OXFORD

Femoral head necrosis and progressive osteoarthritis of a healed intracapital osteotomy in a severe sequelae of Legg–Calvé–Perthes disease with aplasia of tensor fasciae latae

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INTRODUCTION

The Legg–Calvé–Perthes disease (LCPD) is a rare pathology with an incidence of 4–32 cases per 100 000 but represents one of the most common causes of permanent femoral head deformity in childhood [1]. Almost 30–50% of the affected patients will develop progressive osteoarthritis (OA) in adulthood [2].

Early conservative surgery aims to minimize the femoral head deformity and, in case, the acetabular orientation, to try to reduce the risk of secondary OA [1].

The indications for an effective containment surgery are still being debated, with uncertain predicting factors for the outcomes.

In recent times, the ability to safely perform a surgical dislocation of the hip (SHD) has revolutionized the potential of the treatment of LCPD sequelae [2]. Different algorithms to approach the functional analysis of the hip deformities and eventually select the appropriate surgical treatment, especially for young patients, have been described [2, 3].

The intracapital osteotomy is the ultimate limit of conservative hip surgery and consists of an intracapsular osteotomy of the femoral head intending to restore its spherical shape [4, 5]. The most significant expected risk of such surgery is certainly avascular necrosis (AVN). Few cases have been described in literature [5–9].

A case of a young lady treated with an intracapital osteotomy and concomitant periacetabular osteotomy (PAO) for severe and incapacitating sequelae of Perthes Disease and tensor fasciae latae aplasia is presented. Unfortunately, this case ended in progressive hip OA requiring a total hip replacement.

CASE DESCRIPTION

The patient is a Caucasian woman, 17 years old, suffering from deforming LCPD sequelae of the right hip.

She reported the onset of progressive right hip and knee pain with a limited range of motion (ROM) since she was 7 years old.

After a delay of 2 years, the LCPD had been diagnosed and classified at the fragmentation stage according to Waldenström [10], within the B/C group of the Herring classification [11].

In the following years, the patient had been evaluated clinically and radiographically, every 6 months, without showing clinical worsening.

Nevertheless, at the age of 17, the hip pain became completely disabling, compromising her daily activities. On physical examination, the lower right limb was ~1.5 cm shorter. Both active and passive range of motion of the right hip was painful with a maximum flexion of 80°, abduction lower than 30°, adduction of 20° and a complete loss of the hip rotation. At the clinical evaluation, she was rated a Harris Hip Score (HHS) of 10.4 points, an International Hip Outcome Tool 33 (iHOT33) of 4.3% and a Hip Outcome Score (HOS) of 5.9%.

The right hip was investigated with radiographs (Fig. 1) and with a 3 D CT scan reconstruction (Fig. 2), showing a severe femoral head deformity as opposed to an undeveloped acetabulum which entailed a severe articular incongruence and a Tönnis grade 1 OA [12].

The Magnetic Resonance Imaging (MRI) showed an 8 mm focal IV-degree chondropathy associated with an erosion of the apical profile of the head, without signs of osteonecrosis, associated with an erosion of articular cartilage of the acetabulum without signs of a subchondral bone deficit.

