

SOLITARY ENCHONDROMAS : IS RADIOGRAPHIC FOLLOW-UP SUFFICIENT IN PATIENTS WITH ASYMPTOMATIC LESIONS ?

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Enchondromas are benign cartilaginous tumours and rarely transform into chondrosarcomas. Curettage is usually performed, and a low rate of complications is assumed.

We analysed retrospectively data from 73 patients with enchondromas treated by curettage with respect to symptoms, therapy, complications and recurrences. All patients were treated by curettage of the tumour, in most cases followed by cancellous bone grafting. Twenty-three percent of the patients presented a complication. There were two recurrences but no malignant transformations in the follow-up period. During the same time period 29 patients with chondrosarcomas were treated, including two secondary chondrosarcomas. One had a recurrence of a benign enchondroma of the metacarpal and developed a pulmonary metastasis from a chondrosarcoma. The other was a patient with Ollier's disease who had a secondary chondrosarcoma of the radius.

Malignant transformation of a solitary enchondroma to a chondrosarcoma is rare. On the other hand the complication rate of enchondroma curettage is considerable. Regular radiological follow-up of asymptomatic enchondromas may therefore be the better option.

enchondromas (4, 6). The lesion is solitary in most cases ; multiple lesions are found in only 10% of patients. Multiple, predominantly unilateral enchondromas are found in Ollier's disease. Maffucci's syndrome is characterised by the presence of enchondromas and haemangiomas. Enchondromas usually do not cause pain possibly because of their slow growth, insignificant peritumoral reaction and avascularity. They are usually incidental findings, but occasionally the diagnosis is made after a pathological fracture. According to the classification of Enneking, an enchondroma is a benign stage two tumour in children and a benign stage one tumour in adults (7). Malignant transformation of a solitary enchondroma to a secondary chondrosarcoma is extremely rare. However, the risk of malignant transformation in Ollier's disease may be significantly higher : a 10-20% risk has been reported by Brien *et al* and Liu *et al* (1, 12) and up to 50% by Jaffe (10). The risk of malignant transformation in Maffucci's syndrome is 50-100% including the typical additional carcinomas (13, 15). Despite their low malignant potential asymptomatic solitary enchondromas are frequently treated

INTRODUCTION

Enchondromas are intramedullary hamartomas characterised by the formation of well-differentiated hyaline cartilage (2). The tumour involves the medullary canal of the bone, most often in the small bones of the hands and feet, as well as in the proximal femur and the humerus. Ten to fifteen percent of all benign bone tumours are classified as

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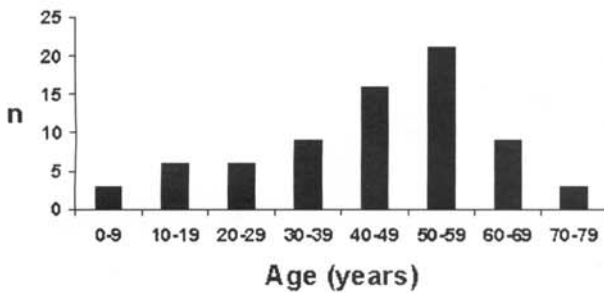


Fig. 1. — Age distribution of the 73 patients with surgically treated enchondromas.

surgically, as the risk of complications of surgical treatment has been perceived as small. The aim of this retrospective study was to determine the indication for treatment, the surgical procedures, the complications and the outcome of enchondroma patients treated surgically. To estimate the risk of malignant transformation, all cases of secondary chondrosarcomas in the same time period have been analysed for comparison.

MATERIAL AND METHODS

From October 1980 to January 1997, 170 patients with benign bone tumours were treated surgically in our department. In 73 of these (43%) an enchondroma was diagnosed and confirmed by histology. The patients were seen on a regular basis and were reviewed with a mean follow-up of 102 months (range: 29 to 228 months). During the same time period, 29 patients with chondrosarcomas were treated. The clinical data were retrieved from medical notes, X-rays and outpatient interviews. The follow-up was conducted by personal contact with the patients or their general practitioners. Recurrence status and postoperative complications, such as fracture, nerve palsy, limited range of motion, swelling or pain were noted. No patients were lost to follow-up.

RESULTS

Of the 73 patients who were treated for an enchondroma, 68 (93%) had a solitary and five (7%) had multiple enchondromas. The average age of the patients was 42 years (range: 3 to 74 years) (fig 1). There were 32 male and 41 female patients; the gender distribution was 1:1.3.

