FUNDACIÓN VALLE DEL LILI

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TYPE III IDIOPATHIC MULTICENTRIC OSTEOLYSIS (IMO)

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HISTORY

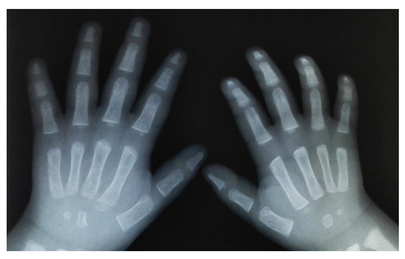
Idiopathic osteolysis was described for the first time by Jackson in the year 1838. In 1937, Froelich and Corret published cases with osteolysis carpo-tarsal and Derot et al. reported the association of osteolysis with renal involvement. In 1985, Hardegger established five subtypes of Idiopathic Multicentric Osteolysis via characterization of the clinical manifestations, the most affected sites, the age of onset and the type of inheritance (See box)

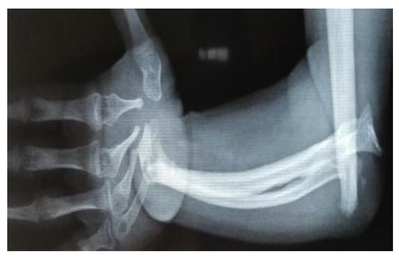
INTRODUCTION

Type III multicentric osteolysis is a rare skeletal disorder characterized by osteolysis of the carpus and the bones of the tarsus and deterioration of the joints that lead to crippling deformities (1), with acute renal failure secondary to the focal segmental sclerosis

Kidney disease usually appears later, progressing to end-stage renal disease (1). Sometimes patients develop secondary hyperparathyroidism with hypercalcaemia.

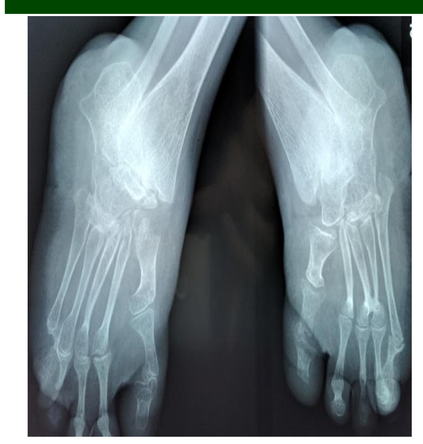
CASE DESCRIPTION



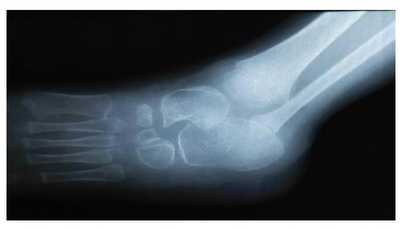


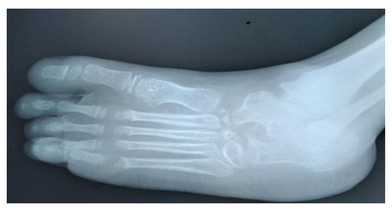
23 year old male patient, with a clinical history of renal transplantation for chronic kidney disease, associated with Type III multicentric osteolysis. The patient does not have a medical history of any importance. He is the product of a third normal pregnancy, with sisters with no phenotypic signs of the disease. There is no family history of any importance.

SIGNS AND SYMPTOMS IN THE PATIENT

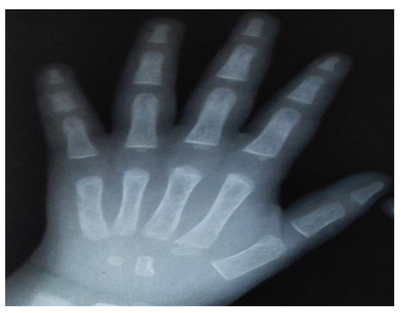


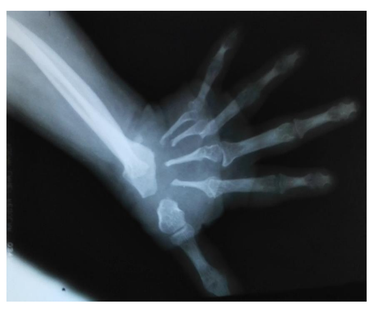
1. Arthralgias
2. Difficulties with the flexo-extension of the joints
3. Difficulties with the march.
4. Contracture of the left knee up to 90°.
5. Epiphyseal osteolysis of the bones of the foot and hand.