Fig. 1. Pelvic antero-posterior (left) and right hip axial (right) view of the young patient. The severe joint deformity is classifiable as aspherical congruency or Class IV (Group 2) according to the Stulberg classification. The proximal femoral epiphysis morphology is definable as a coxa breva and magna with lateral elongation of the head of the femur (Type A according to the Paley Foundation Classification). The Femoral Head Extrusion Index is 55%, with a short femoral neck and a relative higher and larger GT. The acetabulum shows a secondary dysplasia, with a Sharp Angle of 52°, a negative Lateral Center Edge Angle and an Acetabular Index of 12°. The residual dysplasia can be included in the Class IV (subluxation) according to the Severin Classification. The Alpha Angle measured on the Antero-Posterior radiograph is around 58°.
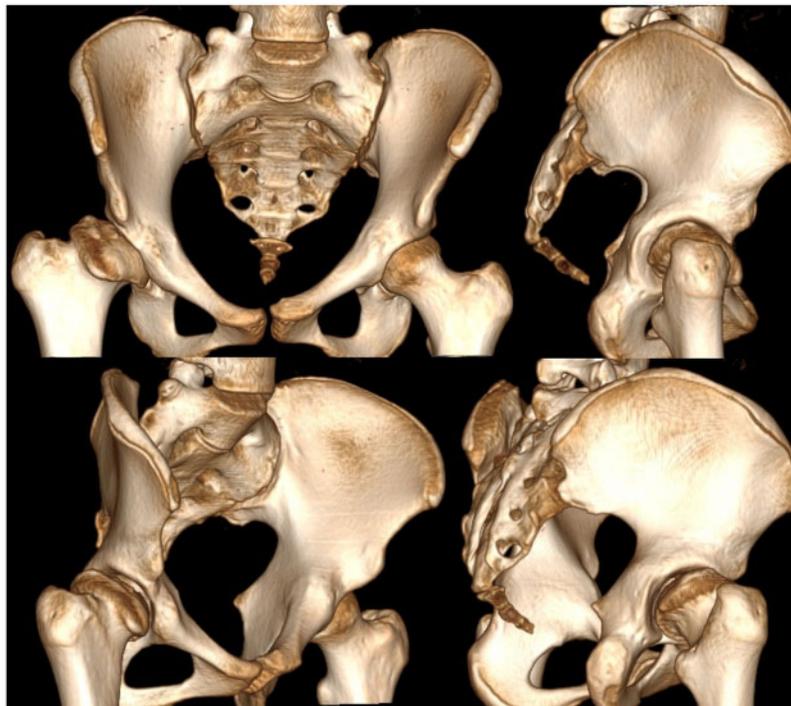

Fig. 2. High-resolution CT scan with 3D reconstruction of the right hip, enlightening the complex intra- and extra-articular deformities.

The patient and her family refused a total hip replacement.

A pre-operative CT based planning of femoral head reduction was performed using the custom-made software application CASPA (Balgrist ROCS, Research in Orthopedic Computer Science, University Hospital Balgrist, Forchstrasse 340, 8008 Zurich, Switzerland; Fig. 3), and a combined surgical dislocation and periacetabular osteotomy planned [3, 13].

Intraoperative findings and surgical technique

The patient and her parents were informed about the procedure and all the possible complications.

The operative plan consisted of a two steps procedure. First, a Femoral Head Reduction Osteotomy (FHRO) and a Relative Femoral Neck Lengthening (RFNL) through a surgical hip dislocation (SHD). Second, a periacetabular osteotomy (PAO) through a modified Smith Petersen Approach.

A longitudinal incision was performed over the anterior third of the greater trochanter (GT), with the patient in a lateral decubitus position. The fascia was then identified and incised just above the Gluteus maximus anterior border, proximally.

First, internally rotating the hip, the posterior edge of the ischial tuberosity was exposed, and the first ischial cut of the periacetabular osteotomy performed under direct visualization and protection of the sciatic nerve.

Then the posterosuperior border of the GT was exposed, and an osteotomy was performed with an oscillating saw. A 1.5-cm thick mobile trochanteric fragment remained attached to the gluteus minimus, gluteus medius and vastus lateralis muscles.

After retracting the trochanter anteriorly, the hip capsule was isolated with the hip externally rotated, and an anterosuperior Z-shaped capsulotomy was performed. The femur was then dislocated, cutting the ligamentum teres.

The femoral head cartilage loss was more severe than expected and a mild degeneration of the labrum and the acetabular cartilage was present as well. Furthermore, the acetabulum showed an irregular triangle-shaped morphology (Fig. 4).

Before proceeding with the planned surgical steps, the hip was reduced, and a cautious extended retinacular soft-tissue flap containing the retinacular vessels to the femoral head was developed and elevated from the femoral neck.

