

## Knee chondrosarcoma secondary to Ollier's disease: a report of one case with twelve-year follow-up

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### Abstract

Management of low-grade chondrosarcomas secondary to Ollier's disease is generally radical. We report here a case of conservative treatment of a knee tumor with reconstruction of the lower femoral extremity with a hinged total knee prosthesis, cemented and coupled with a massive femoral allograft, with a twelve-year follow-up.

### Introduction

Ollier's disease, defined as the simultaneous presence of at least three chondromas with an asymmetric location, leads in 10% up to 25% of cases to a sarcomatous degeneration.<sup>1</sup> Risk factors are early age of onset of the first chondromas, their width in relation to the metaphysis and their location at the distal extremity of the femur. Several authors in these circumstances advise high femoral amputation or coxo-femoral disarticulation because of the malignant nature and the major bony deformities. We report a case of sarcomatous degeneration of a chondroma of the distal end of the femur in progressive Ollier's disease, which resulted in a major loss of bone substance in the involved leg. Treatment was by resection and reconstruction with a hinge prosthesis coupled with a massive distal femoral allograft, and the patient has now been followed up for twelve years.

### Case Report

A 56-year-old woman had Ollier's disease discovered during childhood. She had two pathological fractures of the right upper femoral shaft treated surgically at the age of 6 and 8 years. Also on the right side, she had undergone tibial osteotomy to straighten the leg, which resulted in major proximal metaphyseal malunion in the sagittal plane (anterior slope of about 30°). Epiphysiodesis had been performed on the contralateral leg when

the patient was aged 11 to decrease limb-length discrepancy. She was regularly followed for a metaphyseal chondroma of the distal end of the right femur, where recent changes suggested sarcomatous degeneration (increased pain and volume).

The patient's height was 1.45 m and she had a limb-length discrepancy with the affected limb being 10 cm shorter and with marked genu varum morphology. Walking became increasingly difficult and her walking distance was reduced to the vicinity of her home. She also had a regular activity which required prolonged standing. Radiographic investigation to assess extension (plain radiographs, computed tomography and magnetic resonance imaging) carried out because of the increased pain showed a metaphyseal tumor of the right distal femur with irregular popcorn calcification and antero-posterior cortical osteolysis, and major periosteal reaction without soft tissue infiltration. The tumor measured 12×17 cm in the axial plane and 12.3 cm in height (Figure 1). Secondary locations had been excluded by clinical and radiographic assessment of extension. After multidisciplinary discussion, a surgical biopsy specimen was obtained to determine the stage of progression of the chondroma. Cartilaginous tumor cells were observed, with a predominantly chondromyxoid matrix whose morphological appearance was consistent with a grade I chondrosarcoma. Bacteriological tests were sterile. Amputation at the root of the limb was then proposed by several teams.

Using an anterior midvastus approach (approaching in the medial plane and preserving the vascular axes and extensor mechanism), we carried out conservative resection for prosthetic reconstruction. After total resection of the right distal end of the femur, we replaced the knee joint with a hinge total knee prosthesis, cemented and coupled with a massive distal femoral allograft measuring 17 cm. The prosthesis made it possible to restore an hip-knee-ankle angle within the normal range (180° versus 167° preoperatively) (Figures 2,3). Because of the tibial deformity, it was technically impossible to use an intra- or extramedullary tunnel. Our aim was to obtain a tibial plate that was perpendicular to the anatomical axis of the tibia, at the expense of good adaptation of the tibial pin to the shape of the malunion. We used a navigation system to position the tibial resection. The stem of the tibial implant lay in an unorthodox position with cortical effraction but provided a satisfactory overall axis for the prosthesis. The patella was resurfaced. Surgical margins were clear (pathological findings R0) and no adjuvant treatment was given. The patient was discharged seven days postoperatively. A 2-cm superficial area of necrosis at the lower part of the scar was excised two months postoperatively without sequelae. The patient had post-

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operative physical therapy and resumed full weight-bearing at three months. Six months postoperatively, the result was satisfactory with a functional extensor mechanism and full weight-bearing without assistance. At the 12-year follow-up, the patient was free from pain; she presented a 7-cm compensated leg-length discrepancy, walked without a limp and active knee flexion was 90°. There was no residual flexion deformity. The International Knee Society rating system (IKS) score was 93, IKS function score 80 and total IKS score 173/200. The patient was again fully independent and had no recurrence or malignant degeneration. She considered that she had benefitted from the procedure. Radiographs showed that the allograft was consolidated at the junction without osteolysis.

