Cortical Hyperostosis after Long-Lasting Prostaglandin E1 Treatment in a Newborn with Complex Congenital Heart Disease

A newborn girl was affected by a "ductal-dependent" complex congenital heart disease, which necessitated a long period of prostaglandin (prostaglandin E1) infusion; during this treatment, she developed cortical hyperostosis of the long bones. The patient was originally treated with antibiotics for a neonatal *Staphylococcus aureus* infection complicated with endocarditis and conjunctivitis. After 2 months of prostaglandin infusion, she presented with an antalgic position of the legs and was suspected to have a septic articular complication of the hip joints. Laboratory indexes were normal. Magnetic resonance imaging (MRI) was performed to rule out articular involvement. The examination was performed with the patient under general anesthesia on a 1.5-T MRI scan. T1 and T2 fast spin echo and turbo inversion recovery magnitude sequences, with and without fat suppression, were acquired. The investigation was completed with a fat-suppressed 3-dimensional T1-weighted technique after gadolinium-based contrast medium administration.

Images demonstrated thickening of the soft tissues around long bone shafts, especially the femurs, associated with signal hyperintensity on T2-weighted images; T1 fat-saturated images acquired after contrast administration highlighted cortical hyperemia. No significant intracapsular effusion involved the hips, and diffuse soft tissues thickening was visualized (Figure 1). These seemed to not be pathognomonic of septic articular involvement; however, because of the previous endocarditis and elevated indices of systemic inflammation, we prolonged antibiotic therapy. After 20 days, a repeat scan was performed to evaluate disease evolution, because there was persistence of clinical symptoms along with normalization of inflammatory indices. The second MRI scan revealed diffuse bilateral cortical bone thickening of femurs shaft with the typical aspect of "bone in bone" with a worsening of the soft-tissue involvement (Figure 2).

Cortical hyperostosis is a complication of extended prostaglandin E1 treatments.[1,2] Clinical symptoms could be pretibial soft-tissue swelling of the extremities and reversible proliferation of cortical bone.[3] This syndrome has been described and imaged with conventional radiology.[4] In the present case, a definitive diagnosis was required because septic articular involvement would contraindicate the mandatory surgical correction of the complex heart malformation. MRI soft-tissue involvement in this specific setting should be carefully evaluated and not wrongly addressed as a sign of joint disease. ■

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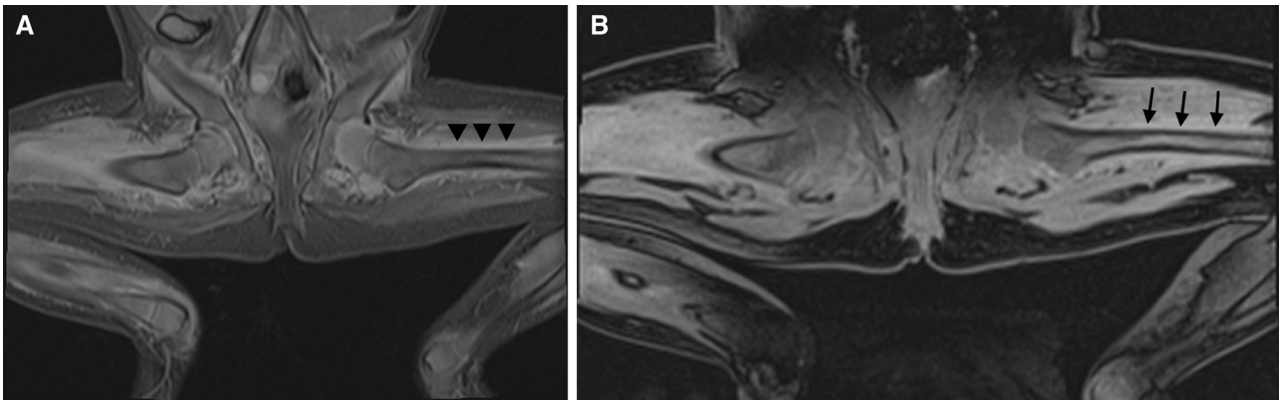

Figure 1. **A,** T2 turbo inversion recovery magnitude (slice thickness 3.0 mm; repetition time 4160 milliseconds; echo time 36 milliseconds; TI 140 milliseconds) image acquired on the coronal plane demonstrates thickening of the soft tissues around long bone shafts, especially the femurs (*black arrowheads*). **B,** T1 volumetric interpolated breath-hold examination 3-dimensional (slice thickness 0.5 mm; repetition time 16 milliseconds; echo time 6.29 milliseconds) image acquired on the coronal plane post-contrast medium administration with fat saturation demonstrates femoral cortical hyperemia (*black arrows*).

