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اسم البحث:-

## Osteoporosis in Iraqi patients with thalassemia

### Abstract

Background:-Thalassemia is defined as a complete absence of one or more of the four globins in the red blood cells due to the deletion of or nonfunctioning of one or more genes .

Osteoporosis is a universal medical problem, affecting both genders . It is generally accepted that its main causes are aging, genetic disorders of osteogenesis, lack of certain nutritional elements or physical activity and endocrine disorders mainly estrogen deficiency . Other causes include neoplastic disorders, gastrointestinal disorders causing malabsorption, liver diseases, inflammatory conditions, and drugs .

Materials and methods:- 74 thalassemic patients 36 male and 38 female below the age of 25 years were included in this study at AL Salaam General Teaching Hospital and in Ibn AL Atheer Pediatric Teaching Hospital, (Thalassemia center) in Iraq / Mosul city. Assessment was done by one rheumatology research physician and by one radiologist for the bone densitometry examination.

The study was a clinical cross-sectional for both genders with thalassemia major and also those which were associated with other types of blood disorder like sickle cell anemia or sideroblastic anemia. Investigation done included a chest x ray, serum iron, total iron binding capacity (TIBC), transferrin saturation, serum calcium, serum phosphorus, serum alkaline phosphatase, blood urea, serum creatinine, and a DXA bone scan .

Statistical analysis:-P-value—S.P.S.S.—chi-square

Results:- We found that the bony disorder in thalassemic patients increased with age (bone pain, carpedal spasm, osteoporosis), and with low serum iron and low T.I.B.C. and with increased transferrin saturation. The compliance of patients with treatment was rated as in 24 good, in 36 fair and in 14 bad. We found that the compliance of patients is good with younger age group while the fair and bad compliance was more with older age ,also good compliance was found with those early diagnosed cases and earlier administration of chelating therapy and blood transfusion ,also good compliance was more in patients with normal serum iron and normal T.I.B.C. ,also we found that a normal BMD was found more in patients with good compliance.

The prevalence of osteoporosis in thalassemic Iraqi patients DXA scans was found to be 67.5% while osteopenia was found in 9.4% and normal BMD in 22.9%

Discussion:- The modern radiologist is unlikely to encounter the classic radiographic features of thalassemia other than in teaching files . The main pathological change that leads to radiological skeletal changes in beta thalassemia major is extensive marrow proliferation .



The severity of the skeletal responses is related to the type of thalassemia, the extent and duration of the disease, the type of treatment and the volume of blood transfusions given to

the patient, as well as the side effects of transfusion-chelating therapy, and also depends on the bone involved. The radiographic features can be divided into those affecting the skeleton (axial and appendicular) and those occurring extra-medullary. Axial skeletal changes mainly include skull and facial bones, para nasal sinuses, vertebral bodies and in weight-bearing bones, while appendicular skeleton manifestations are more pronounced in peripheral bones, mainly hands and feet as well as ribs. Patients on repeated blood transfusion and iron-chelating therapy may demonstrate variable range of manifestations than in the scope of untreated patients.

During the last decade, the presence of osteopenia and osteoporosis in well-treated thalassaemics has been described in different studies with high prevalence up to 50%. The pathogenesis of osteoporosis in thalassaemia major is complicated and differs from the pathogenesis of bone deformities characteristically found in non transfused patients who develop bone distortion mainly due to ineffective haemopoiesis and progressive marrow expansion .

Several factors are implicated in reduction of bone mass in thalassaemia major. Delayed sexual maturation, growth hormone (GH) and insulin growth factor-(IGF)-1 deficiency, parathyroid gland dysfunction, diabetes, hypothyroidism, ineffective haemopoiesis with progressive marrow expansion, direct iron toxicity on osteoblasts, as well as liver disease have been indicated as possible etiological factors for thalassaemia-induced osteoporosis. Furthermore, iron chelating has correlated with growth failure and bone abnormalities, and high desferrioxamine dosage has been associated with cartilage alterations .

Conclusions:- Osteoporosis in thalassaemic Iraqi patient was too high and even more in those patients with bad compliance regard attendance to the Thalassemia centre.

Recommendations:- We need to inform the thalassaemic patients about the risk of osteoporosis and the need for their awareness regard such complication and the importance of their compliance with therapy .