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Ollier's Disease Treated with Grafting Using Alpha-tricalcium Phosphate Cement. A Case Report

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ABSTRACT

Ollier's disease is a rare disorder characterized by multiple enchondromas with a unilateral predominance, especially in fingers in early childhood. We experienced a case of Ollier's disease treated four times with simple curettage from the age of 2 years and alpha-tricalcium phosphate cement grafting at the age of 21 years. The forth curettage was performed when the patient was 15 years old and preoperative X-rays had shown remarkable finger deformities. Postoperative casting of the involved joints was necessary to prevent fractures but led to some finger contractures. Following X-rays demonstrated incredible improvement of the appearance. This suggests that simple curettage alone at an early stage of Ollier's disease provide cosmetic improvement.

At the age of 21 years our patient showed enlargement of the intramedullary finger lesions again. Some lesions seemed to be impending pathological fractures. This time we chose alpha-tricalcium phosphate cement to fill the cavities after curettage had been carried out. Harvesting a large amount of autologous bone was not required. All activities of daily life could be resumed immediately after surgery and none of the finger joints showed further restriction of motion. X-rays taken three years after the operation showed new bone ingrowth surrounding the material with little evidence of absorption. To our knowledge, this may be the first case of Ollier's disease treated with artificial bone grafting reported in the English literature.

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INTRODUCTION

Ollier's disease is a rare disorder characterized by multiple enchondromas with a unilateral predominace that usually occurs in infancy or early childhood. Diaphysis and metaphysis of tubular or long bones, particularly fingers, are usually affected, and the lesions are often associated with remarkable deformities and dysfunctions. We experienced a case of Ollier's disease in a patient with multiple enchondromas in the upper and lower extremities from the age of 2 years. After simple curettage of the finger lesions had been carried out four times, the patient was recently operated on with artificial bone grafting using alpha-tricalcium phosphate cement. To our knowledge, this is the first case of Ollier's disease treated with artificial bone grafting reported in the English literature. In this case report, the clinical results are presented.

CASE REPORT

The patient was a 21 year-old male. He had undergone an auriculoplasty of the right coloboma lobuli at the age of 10 years and conservative treatment of spontaneous pneumothorax at the age of 20 and 21 years respectively. His mother noticed deformities of his right index finger when he was 2 years old. Bone tumors on the proximal phalanx of the right index finger and second matacarpal bone were found and curetted away. Microscopical examination revealed enchondromas. He was diagnosed as having Ollier's dis-

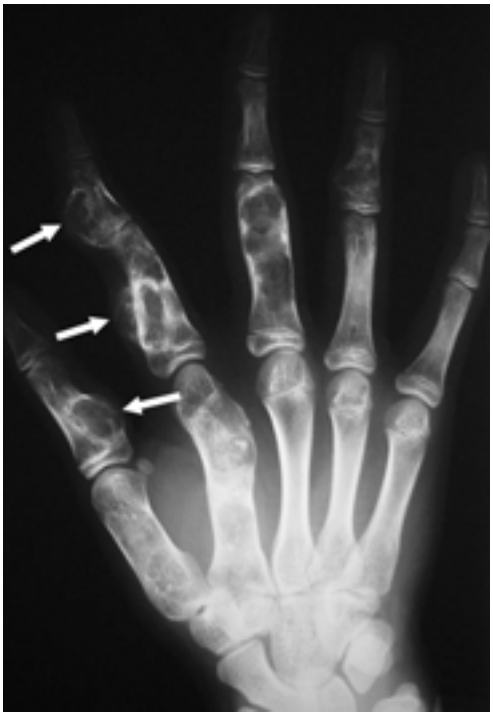

Fig 1. Preoperative X-ray at the age of 15 years. Multiple enchondromas of the right fingers are shown. Some lesions are associated with severe deformities (arrow).

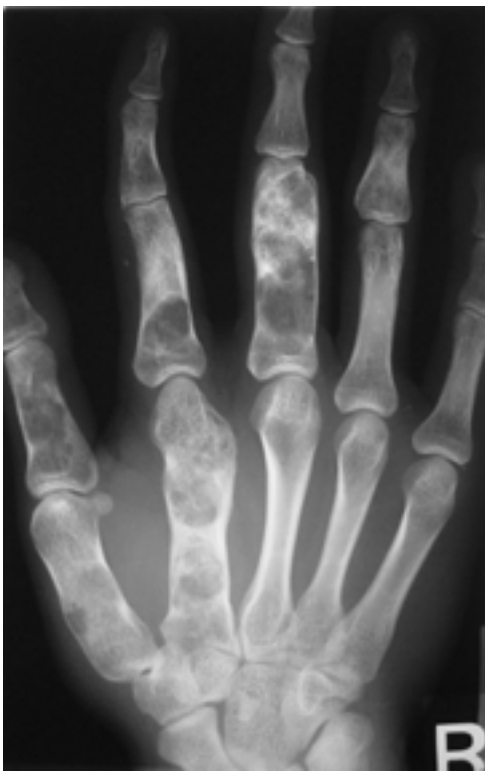

Fig 2. Preoperative X-ray at the age of 21 years. Although the severe deformities were apparently improved, intramedullary radiolucent lesions had increased in size.