The hip was dislocated again, and the double femoral head-neck osteotomy was performed safely following the computerized pre-operative planning. An enlarged necrotic central portion of the femoral head was then removed (Fig. 5).

The superior femoral head-neck vascularized fragment was then perfectly reduced to the stable inferior head-neck with two 3.5 mm screws. The residual bone gap at the superior base of the neck was filled with the bone fragments conserved from the central femoral head fragment removed.
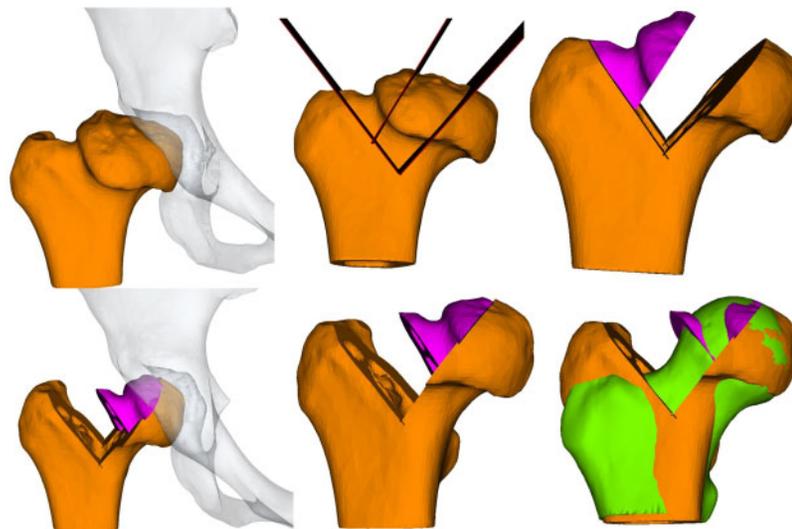

Fig. 3. Pre-operative computerized planning simulation of the femoral head reduction osteotomy with the custom made software application CASPA (Balgrist ROCS, Research in Orthopedic Computer Science, University Hospital Balgrist, Forchstrasse 340, 8008 Zurich, Switzerland). The bottom right image shows a comparison between the postoperative morphological result (purple and orange) and the contralateral hip (green).

This procedure produced a smaller and round femoral head, with an improved range of motion, but still insufficiently covered by the acetabular roof.

The trochanteric distalization was competed and fixed with two 3.5 mm screws obtaining a relative neck lengthening. The lateral approach was closed.

The patient was then positioned in supine decubitus, and a periacetabular osteotomy (PAO) was performed (Fig. 5) through a modified Smith–Petersen approach. Interestingly, the tensor fasciae latae (TFL) could not be
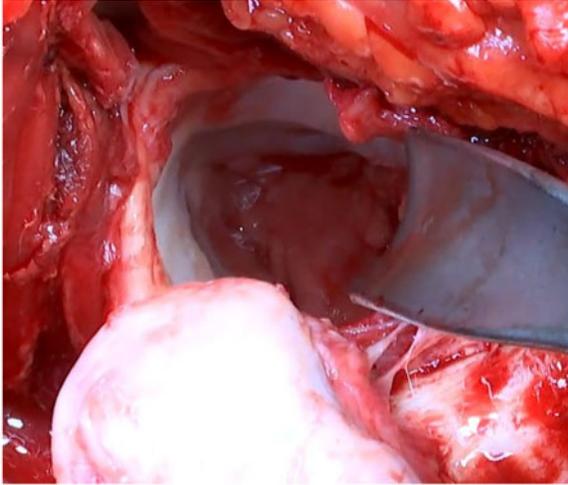

Fig. 4. Intraoperative view of the acetabulum through the surgical dislocation characterized by a wide acetabular fossa and a pathological triangular shape.

found during the approach due to an apparent previously unrecognized congenital aplasia. Bilateral TFL aplasia was then confirmed with MRI.

At the end of the procedure, a round femoral head well covered by the acetabulum was confirmed with a pelvis X-ray (Fig. 6).

