Patient with Klippel-Trenaunay Syndrome. Case Report of an Intraoperatively Aborted Hip Joint Replacement.

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ABSTRACT
Klippel-Trenaunay Syndrome is a rare congenital disorder. It usually affects one of the lower limbs. It is characterized by the presence of angiomas, venous malformations, hypertrophic varices with arteriovenous communications and hypertrophy of the affected limb, altering in some cases the bone structure. The aim of this presentation is to describe the case of a 54-year-old patient for whom a joint replacement of the left hip had been scheduled for one year and the surgery had to be aborted due to the risk of exsanguination of the patient, and to analyze the literature in this regard.
Key words: Klippel-Trenaunay Syndrome; arteriovenous malformations; massive bleeding; hip.
Level of Evidence: IV

Paciente con síndrome de Klippel-Trenaunay. Reporte de un caso de reemplazo articular de cadera suspendido durante la cirugía

RESUMEN
El síndrome de Klippel-Trenaunay es un trastorno congénito infrecuente. Por lo general, afecta uno de los miembros inferiores. Los hallazgos típicos son angiomas, malformaciones venosas, várices hipertróficas con comunicaciones arteriovenosas e hipertrofia del miembro afectado que, en algunos casos, altera la estructura ósea. El objetivo de esta presentación es describir el caso de un paciente de 54 años en el que se programó un reemplazo articular de la cadera izquierda durante un año y la cirugía debió suspender por el riesgo de exanguinación, y analizar la bibliografía al respecto.
Palabras clave: Síndrome de Klippel-Trenaunay; malformaciones arteriovenosas, sangrado masivo, cadera.
Nivel de Evidencia: IV

INTRODUCTION

Klippel-Trenaunay syndrome is a rare congenital disorder, described in 1900 by French physicians.[1] It usually affects one of the lower limbs. The characteristic findings are angiomas, venous malformations, hypertrophic varices with arteriovenous communications and hypertrophy of the affected limb.

In some cases, the malformation compromises the bone structure and generates intraosseous arteriovenous communications with alteration of the bone architecture. Recurrent hemarthrosis is mentioned as the cause of joint degeneration.

So far, four patients with hip compromise who have been scheduled for joint replacement have been published.[2-5] One of the procedures had to be suspended due to the risk of exsanguination.[5]

The aim of this report is to present a case similar to the one described, in a 54-year-old patient who was scheduled for a left hip joint replacement for one year. The surgery had to be suspended due to the intraoperative risk of exsanguination of the patient.

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CLINICAL CASE

A 54-year-old patient with a diagnosis of Klippel-Trenaunay syndrome that affected the left lower limb and ipsilateral hip osteoarthritis of five years of evolution (Figure 1). The left lower limb had a larger diameter than the contralateral one, and multiple varicose cords with palpable pulse were detected in the groin, ankle, and posterior gluteal region (Figure 2). The patient walked with a cane and the hip flexed; the limb was shortened. He had lumbar scoliosis and lumbago, and the ipsilateral knee was unaffected.