ease, and underwent curettage of the lesions in the second metacarpal on the right hand at the age of 6 years, the middle phalanx of middle finger and second metacarpal when he was 9 years old, and the proximal phalanx of the thumb and middle / proximal phalanx of the right index finger at the age of 15 years. At that occasion, preoperative X-ray showed radiolucent bone tumors in the first metacarpal and proximal phalanx of the right thumb, middle / proximal phalanx of the right index finger, and the right second metacarpal, and proximal phalanx of the right middle finger, and middle phalanx of the right ring-finger. Some of these lesions demonstrated ballooning of the surrounding bone cortices and severe deformity (Fig 1). After tumor curettage, casting for 4 weeks was employed to prevent postoperative pathological fractures.

At the age of 21 years, he visited our clinic again with the complaint of swelling of the thumb, index / middle / ring-finger of the right hand. There were operative scars on his thumb, index / middle finger and mild restriction of motion in these involved joints. Plain X-ray showed intramedullary radiolucent lesions in the proximal phalanx of the thumb and first metacarpal bone, middle / proximal phalanx of the index finger, second metacarpal, and proximal phalanx of the middle finger, and middle phalanx of the ring-finger (Fig 2). Bone scintigraphy identified high-intensity radionuclide uptake not only in the right hand but also in the right scapula / humerus / radius / rib cartilage and left rib / femur (Fig 3). The lesions in the right hand seemed to be impending pathological fractures. He underwent tumor curettage of the right proximal phalanx of the thumb, first

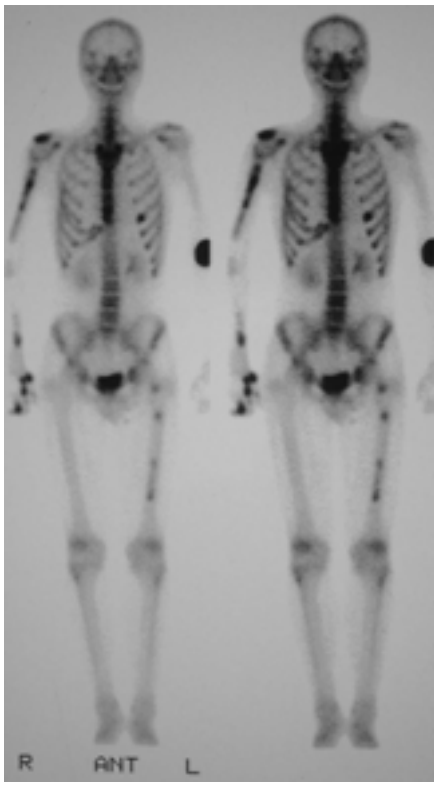

Fig 3. Bone scintigraph.

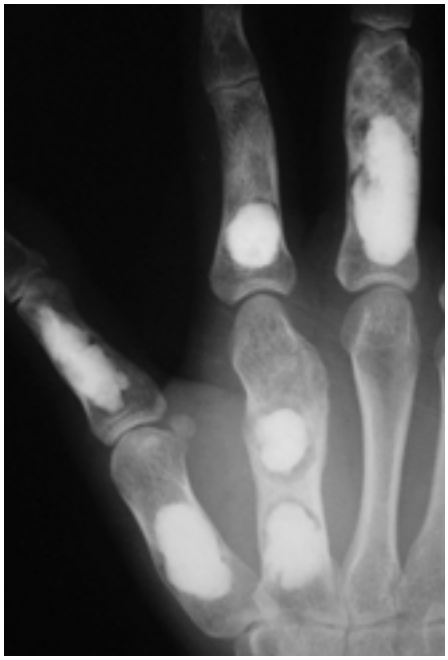

Fig 4. X-ray after curettage of the bone tumors and filling of the cavities with alpha-tricalcium phosphate cement.

metacarpal and proximal phalanx of the index finger, and second metacarpal and proximal phalanx of the middle finger. An upper arm tourniquet was used to produce a bloodless field. The cortex over the tumor was fenestrated into a square shape of approximately 5 x 8 mm. Careful curettage was then performed using a sharp spoon. The gross appearance of the resected tumor showed cartilage-like tissue. Alpha-tricalcium phosphate cement was used to fill the resultant bone cavity and to prevent postoperative pathological fractures (Fig 4). The cortical bone piece excised for the fenestration was used to cover the material. The patient could resume all activities of daily living immediately after the wound healing. Microscopical examination showed that the specimen consisted of cartilage matrix and multiple chondrocytes, the nuclei of which were not enlarged, and mitoses were not seen. These tumors were diagnosed as Ollier's disease. X-rays taken three years after the operation showed new bone ingrowth surrounding the material with little evidence of absorption (Fig 5). None of the finger joints showed further restriction of motion.

