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PEDIATRIC RHEUMATOLOGY

POSTER PRESENTATION Open Access

Mc Cune Albright syndrome: estimation of bone strength parameters and response to treatment using peripheral Quantitive Computed Tomography (pQCT) of the tibia

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Aim

We assessed bone strength parameters and response to treatment in a girl with McCune -Albright syndrome (MAS) using tibia pQCT.

Methods

We present a 14y old girl with polyostotic fibrous dysplasia (right humerus, femur, tibia, skull), precocious puberty and café au lait skin spots, diagnosed as MAS with a confirmed heterozygous c.601C>T mutation of the GNAS1 gene. Due to initial bone pain and continuously increasing bone turnover, the patient was treated with iv bisphosphonates for 4 years. We used pQCT to estimate bone strength parameters at the site of fibrous dysplasia lesion of the right tibia (38% of tibia length) vs the same site of the left (healthy) tibia at baseline and after treatment. A Stratec XCT-2000 scanner was used (Stratec Medizintechnik, Pforzheim, Germany) and we specifically assessed for the 38% site cortical BMC (Cort_CNT), cortical BMD (Cort_DEN), cortical area (Cort_A),cortical thickness (Cort_THK) and Stress Strain Index (SSI) as an indicator of bending/torsional strength.

Results

At baseline all parameters were lower at the right (lesional) tibia: Cort_DEN (right 916.53 vs left 1154.47 mg/cmÂ³), Cort_CNT (0.78 vs 2.65 gr/cm), Cort_A (85 vs 230.25 mm^2) , Cort_THK (0.99 vs 4.76 mm), SSI (941 vs 1110.35 mm^3). All parameters increased significantly after 4 years of therapy at both legs with maximal increases at the lesional tibia: Cort_DEN (left +10.16% vs right + 13.54%), Cort CNT (+13.9% vs +34.6%), Cort_A (+3.14% vs +19.7%), Cort_THK (+4.2% vs +19.19%), SSI (+16.77% vs +26.4%).

Conclusions

1) With 3- dimensional densitometry we can actually measure the loss of cortical bone and derived strength of lesional sites in MAS 2) All bone strength parameters improved with iv bisphosphonates. P QCT, where applicable, is an easy, safe and accurate method for non invasive monitoring of disease progress.

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