimentation rate (ESR), negative auto-antibodies, leukocytosis, liver dysfunction and hyperferritinemia are major laboratory findings.[1,2] A very high level of ferritin is frequently observed and could be a marker of the disease. Ferritin is a major iron storage protein that has a diagnostic value for acute SD and a key factor in follow-up.[3] Serum ferritin level in SD was reported to be higher than one could expect for a simple inflammatory state, especially in adult onset SD.[4]

CASE

A 15-year-old girl was admitted to the hospital because of worsening joint pain and a low-grade intermittent fever. The patient was in a stable state of health until one year earlier, when she was admitted to another hospital due to severe pain, erythema and swelling in joints namely the knee, diarthroidial joints of spine, wrist, elbow, metocarpophalangials and sacroiliacs. She was complaining of fatigue, mild anorexia, weight loss, and progressive early morning stiffness. She did not improve on non-steroidal anti-inflammatory drug treatment. Physical examination revealed growth failure. There was a generalized rash and local pain in all of the joints. There was a mobile 1x1 cm sized lymphadenopathy in the right axillary region. A fine-needle aspiration biopsy was performed and cytological examination revealed reactive lymphocytic hyperplasia. Ultrasonography of abdomen revealed hepatosplenomegaly. Ophtalmological examination was within normal limits. Serum ferritin level was 5440 ng/ml (N:15-200 ng/ml). Serum immunoglobulin levels were evaluated as normal. All laboratory findings are presented in Table 1. The patient was diagnosed as SD upon clinical and laboratory findings. Prednisolone 1 mg/kg was started. She improved gradually. Serum ferritin and CRP levels were found to be decreased in two weeks. She was discharged from the hospital and is doing well with the disease at one year of follow-up.

Table 1. Laboratory findings on admission.

Hematocrit (%):	26
Hemoglobin (g/dl):	9.3
White-cell count (Per mm^3):	8500
Platelet count (Per mm^3):	409000
Serum iron level (μgr/dl):	28
Aspartat Aminotransferase (U/L):	438
Albumin/Globulin (g/L):	3.8/4.7
Lactate Dehydrogenase(U/L):	438
Alkaline Phosphatase (U/L) :	115
Iron binding capacity (μmol/dl):	250
Ferritin (ng/ml):	5480
ESR (mm/h) :	118
C-reactive protein (mg/dl) :	24
Anti nuclear antibody:	Negative
Complement-3 (mg/dl):	171
Complement-4 (mg/dl) :	41
Ceruloplasmin:	Normal
Anti-ds DNA:	Negative
Brucella agg. Test:	Negative
Rheumatoid factor :	Negative
Immunoglobulin A (mg/gl):	390
Immunoglobulin M (mg/gl):	137
Immunoglobulin G (mg/gl):	1340
Chest x-Ray:	Normal
Pelvic x-Ray :	Normal
ECG:	Normal
Abdominal USG:	Hepatosplenomegaly
Pelvic USG :	Normal
Torax CT:	Normal
Hypophyseal CT:	Normal
Adrenal CT:	Normal
Echocardiography:	Mitral regurgitation
Urine analysis:	Normal

DISCUSSION

Diagnosis of SD is difficult because of the great diversity of clinical and biological signs. Children with SD are most likely to be admitted to hospital with fever of unknown

origin to be assessed for spiking temperatures, which might antedate the onset of arthritis by several weeks. The migratory, maculopapular, salmon-colored rash that coincides with fever spikes can persist long after although other symptoms have abated. Activation of the reticulo-endothelial system with lymphadeno-pathy can be seen as a reactive hyperplasia. Joint manifestations at onset vary from none to severe polyarticular arthritis. Ninety-five percent of patients develop arthritis by the end of the first year of disease.[5] In 20 percent of the acute cases, high level of serum ferritin is associated with a hemophagocytic syndrome which responds well to immunosuppressive treatment.[6] Serum ferritin level is helpful in monitoring disease activity and guiding decisions about treatment. Detection of ferritin levels exceeding 3000 ng/ml should lead to the consideration of SD when there is an acute febrile illness.[7] Ferritin level helps to discriminate SD from arthritis of other systemic diseases. In patients with active SD, serum ferritin level was higher than that in patients with inactive SD and other diseases.[8,9,10] Additionally, the glycoform of isoferritins and the percentage of glycosylation offers an additional tool for the diagnosis of SD.[4]

In conclusion, we report a case of SD in whom serum ferritin level was correlated with the active disease. Although high serum ferritin level is not a pathognomonic finding for SD, it is an important laboratory test for diagnosis and follow-up. Serum ferritin level is a beneficial finding in discrimination of SD.

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