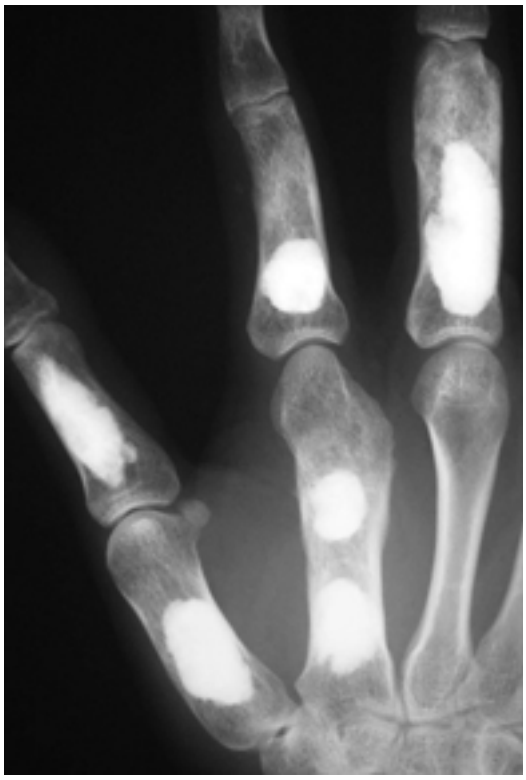

Fig 5. Present X-ray 3 years after the operation. The figure shows that there is a new bone ingrowth surrounding the alpha-tricalcium phosphate and a minimal absorption of this material.

DISCUSSION

Olleir's disease was first described by Ollier in 1899, in a 14 year-old child with multiple enchondromas showing a marked unilateral predominance [13]. Multiple enchondromas tend to occur in infancy or early childhood, while solitary enchondromas are

most often seen in individuals from 10 to 30 years of age. Although solitary enchondromas usually present with the complaint of pain and, occasionally, an associated pathological fracture, Ollier's disease is initially more often noticed because of the deformity and swelling of the involved limb, especially when fingers are affected. In cases affecting the tubular bones of the upper and lower limbs, patients have unilateral angular deformities, leg-length discrepancy, and joint maladaptation. The pathogenesis is still unknown. Recent cytogenetic studies have revealed several chromosomal abnormalities in patients with multiple enchondromatoses, leading to sarcoma transformation [4, 14]. Liu et al. have found that approximately 30% of patients (mean age at diagnosis was 40.5 years) with Ollier's disease will develop a malignant bone neoplasm, most probably chondrosarcoma [11]. Schwartz et al. have reported that the incidence of secondary chondrosarcoma in patients who have Ollier disease is about 25 per cent at the age of forty years [15]. For the hand lesions, malignant transformation is considered to rarely occur [5, 8].

The reported standard treatment is curettage of the tumor and filling of the resultant bone cavity with an autologous bone graft. However, it is often difficult to obtain a sufficient amount of cancellous bone to fill the large cavities after removal of multiple lesions. Contractures during casting is another concern, because several joints are usually involved. In cases with severe deformity, the treatment of choice is amputation. Blauth and Sonnichsen have stated that the defect should be filled with loosely emplaced autologous spongiosa [3]. Yahagi, however, has described that in cases of pediatric patients who have excellent ability of bone formation, simple curettage alone can yield good clinical results [18]. In our case, the patient has undergone simple curettage four times so far. Although preoperative plain X-rays at the age of 15 years showed remarkable deformities in the fingers, preoperative X-rays at the age of 21 years demostrated incredible improvement of the appearance. Judging from this finding, curettage alone can be considered to provide a reasonable cosmetic improvement. Therefore, we recommend simple curettage of multiple lesions at an early stage when Ollier's disease presents.

The present patient had intramedullary lesions that enlarged again 6 years after the last of the previous operations. We used alpha-tricalcium phosphate cement to fill the cavities caused by curettage of the lesions. There has been no description of artificial bone grafting for Ollier's disease published in the English literature. In cases of solitary enchondroma, some authors have recommended simple curettage without bone grafting and have reported good clinical results, especially in younger patients. However, such patients must undergo casting and wait for 4 to 6 weeks before they can resume unrestricted daily activity, and some patients experience joint contracture of the affected limb [7,16,17]. Autologous bone grafting is performed conventionally, but the contribution of cancellous bone to the structural strength in the short term is considered to be very low [9]. Recently, bone substitutes have also been used instead of autologous bone grafting. Although hydroxyapatite, beta-tricalcium phosphate, and various granular materials have excellent biocompatiblity, these can not be considered to provide sufficient structural strength before consolidation [1,6,12]. PMMA cement provides

immediate mechanical stability [2], but the heat of polymerization and monomer toxicity are concerns. Our present case needed initial mechanical stability to prevent restrictions in the activities of daily living. In addition, such a large amount of autologous bone was not easy to obtain. Alpha-tricalcium phosphate cement is an easily injectable material for filling cavities and can be used alone to provide excellent stabilization and allow early mobilization [10]. However, resorption seems to occur quite little. A possible concern is that the remaining material may make it difficult to re-operate, if there is a relapse of the tumor in the future.

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