Post-operative rehabilitation

The patient was allowed to walk with restricted toe-touch weight-bearing for 8 weeks from the third post-operative day. She started, immediately, an assisted physiotherapy rehabilitation. Active flexion, abduction and rotation of the right hip were prohibited for 6 weeks. At 10 weeks postoperatively, the patient achieved a normal ROM and was able to walk with full weight bearing.

Clinical result

The patient has been evaluated with a strict follow-up reporting good pain management, good improvement of the hip ROM and strength. Four months after the surgery, she could go up and down the stairs, sit comfortably and take short walks. The clinical scores showed an improvement in her daily life (HHS: 41.8; IHOT33: 13.5%; HOS: 30.9%).

In the meantime, a progression of the OA was visible on the X-rays (Fig. 6).

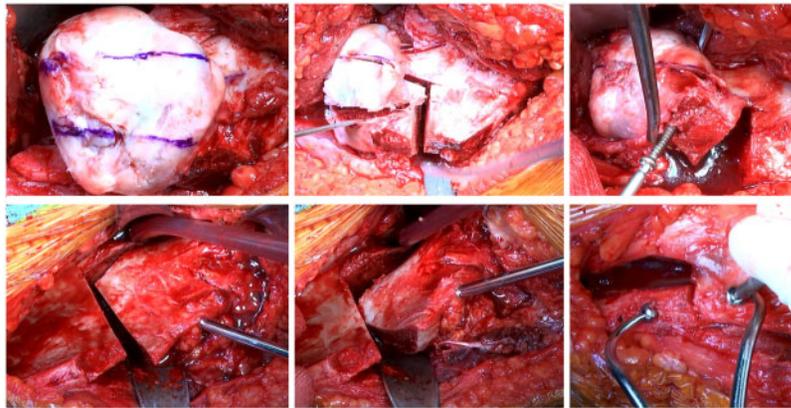Unfortunately, 6 months after the surgery, the patient reported an abrupt clinical worsening.

Fig. 5. Intraoperative findings of the first surgery. The upper row images show the surgical steps of the Femoral Head Reduction Osteotomy (FHRO). The femoral head osteotomies have been performed on the sagittal plane as illustrated in the computerized planning. In the first picture on the left, the femoral head is viewed from the medial edge, leaving on top of the picture its anterior edge. The middle and right pictures show the femoral head from an anterior view, leaving on top of the picture the inferior edge of the femoral head and neck. The bottom row images show the surgical steps of the Periacetabular Osteotomy, performed through the modified Smith–Petersen approach. In all the bottom row pictures, the proximal side is on the left and the distal side on the right.

The MRI showed good sphericity of the femoral head but marked edema of the cancellous bone of the head and neck with a small apical osteonecrosis area associated with a further progression of the OA were present (Fig. 7).

The patient followed one cycle of hyperbaric oxygen therapy without any clinical or MRI improvement.

Almost 12 months after the first surgery, the right hip pain became utterly disabling. She could not walk without crutches. On physical examination, ROM was severely limited with low clinical scores.

The patient underwent a total hip arthroplasty (THA) conversion, with a polyethylene-ceramic bearing surface, through an extended direct anterior approach (Fig. 8). The acetabular osteotomies were healed entirely, and all the hardware was removed. The acetabulum showed a double floor due to medial wall osteophytes and a critical triangular shape. There was complete healing also of the femoral osteotomy but severe apical wear (Fig. 8).

The recovery of the right hip was completed during the following 4 months. The patient returned to practice physical activities such as swimming, yoga and fitness classes.

One year after the total hip arthroplasty, the patient was happy and had complete and painless ROM with satisfactory clinical scores (HHS: 83.1; IHOT33: 62.7%; HOS: 70.0%; HOS ADL 91.2%; HOS Sport: 25.0%; Fig. 9)

DISCUSSION

In this article, a case of incapacitating LCPD sequelae in a young female who failed either for progressive OA or limited AVN is presented.