Multiple malformations in the joints of the ankles of the tarsus, tarsometatarsal and interphalangeal joints with varus deformity in both metatarsals





Patient with absence of the bony structures of the carpus with hypertrophy and consequent shortening of the metacarpals. There is also the presence of phalangyal metacarpus joints with an absence of fracture lines.

DIAGNOSTICO

1. Clinical condition (2)

2. Biochemical analysis, labs, autoimmune diseases, kidney function tests, parathyroid hormone (PTH), Vitamin D and bone markers

3. KIDNEY BIOPSY

4. **BONE DENSITOMETRY**

5. DIAGNOSTIC IMAGING:
Computed tomography (CT) scan
X-Ray.

IDIOPATHIC MULTICENTRIC OSTEOLYSIS. HARDEGGER CLASSIFICATION, 1985.

TYPE	AGE OF ONSET	SITE WITH THE HIGHEST DEGREE OF OSTEOLYSIS	INHERITED	ASSOCIATED CHARACTERISTICS
I Carpo-tarsal Osteolysis	2 to 7 years Sometimes reappears in the 3rd decade	Carpus and tarsus distal epiphysis ulna and radius elbows Osteolysis of the phalanges	AD	Is self-limited in adolescence Deformities in hands and feet Rarely nephropathy
II Carpo-tarsal Osteolysis Torg's Disease	1 to 5 years	Carpus and tarsus distal epiphysis ulna and radius elbows Osteolysis of the phalanges	AR	Generalized Osteoporosis with thinning of the cortical Rarely nephropathy
III Non-hereditary multicentric osteolysis associated with nephropathy	Infancy	Carpus to a greater degree than metacarpals and phalanges Osteolysis of the centers of ossification of the radius and ulna	Not hereditary	Normal Size. It is associated with nephropathy (glomerulonephritis) and malignant hypertension
IV Gorham massive osteolysis	Young adult age	Massive due to malformations: angiomatosis or hemangiomatosis.	Not hereditary	Unifocal and massive with slow progression, The destructive process ceases after a few years
V Winchester Disease	Infancy	Carpo-tarsal and elbows	AR	Low height, contractures, osteoporosis, skin lesions

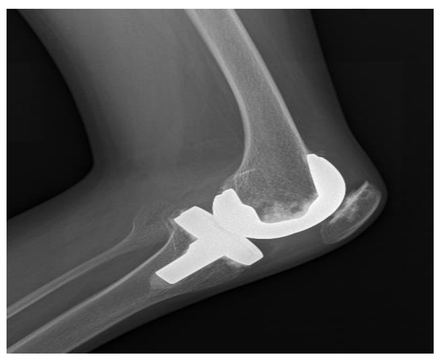
HOSPITAL STAY

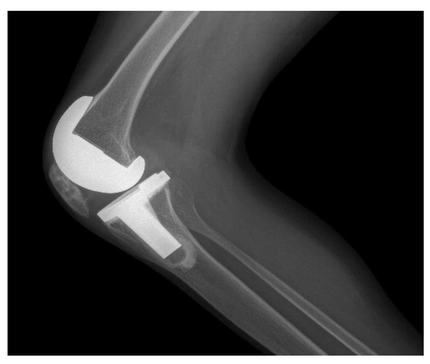
Paraclínicos	SCr	BUN	Na	K
Admission	7.0	--	142.0	2.94
Before the surgical procedure	4.03	24.60	142.0	2.96
24 hours before the procedure.	1.13	12.9	141	4.36
48 hours after the procedure.	0.47	9.7	141	4.12
72 hours after the procedure.	0.57	9.7	---	3.7
Discharge	0.57	9.7	---	3.73
15 days after discharge	0.69	---	141	4.73
Currently	0.73	12.3	139	3.93

TREATMENT

The treatment is palliative (non-steroidal anti-inflammatory drugs, NSAIDS, physical therapy and rehabilitation)
Terminal renal failure should be treated via kidney transplant.
Hypertension: use of antihypertensive drugs (1.2, 3).
In other cases, patients have required surgical intervention, for the correction of bone deformities with the objective of rehabilitating function
Ferreira-Garrott L et al. date have reported treatment of kidney disease with renal transplantation, with good results.