### Discussion

Ollier's disease is a rare and non hereditary bone disease that affects men and women equally. It results from a somatic mutation (non-mendelian transmission) and the question of the gene or genes involved (mutant PTHR1 (R150C))<sup>2</sup> is still debated.<sup>3</sup> Chondroma is a benign cartilaginous tumor that escapes growth regulation and impairs normal bone development, often in a metaphyseal location.<sup>4</sup> Its gravity is related to potential malignant progression to chondrosarcoma.<sup>5</sup> Involvement often predominates on one side and can lead to varying degrees of deformity, limb length differences or pathological fractures.<sup>6-8</sup>

As Ollier's disease is rare, the surgical literature on its management in adults is sparse, and there are even fewer publications on the

degenerative stage of low-grade chondrosarcoma. These tumors are distinguished by their lower histological grade (60%) according to the classification of O'Neal and Ackerman compared with other chondrosarcomas.<sup>9,10</sup> Surgical intervention is central to the management of these patients because of the low efficacy of adjuvant treatments (chemotherapy and radiotherapy).<sup>11</sup> Ten-year survival is  $73\pm 16\%$  whatever the tumor site, and  $87.5\pm 10\%$  in distal femoral sites and lower histological grade tumors.<sup>12</sup> Degenerative changes occur at a mean age of 30.7 years to 45 years depending on the series.<sup>4,13</sup>

Although such results could encourage conservative treatment, we mainly find radical amputation reported in the various series on grade I chondrosarcomatous degeneration of the distal femur in adults with enchondro-

matosis. This appears surprising, as we know that amputation and conservative surgery yield comparable oncological results in malignant bone tumors and that secondary chondrosarcomas are generally less aggressive,<sup>14,15</sup> allowing conservative treatment if carcinological resection leaves clean margins. Conservative surgery enables restoration of functional joint mobility, better than amputation and knee replacement or disarticulation,<sup>16</sup> with the advent of hinged prostheses.

In the literature, we find six patients with Ollier's disease surgically treated for low-grade (grade I) chondrosarcoma of the distal end of the femur. Four of these patients were reported by Liu *et al.*<sup>17</sup> Treatment consisted of two transfemoral amputations, one coxofemoral disarticulation and one tumor resection. Reconstruction procedures were not specified. Survival was not correlated with surgical procedure, and at the minimum follow-up of 8 years there was a single death, the patient who had undergone amputation.

Vázquez-García *et al.*<sup>4</sup> more recently reported two similar cases treated by resection and who were both living at 17 years, although no details on reconstruction procedures were given. With regard to grade II chondrosarcomas, Schaison *et al.*<sup>13</sup> reported 4 patients who underwent two disarticulations, one amputation and resection followed by reconstruction with a total knee prosthesis, but with no indication of the nature of the procedure. One patient was lost to follow-up, two are living at 60 and 83 months follow-up while one patient who had undergone disarticulation died at 14 months.

The hinged total knee prosthesis coupled with a massive allograft of the distal end of the femur is an interesting treatment, even

though it has been abandoned by some teams after several failures (infections, pseudoarthrosis and fractures).<sup>18</sup> The survival curve of massive distal femur allografts is moderate to good, depending on the series (65% to 78% at 10 years).<sup>19,20</sup> This treatment restores bone stock that is useful for the future management of these young patients, avoids an amputation and allows early recovery (total weight-bearing at 3 months postoperatively).

## Conclusions

In conclusion, Ollier's disease is a rare disease whose principal complication is chondrosarcomatous degeneration which needs to be monitored and also treated. Low-grade degeneration of distal femur can be treated conservatively, but we have found no previous report in the literature. In spite of the technical difficulties, we need to be able to propose an alternative to radical treatment for these patients.

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Figure 1. Magnetic resonance imaging of sagittal section. Increased signal intensity of the tumor on T1 and T2-weighted sequences with popcorn appearance and cortical osteolysis, without soft tissue infiltration.



Figure 2. Satisfactory bone consolidation, no osteolysis at the junction.



Figure 3. The tibial slope persists due to the severe structural defect of the metaphysis.

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