There is a massive lack of predictive factors for conservative surgical treatment of LCPD sequelae [13].

The ability to safely dislocate the hip revolutionized the potential for conservative treatment of hip deformities related to the LCPD sequelae, offering the ability to address any possible source of intra- and extra-articular irregularity [14].

The acquired in-depth knowledge of the vascular supply of the femoral head also allowed to design a technique to develop the extended retinacular flap [4, 15]. The
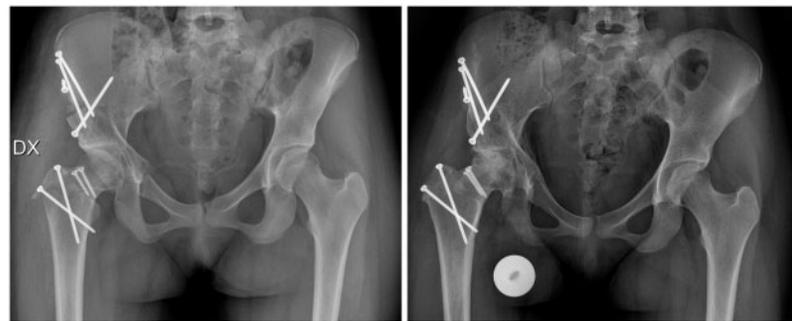

Fig. 6. Radiological follow-up. On the left, the antero-posterior pelvic radiograph at 4 weeks after the surgery, showing a good healing phase of the osteotomies and an excellent appearing round contour of the right femoral head. On the right, the antero-posterior pelvic radiograph at 10 months after the surgery, showing a complete healing of the osteotomies but a severe progression of the right hip osteoarthritis.
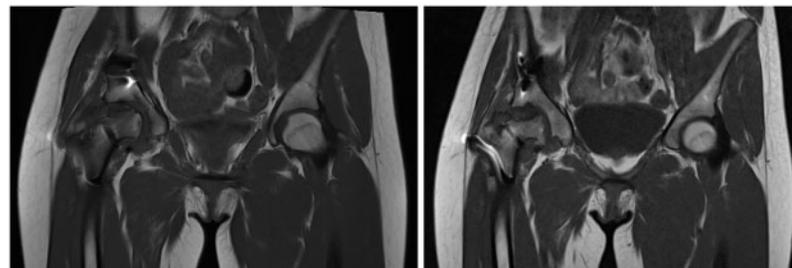

Fig. 7. Right hip Magnetic Resonance Imaging on coronal planes. On the left, at 6 months after the surgery showing a good sphericity of the femoral head but a marked edema of the cancellous bone of the femoral epiphysis with an osteonecrosis area. On the right, 8 months after the surgery, and after the hyperbaric oxygen therapy, showing a right hip osteoarthritic progression and the osteonecrosis area at Stage 4 according to Ficat and Arlet classification.
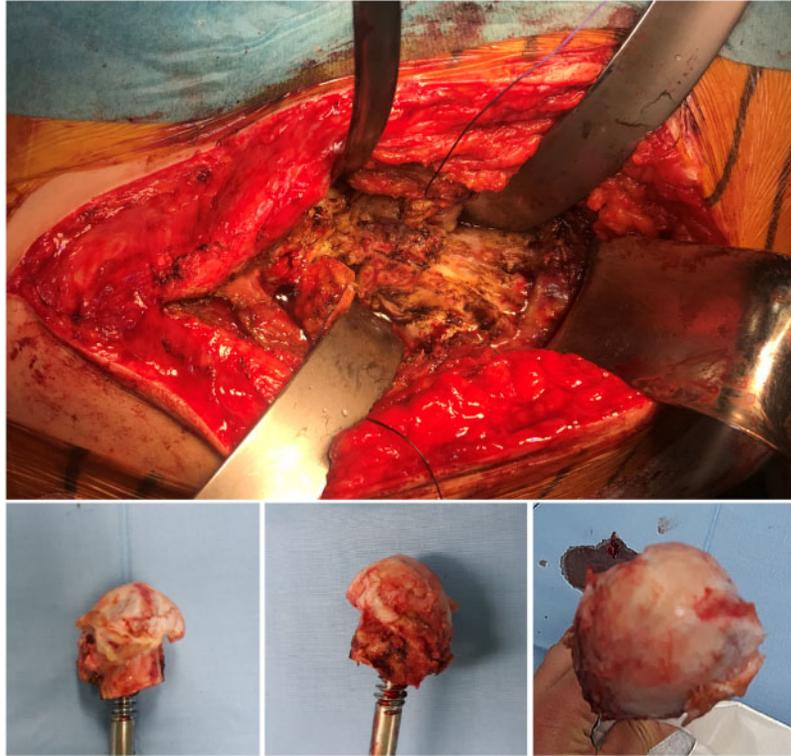
Fig. 8. Intraoperative findings of the second surgery through an extended direct anterior approach. On the bottom line images, the femoral head showed a complete healing of the osteotomy and of the cartilage junction but a severe apical wear.

