Low Bone Mass in Ehlers-Danlos Syndrome

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A 14-year-old girl with type-III Ehlers-Danlos syndrome (EDS) underwent a dual energy X-ray absorptiometry (DXA) evaluation of the bone mineral density (BMD) in the scoliotic lumbar spine (L-spine), which measured 0.464 g/cm² (T-score: -5.3, Z-score: -4.23) (Picture 1). The BMD was found to be 0.230 g/cm² (T-score: -4.5) in the deformed distal forearm and 0.181 g/cm² (T-score: -4.1) in the ultra-distal forearm using single energy X-ray absorptiometry (SXA) (Picture 2). The patient was treated with teriparatide and her BMD in the distal radius thereafter consistently increased without showing any increase in the BMD in the L-spine. The total BMD was found to be 458 mg/cm³ (T-score: 1.2, Z-score: 1.4) at the radius-4% distal site, and the trabecular BMD was found to be 220.5 mg/cm³ (T-score: 0.5, Z-score: 0.6) at the same site using peripheral quantitative CT (pQCT). The volumetric BMD was found to be 71.2 mg/cm³ (T-score: -3.74, Z-score: -4.51) in the lordotic L-spine on QCT (Picture 3). We herein report the occurrence of marked osteoporosis in the peripheral skeleton of EDS patients as an additional, yet heretofore unrecognized, manifestation of...
EDS (1, 2).

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References


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