Araştırma Makalesi/Research Article

Orak Hücreli Anemi'de Kemik Yoğunluk Anomalileri ve İlişkili Faktörler

Bone Density Abnormalities and Related Factors in Sickle Cell Anemia

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ÖZET

AMAÇ: *Orak hücreli anemi (OHA)'de osteopeni ve osteoporoz riski net olarak ortaya konmamıştır. Bu çalışmada OHA hastalarımızda anormal kemik yoğunluğu sıklığını ve bunun klinik/laboratuvar bulgularla olan ilişkisini saptamayı amaçladık.*

YÖNTEMLER: *Çalışmamıza hemoglobinopati merkezimize rutin kontrole gelen, vazookluziv krizi olmayan OHA hastalarını aldık. Kemik mineral yoğunluğu (KMY) "dual energy x-rayabsorbtiometry (DEXA)" kullanılarak spinal kolon ve proksimal femurdan ölçüldü.Osteopeni ve osteoporoz WHO kriterlerine göre değerlendirildi.*

BULGULAR: *Altmış üç hastanın sonuçları değerlendirildi. Hastalarda %39,7 oranında osteopeni, %36,5 osteoporoz ve %23,8 normal KMY bulundu. Anormal KMY ile hemoglobin, lökosit, laktat dehidrogenaz, total bilirubin, ürik asit, alkalen fosfataz ve vitamin B12 düzeyleri arasında ilişki bulunamadı.*

SONUÇ: *Bu çalışmaya göre osteopeni ve osteoporoz OHA hastalarında sıktır. Klinisyenler osteoporoz hakkında dikkatli olmalı ve bu hastalar kırıklardan korunmak için KMY ölçümleri ile izlenmelidir.*

Anahtar Kelimeler: *Orak hücreli anemi, osteopeni, osteoporoz*
Türkçe Kısa Makale Başlığı: *Orak Hücreli Anemide Kemik Yoğunluk Anomalileri*

ABSTRACT

OBJECTIVE: *The risk of osteopenia and osteoporosis has not been clearly found in sickle cell anemia (SCA) patients. In this study, we aimed to find the frequency of abnormal bone mineral density and determine the relationship of abnormal BMD with clinical/laboratuary findings of our SCA patients.*

METHODS: *We added sickle cell anemia patients without vasooclusive crisis during rutine control in our hemoglobinopahy center. Bone mineral danistometry measurements were done with dual energy x-rayabsorbtiometry (DEXA) at lumbar spinal colon and proximal femur regions. Osteopenia and osteoporosis were evaluated according to WHO criteria.*

RESULTS: *We evaluated 63 patients' results. There were 39,7% osteopenia, 36,5% osteoporosis, 23,8% normal BMD. There was no relationship among abnormal BMD with hemoglobin, leukocyte, lactate dehydrogenase, total bilirubin, uric acid, alkaline phosphatase and vitamin B12 levels.*

CONCLUSION: *According to our study, osteopenia and osteoporosis are frequent in our SCA patients. Clinicians sould be careful about osteoporosis and these patients should be screen with BMD measurements for avoiding fractures.*

Key words: *Sickle cell anemia, osteopenia, osteoporosis*
İngilizce Kısa Makale Başlığı: *Bone Density Abnormalities in Sickle Cell*

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INTRODUCTION

Bone diseases in sickle cell anemia are microenfarcts, osteopenia, osteoporosis, osteomyelitis and osteonecrosis. Although mechanism of osteoporosis is not fully understood, etiologic factors may be unappropriate diet, hemolysis inflamation, limited vitamin d3 production at skin, and perhaps hospitalizations for vasooclusive crisis (1). Bone marrow hyperplasia is important role in pathogenesis (2). These bone complications don't cause mortality but play role in morbidity and low life quality. Although osteopenia and osteoporosis are generally asmptomatic; pain, fracture, deformity and vertebral collapse can be seen and chronic analgesia, mechanical / surgical support can be necessary.

MATERİALS and METHODS

We added sickle cell anemia patients without vasooclusive crisis during rutine control in our hemoglobinopahy center. Bone mineral danistometry measurements were done with dual energy x-rayabsorbtiometry (DEXA) at lumbar spinal colon and proximal femur regions. Osteopenia and osteoporosis were evaluated according to WHO criteria.