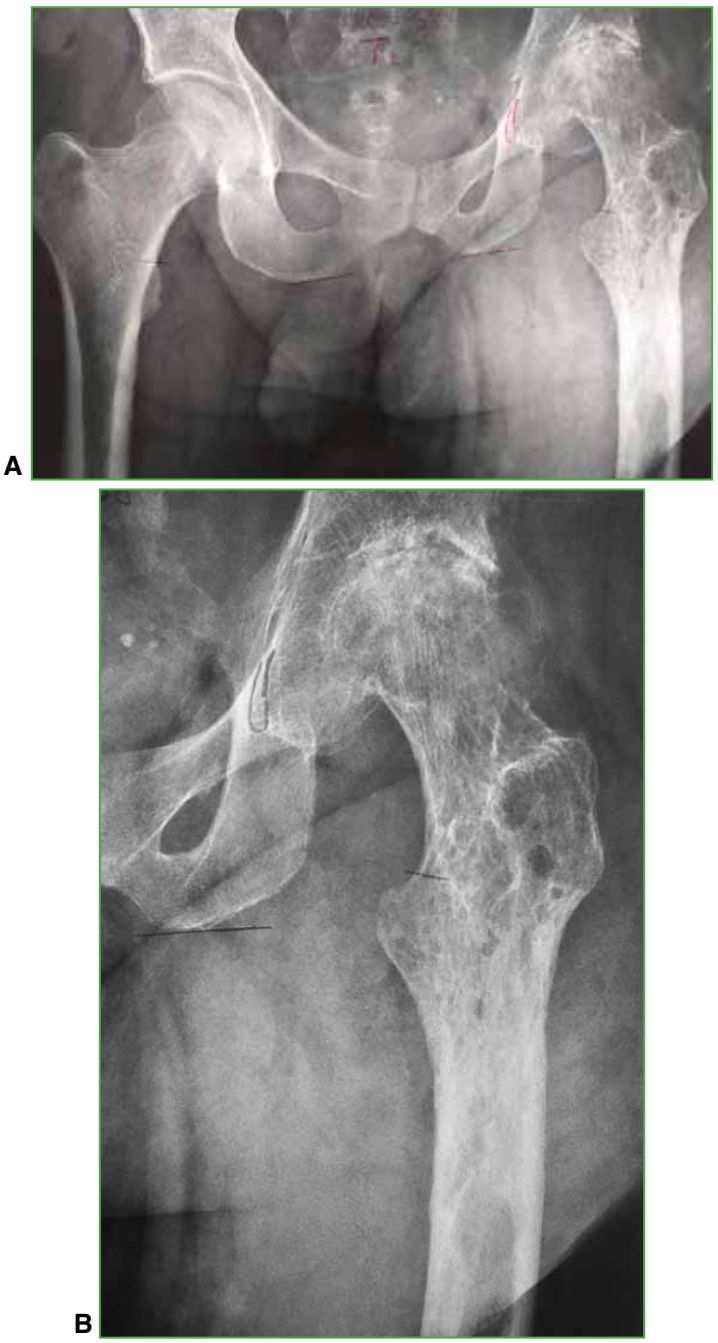

Figure 1. A. Frontal radiographs of both hips. **B.** Frontal radiograph of the left hip. The left hip osteoarthritis is observed, with lytic images in the head, neck and pertrochanteric region that would correspond to the arteriovenous anastomoses.