Fig. 9. Pelvic antero-posterior radiograph at 1 year after the total right hip arthroplasty.

posterior extended retinacular flap contains a single tissue sleeve, the medial femoral circumflex artery (MFCA) and the retinacular vessel.

This technique provides safe access to the femoral neck, allowing the safe performance of new intracapsular osteotomies such as the femoral head reduction osteotomy [5, 16].

Tannast and colleagues offered a systemic approach to carefully analyse the femoral and acetabular deformities better to understand the underlying functional problem [2]. They also suggested detailed surgical plans to address each and single deformity, knowing that restoration of completely normal anatomy is rarely possible.

An algorithm to help clinicians assessing patients with complex hip deformities and identify patients who can benefit from proximal femoral and periacetabular osteotomies was published [3]. According to this algorithm, the clinical results of 53 consecutive patients showed a significant improvement of pain, range of motion, limping, with no case of osteonecrosis of the femoral head. In another study, patients treated according to the same algorithm showed a cumulative survival of 86% at 5 years and 61% at 8 years of follow-up. The authors concluded that the restoration of normal hip anatomy is rarely possible, but the progression of OA is related to the preexisting articular damage [13].

However, in their case series, the typical surgical treatment rarely consisted of a femoral head reduction osteoplasty, and the main contraindication for surgery was an OA grade 1 or greater, according to Tönnis. They also described 11 univariable predictors for poor outcomes with corresponding hazard ratios for each one, divided into three categories: demographic, pre-operative and post-operative [13].

They suggested that the post-operative midterm results of the joint preserving surgery are mostly affected by the class of deformation of the hip according to the Stulberg Classification [17], the pre-operative grade of OA, the age of the patient, the accuracy of the correction, and the presence of a post-operative broken Shenton's line.

According to these findings, the patient present here should have benefited from a hip preserving surgery because of her young age and good preservation of the hip abduction. Unfortunately, the patient presented also plenty of poor predictive factors: a Tönnis grade of OA greater than one, an acetabular index lower than 14°, an alpha angle higher than 56°, low clinical scores, a dysplastic morphology with subluxation of the femoral head (Class IV according to the Severin classification [18]) and a particularly severe deformity of the hip (Class IV according to the Stulberg Classification).

Furthermore, as this case demonstrated, another major challenge, managing hip joint incongruence in LCPD sequelae, lies in the acetabular deformity [19]. In fact, in these cases, the acetabulum, in addition to flattening, can develop a triangular shape as a consequent adaptation to the deformity of the head, making it impossible to restore a correct congruency with the new femoral head, despite the reorientation obtained by the osteotomy.

Therefore, the dysmorphism of the acetabulum appears to be another parameter, which should be considered before undertaking such a complex conservative surgery. However, the triangular shape of the acetabulum could not be considered a crucial factor for failure or rapid articular degeneration since containment has a higher value than articular congruency even in long-term outcomes [9].