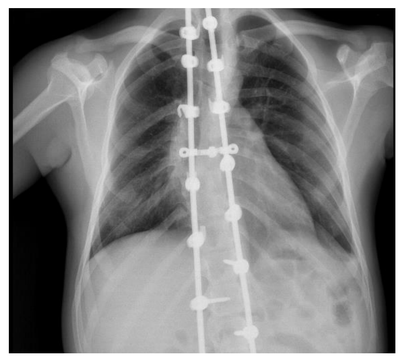
PATIENT IMAGES

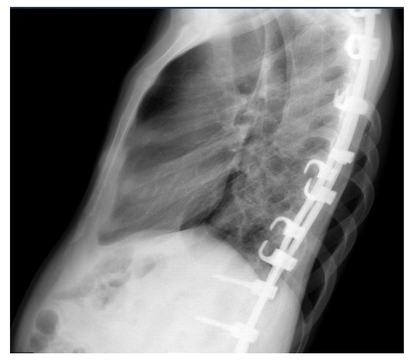




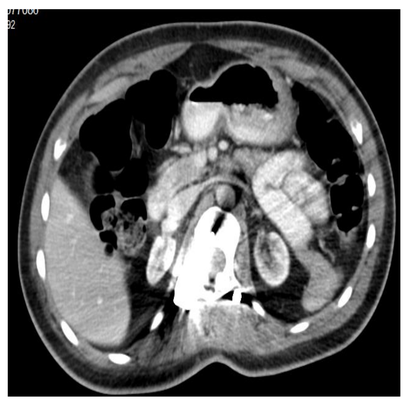
Condroplasty, synovectomy of the knee.

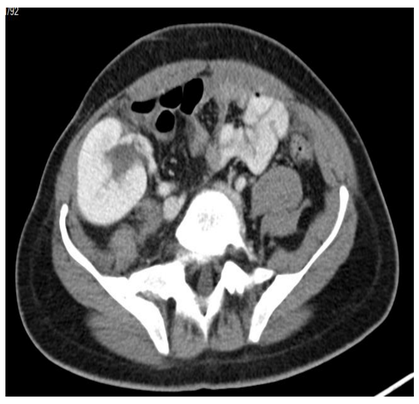
The patient showed deteriorating total bone mass with degeneration of the vertebrae and scoliosis. Underwent surgery for the implantation of screws, hooks and reconstruction, with the aim of rehabilitation.





The patient progressed to terminal renal failure despite medical management, for which a kidney transplant was considered. This was performed with induction of thymoglobulin with the aim of having maintenance treatment free of steroids. Excellent clinical evolution after transplantation, with no immunological events or rejection or other associated complications.





Graft in the right iliac fossa of normal size, contours and density. Slight pyelocalyceal ectasia with renal pelvis of 9mm, with no dilatation of the neo ureter. Graft vascular structures are adequate. Changes indicative of chronic kidney disease can be observed in the native kidneys.

DISCUSSION

1. Multicentric osteolysis type III is a rare skeletal disorder characterized by osteolysis of the carpus, bones of the tarsus and deterioration of the joints that lead to crippling deformities.
2. We have studied renal failure in a patient with multicentric osteolysis, where the presence of GEFS (1,2,4)
3. With progressive nephropathy leading to end-stage renal failure. It is characterized by the onset of spontaneous bone resorption without causal factors. In this case, the patient has multicentric osteolysis (5)
4. Kidney disease that has benefited from the management with ACEI, RRT or renal transplant
5. The appearance of the disease can occur in a period of 9 months to 14 years. Most of the patients present with carpal bone resorption (100%) and the tarsus (60%); it should be noted that all of the bones and joints of the body can be compromised.
6. The patient underwent multiple surgeries due to affected bones and joints. They received a kidney transplant, presenting a successful clinical improvement of renal function and the symptomology.
7. There is only one report of a renal transplant patient with IMO III, from Argentina. With this report, we are reporting the second case in the literature of a patient who underwent a successful kidney transplantation, which is often the only therapeutic alternative for these patients.

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