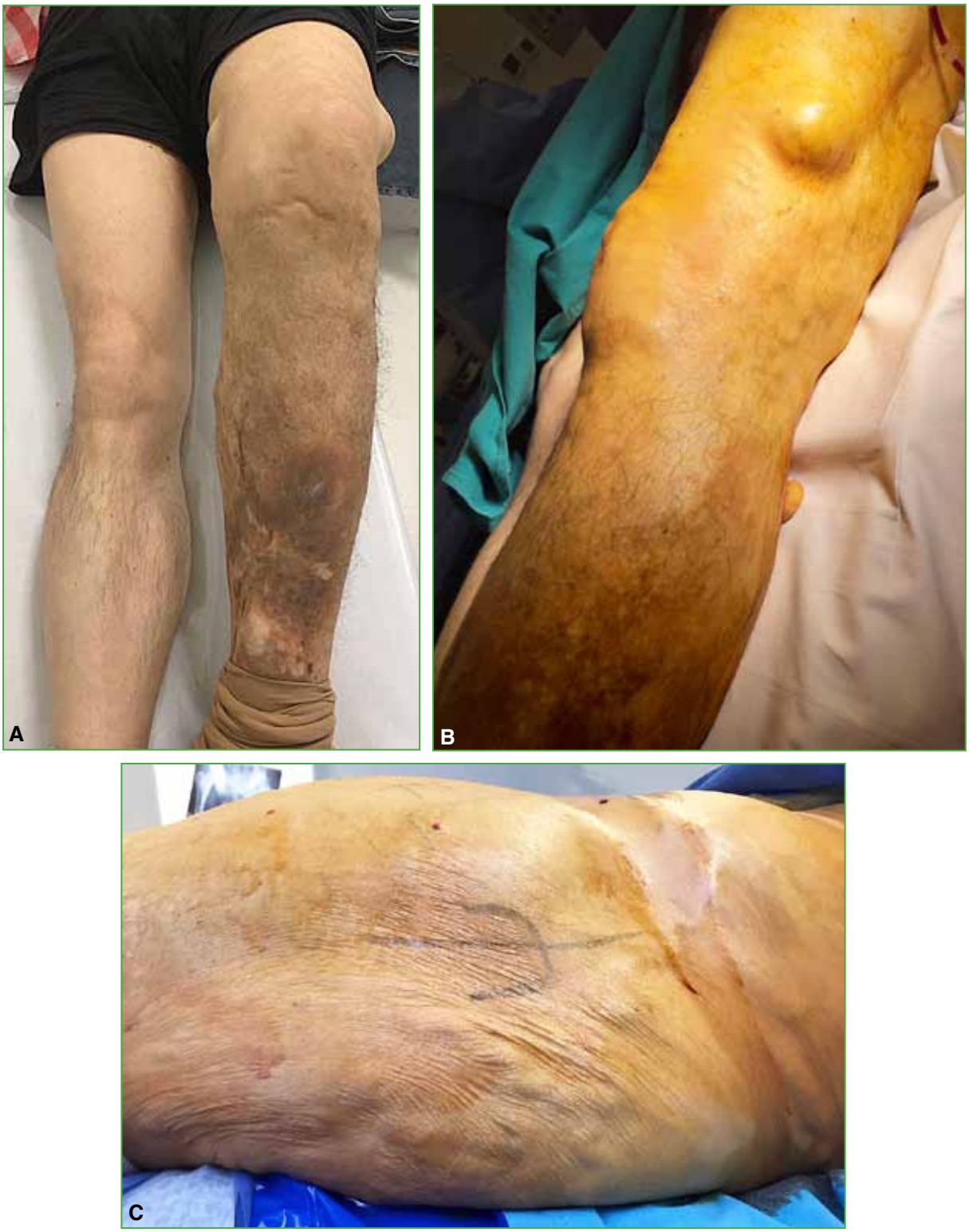

Figure 2. A and B. Image of the lower left limb. It is possible to observe the change in the color of the skin of the leg and tumors on the external and internal aspects of the thigh that correspond to the arteriovenous anastomoses. **C.** The area of the hip to be operated on is observed. The greater trochanter and the anterolateral incision are marked with the patient in dorsal decubitus position. A varicose area is observed in the gluteal region, which makes the posterolateral approach difficult.

The patient underwent arteriography and Doppler ultrasound by the Cardiovascular Surgery team at the hospital. The lateral region was defined as the best approach to the hip (Figure 2), as there were no varicose formations or arteriovenous anastomoses detectable clinically or in the studies.

An MRI was performed, in which the alteration of the bone architecture due to vascular malformations in the head, neck and femoral metaphysis was verified (Figure 3).

Surgery through a direct anterolateral approach was scheduled, with blood supplements and units of blood available for the procedure. The patient was anticoagulated, so the Hemotherapy Team suspended said prophylaxis before surgery.

The placement of a hybrid total hip prosthesis—uncemented cup and cemented stem—was programmed, considering the architecture of the proximal femur (Figure 1) and that, in the MRI (Figure 3), multiple intraosseous images corresponding to arteriovenous fistulas were observed. The idea was to reduce intracanal bleeding with cementation.