Considering this critical biomechanical configuration, the cranial aspect of the acetabular roof would have less load transmission, leaving unexplained the arthritic changes on the apex of the head and the absence of damage to the acetabular cartilage. As a matter of fact, in our case, the acetabular damage lies in the joint periphery due to an inclusive impingement, of the flattened head, in flexion and abduction.

To the best of our knowledge, this is the first documented case of AVN after a femoral head reduction [7]. Interestingly, during surgery, the mobile head fragment was tested viable through small microfracture holes that showed subchondral bone bleeding.

Nevertheless, in hips affected by LCPD, the lateral epiphyseal arteries' blood supply may be impaired at their origin, leading to underrepresented and compromised retinacular vessels [20]. The vascular penetration regenerative process may also be absent in the femoral head's weight-bearing portion, right below the acetabular roof, enlightening the need for special care and effort required to assess, pre- and intra-operative, the femoral head perfusion before the FHRO [20].

To be emphasized finally, the young patient also presented bilateral agenesis of the tensor fasciae latae. This aplasia was discovered intraoperatively, confirmed on the MRI before the THA (Fig. 10), and retrospectively observed on the MRI before the first surgery. This is an exceedingly rare condition that should be known before any anterior approach to the hip where the tensor fasciae latae is the prominent landmark. Neither the patient nor her parents suffered from other congenital known muscular aplasia or signs of neuromuscular diseases [21]. The aplasia of the tensor of the fasciae latae muscle could be only mere randomness but led us to question if this anatomic vascular aberration could influence the entire hip region thus, on the mobile fragment insufficient perfusion and AVN development.
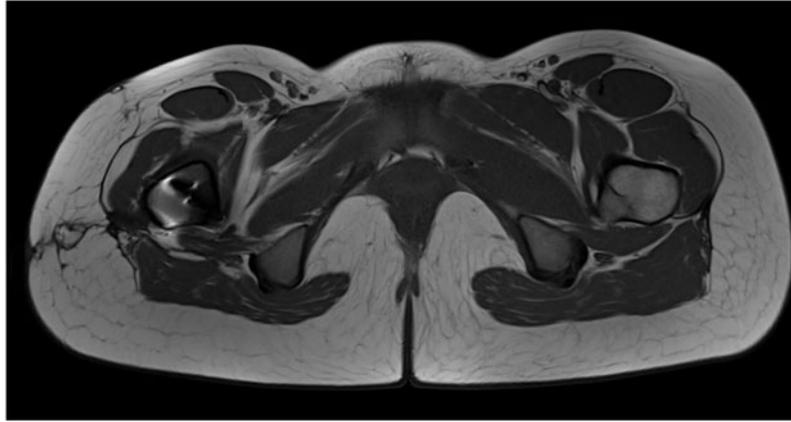
Fig. 10. Pelvic Magnetic Resonance Imaging showing the complete bilateral agenesis of the tensor of the fasciae latae muscles.

As suggested by the peculiarity of this case, before performing an FHRO, a super-selective MRI perfusion study of the retinacular vessels should have been recommended. Notwithstanding, still unclear remains the question if the AVN was a result of the surgical procedure, a consequence of the femoro-acetabular incongruency, a fragile and unfavorable anatomical hip vascularization somehow related to the tensor aplasia, or else. Impossible to be known. Neither is possible to know the main reason for this structurally well-performed osteotomy failure. The excessive, not reversible articular cartilage damage, the overcoming AVN or both?

CONCLUSION

LCPD sequelae can be disabling and complex to treat, especially in a younger patient where a replacement is not well accepted.

The FHRO, associated with PAO, is a valid surgical indication in patients with congruent acetabular morphology and without substantial degenerative joint changes. Unfortunately, in cases with unfavorable prognostic factors, the conservative surgery chances of success fall off. Even though a failure for a femoral head avascular necrosis is a rarity, the condition of agenesis of the TFL, having never been described before, may imply the entire region's blood supply affecting this FHRO outcome.

DATA AVAILABILITY

The authors confirm that the data supporting the findings of this study are available within the article.

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