RESULTS

We evaluated 63 patients' results. Thirty patients are male, 33 patients were female. Mean age of males and females are 27,9 ±10,1 and 31,5±11,7 years respectively. Fourty eight of 63 patients (76,1%) had abnormal BMD. Fifteen patients (23,9%) had normal BM. In 48 patients with abnormal BMD , 25 patients had osteopenia and 23 had osteoporosis. Twelve of 30 male pateints had osteopenia and 8 of them had osteoporosis. Thirteen of 33 female patients had osteopenia, 15 of them had osteoporosis. Abnormal BMD frequency 66,7% in males, 84,9% in females.Osteoporosis in femur was seen 16,7% of males, 54,5% of females. Osteoporosis in lumber spinal region was seen in 36,7% of males and 51,5% of females. We didn't find any relationship among abnormal BMD with sex, mean levels of hemoglobin, leukocyte, lactate dehydrogenase, total bilirubin, uric acid, alkaline phosphatase and vitamin B 12. Similarly there was no relationship among abnormal BMD with hydrea using and renal disease (Table 1, Table 2)

Table 1. Abnormal BMD and sex, hydrea using, renal disease

	ABNORMAL BMD			p
	Yes	No	Total	
Sex	20	10	30	0,081
Male	28	5	33	
Female				
Hydrea using				
Yes	27	8	35	1
No	21	7	28	
Renal disease				
Yes	7	0	7	0,182
No	15	41	56	

Table 2. Abnormal BMD and laboratuary findings

Parameter	ABNORMAL BMD		p
	Yes	No	
Hemoglobin (g/dl)	8,4	9,7	0,120
Leukocyte (10^9/L)	11,7	10,5	0,528
Lactat dehydrogenase (mg/dl)	762	652	0,282
Total bilirubin (mg/dl)	1,92	2,89	0,058
Uric acid(mg/dl)	5,5	5,6	0,886
Alkaline phosphotase(mg/dl)	88,9	99,3	0,712
Vitamin B_{12} (pg/ml)	233	377	0,052
Vitamin D_3 (ng/ml)	14,4	19,1	0,217
Ferritin (pg/L)	458	701	0,270
C-reactive protein (mg/dl)	6	4,1	0,122

DISCUSSION

In a study with 14 sickle cell patients with osteopenia, osteoporosis and vitamin D deficiency, after calcium and vitamin D replacement for 12 month, 25 hidroxy vitamin D and BMD were improved. Bone resorption markers didn't change. Vitamin D screening was emphasized in this study (1). In our study, vitamin D3 levels were low in both normal BMD and abnormal BMD groups.

There were no statystical meaningful difference between two groups.

Low sex steroids were found as major role player for developing osteopenia and osteoporosis in a study conducted with 103 sickle cell patients (2).

In study conducted in brasil with 65 SCA patients, low BMD was found in relationship with hemolysis parameters like high lactate dehydrogenase (LDH), low hemoglobin level (3).

In a study done with 103 SCA patients, abnormal BMD was found in no relationship with age, sex, menarge, vasooclusive crisis complications, iron deposition, hydroxyurea/ desferoxamin using, renal disease, smoking or alcohol using. Abnormal BMD was seen more in patients taking hydroxyurea at least 6 months. Body mass index, ferritin, hemoglobin type and level were found major rol players for abnormal BMD (4).

In another study done with 17 SCA patients, osteopenia frequency was 47% and serum iron and ferritin levels were found higher in osteopenic patients than patients with normal BMD (5).

Another study done with 60 SCA patients showed that magnesium deficiency contributed to osteoporosis and rutin screening of magnesium level and treatment of low magnesium levels were recommended (6).

In a study with 50 children with SCA, osteopenia and osteoporosis frequencies were 11% and 2,2% at vertebral area. Osteopenia was 11% of patients at femur neck. The most endocrine disorder was found as vitamin D deficiency. When age increased, BMD of femur neck was found lower (7).

In conlusion, osteopenia and osteoporosis is not a rare complication of SCA in our patients. BMD screening should be done for preventing fractures in these patients.

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