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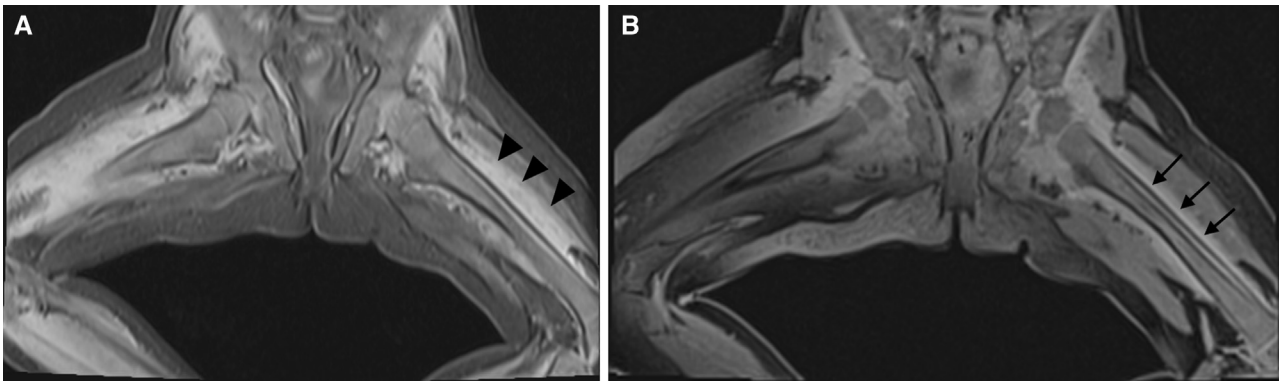

Figure 2. **A,** T2 turbo inversion recovery magnitude (slice thickness 3.0 mm; repetition time 4160 milliseconds; echo time 36 milliseconds; TI 140 milliseconds) image acquired on the coronal plane after 20 days highlights a worsening of the soft-tissue involvement (*black arrowheads*). **B,** T1 volumetric interpolated breath-hold examination 3-dimensional (slice thickness 0.5 mm; repetition time 16 milliseconds; echo time 6.29 milliseconds) image acquired on the coronal plane postcontrast medium administration with fat saturation highlights a diffuse bilateral cortical bone thickening of femur shaft with the typical aspect of "bone in bone" (*black arrows*).

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Reversible Myelofibrosis in Pediatric Renal Osteodystrophy

A 9-year-old female with no medical care for 3 years presented with recurrent emesis, epistaxis, and pallor. Laboratory evaluation showed renal failure, with a blood urea nitrogen level of 216 mg/dL and a serum creatinine concentration of 17.5 mg/dL. A hematologic workup showed severe normocytic anemia, with a hemoglobin concentration of 4.3 g/dL and an initial platelet count of 108×10^9/L that decreased to 60×10^9/L over 5 days. Her total white blood cell count was normal, but she had mild lymphopenia (1.3×10^9/L). Parathyroid hormone (PTH) levels were markedly elevated at 1600 pg/mL (normal range, 15-87 pg/mL). A peripheral blood smear review showed normocytic normochromic anemia with moderate anisopoikilocytosis, including rare acanthocytes, teardrop cells, and rare schistocytes. No blasts were present.

The cause of renal failure was not determined, but the patient had small bilateral kidneys on ultrasound, consistent with long-standing chronic kidney disease (CKD), possibly secondary to renal dysplasia. Bone marrow biopsy revealed hypocellularity (30%) and grade 2 nonuniform, patchy peritrabecular myelofibrosis with increased osteoclastic and osteoblastic activity (**Figure**, A). No immunophenotypic or cytogenetic abnormalities were detected, and testing for *JAK2V617F* was negative. The bone marrow findings in the setting of hyperparathyroidism due to long-standing CKD and the absence of other etiologies were consistent with nonmalignant myelofibrosis due to secondary hyperparathyroidism/renal osteodystrophy.

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