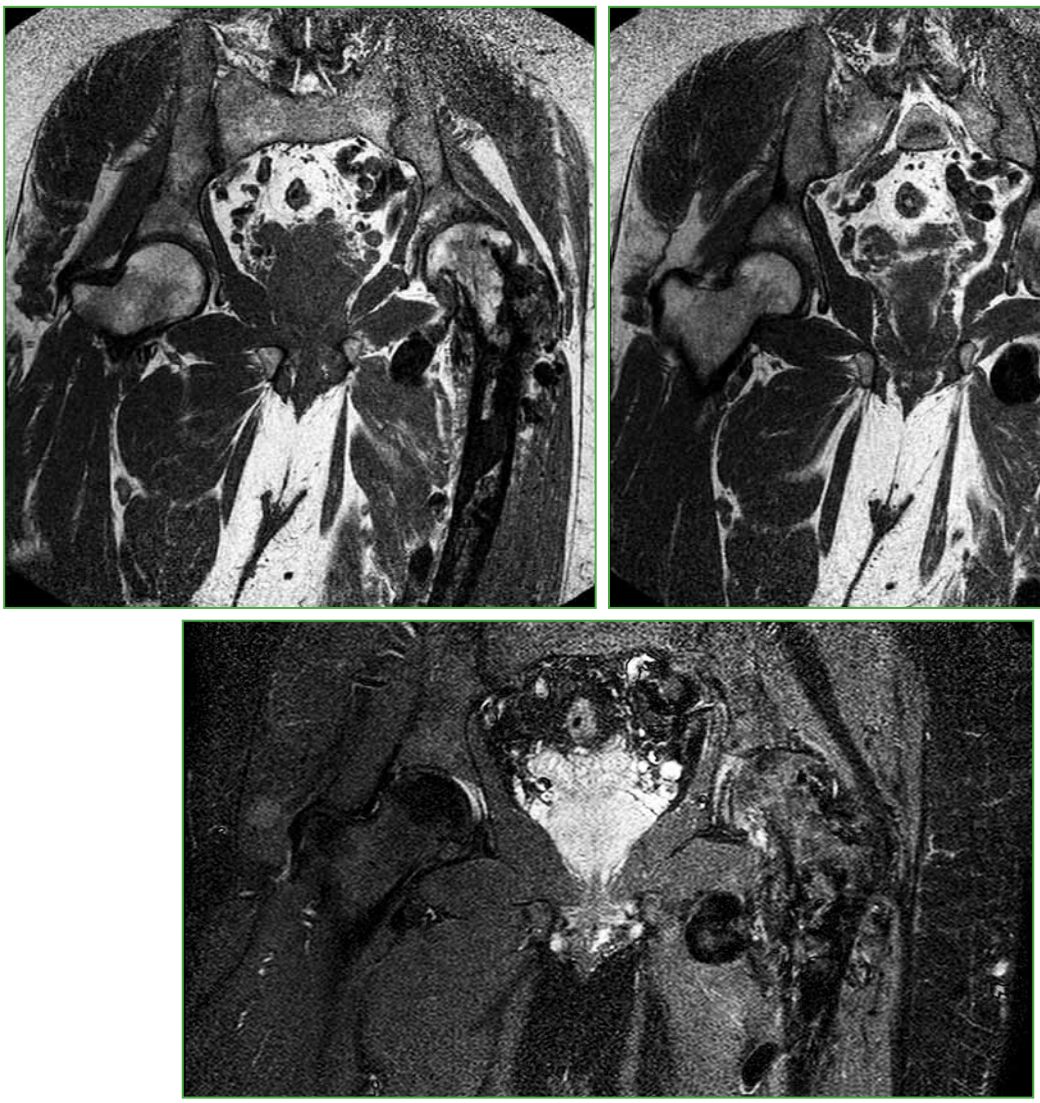

Figure 3. MRI, coronal planes. Blood islets can be verified, especially in the pertrochanteric region, with alteration in bone architecture.

When the approach was performed in the subcutaneous cell tissue, there was abundant bleeding from two venous formations of 1 cm in diameter each, which had to be ligated. During the capsulotomy, there was an intense bleeding stemming from the nutrient foramina of the femoral neck, which was difficult to control. At that time, 1.5 liters of blood had already been collected in the aspiration flask. The vascular surgeons who were in the adjacent operating room were consulted, since it was anticipated that, when performing the osteotomy of the femoral neck, the bleeding could be cataclysmic and incoercible due to the intraosseous vascular malformations. It was jointly decided to suspend the procedure.

The patient had to be transfused during surgery and in the immediate postoperative period. He recovered favorably and was discharged after 72 hours.

DISCUSSION

According to the four published articles, in three cases it was possible to perform the programmed total hip arthroplasty and all had significant bleeding.[2-4]

In the fourth patient, the procedure was suspended at the same moment as in ours—when performing the capsulotomy—due to the risk of exsanguination, and multiple sources of intraosseous bleeding were identified that had not been detected in the arteriography.[5] It should be noted that this was the third attempt; according to the authors, the approach route had been modified by an anterior one, because there were fewer arteriovenous fistulas.

Analyzing the hip radiographs of the published cases, in the three patients in whom joint replacement was completed, the intraosseous alteration was not as significant as in the one in which the procedure had to be suspended and in ours (Figure 1).

A careful approach with dissection and ligation of vessels can limit bleeding, but intraosseous malformations may not be managed during surgery, so the risk of unrelenting bleeding is high. For this reason, it is important to have blood units and blood supplements to replenish, and where possible, a recuperator.

In these patients, thromboembolic events are frequent, with a risk of pulmonary embolism (14%) and deep vein thrombosis (16%).[6,7] Our patient was anticoagulated; therefore, it was necessary to regularize the coagulation values before surgery.

Another issue to consider is the implant to be placed, since the alteration of bone architecture can be considerable. In the case published by Mallick et al.[2], a total cemented arthroplasty was performed in a 58-year-old patient to reduce bone bleeding. García-Juárez et al.[4] used a cementless implant, and highlighted that this could have been the cause of postoperative bleeding in the patient, who had a hemoglobin level of 4 mg on the third day after surgery. It was only possible to achieve hemodynamic stability with transfusions on postoperative day 14, with a hemoglobin of 9 mg. The patient was discharged after 23 days. In our case, the idea was to place a hybrid prosthesis—with a cemented stem—keeping in mind the poor quality of the femoral bone and the arteriovenous fistulas observed in the radiographs and in the MRI of the proximal femur and, in this way, fill the bleeding bone with the cement. Certainly, the cementation would be of poor quality and could alter the survival of the implant.

CONCLUSIONS

These patients are at high risk of bleeding and, consequently, must be carefully studied. The approach must be properly planned and prepared for the ligation of the great venous vessels.

Likewise, it is essential to identify the magnitude of intraosseous malformations—since the decision to perform surgery may depend on it—and the type of implant fixation, bearing in mind that, once the femoral neck osteotomy has been performed, the bleeding can be uncontrollable and life-threatening.

We agree with Cirstoiu et al.[5] in recommending the nonsurgical management of these patients, as long as possible.

Conflict of interests: The authors declare they do not have any conflict of interests.

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