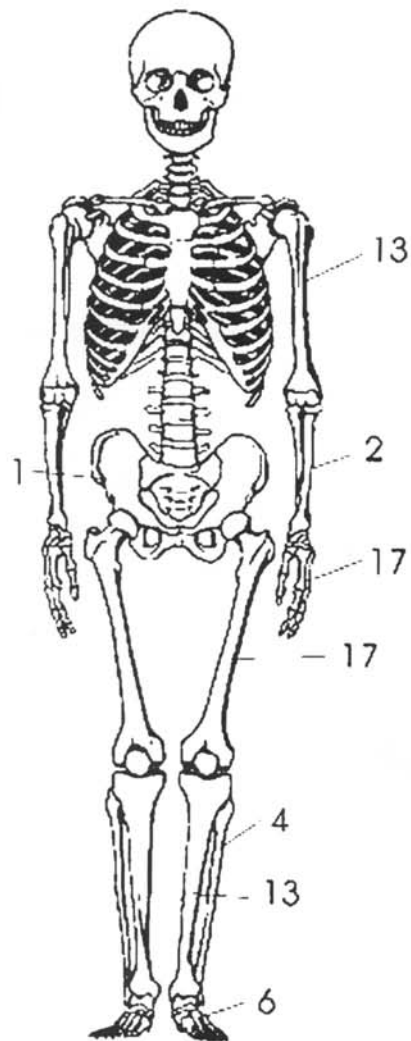


Fig. 2. — Location of the enchondromas in 73 surgically treated patients. In cases of multiple enchondromas the site of surgery is shown.

The majority of patients ($n = 33$) presented with pain, 14 with swelling, four with restriction of movement, four with neurological symptoms and three with a pathological fracture. In 19 patients (26%) the enchondroma was an incidental finding.

Most of the lesions ($n = 40$) were found in the lower limbs, 32 in the upper limbs, one in the acetabulum (fig 2). In all cases radiographs were obtained before surgery (fig 3). In 19 cases magnetic resonance imaging (MRI) and in 10 cases computed tomography (CT) scans were performed. Of



Fig. 3. — AP X-rays of the right hand showing an asymptomatic enchondroma of the second metacarpal in a 41-year-old woman. The enchondroma was an incidental finding.

33 bone scans, only one in a patient with a proven enchondroma showed no uptake. In 15 patients a biopsy was performed before surgery because of suspected malignancy.

In all cases the cortex of the bone was fenestrated and a thorough curettage of the tumour was performed. In 53 patients the bone cavity was filled with autologous cancellous bone graft. In one patient reconstruction of the bone with a fibular graft was necessary; in five cases an additional plate osteosynthesis was performed.

According to the classification of Enneking, all tumours were stage G0. Twenty-six tumours showed a latent, 43 an active and four an aggressive growth.

Seventeen patients (23%) presented a complication. Three patients needed revision surgery because

of postoperative bleeding. In three cases an injury of the femoral nerve (1) or the tibial nerve (2) was observed. Five postoperative fractures at the site of the lesion required a reoperation or external fixation (fig 4). Four patients complained of reduced range of motion. In two patients (2.7%) a recurrence was observed, which required a second operation. Sixty-five patients were free of symptoms after treatment; in six patients negligible and in two patients major problems were caused by nerve injury as a result of surgery. The average follow-up period was 102 months from 29 to 228 months.

During the same time period 29 patients with chondrosarcomas were treated surgically. Two had a secondary chondrosarcoma. The first patient (female, 61 years) had undergone curettage and bone grafting for an enchondroma of the left fifth



Fig. 4. — X-ray of a 48-year-old patient with a pathological fracture of the proximal left humerus after curettage and bone grafting of an enchondroma. Treatment consisted in a second bone grafting and osteosynthesis with a plate.

metacarpal elsewhere. Two and a half years after the initial surgery the patient came to us complaining of pain and swelling of the surgically treated region for one year. X-rays showed recurrence of the lesion; a second curettage was performed. Three months later recurrence of the tumour now with soft tissue infiltration was observed. Because of the risk of malignancy a wide resection was performed. Histologically the tumour was classified as a benign enchondroma with soft tissue extension. Due to the clinical and radiological malignant features the patient was seen regularly with follow-up X-rays of the hand and a CT-scan of the chest. Fifty-two months after the initial surgery a round

pulmonary lesion was detected by routine chest X-ray and confirmed by CT-scan. The lesion was resected and histologically classified as a chondrosarcomatous metastasis.

The second case was a 39-year-old male patient who had a pathologic fracture of the right radius in 1993 (fig 5). Biopsy showed a secondary chondrosarcoma. Bone scan and radiographs showed Ollier's disease with lesions in the right clavicle, the right humerus and the right first metacarpal. Biopsy of the largest lesion (humerus) showed a benign enchondroma. The patient was treated by resection of the distal radius and reconstruction with a vascularised fibula (fig 6). During follow-up



Fig. 5. — AP and lateral X-rays of the right radius of a 39-year-old patient with Ollier's disease showing a grade I chondrosarcoma (histologically confirmed) with pathological fracture.

no local recurrence or progression of the disease was noted.

DISCUSSION

Enchondromas are benign, usually painless cartilaginous tumours. Malignant transformation of solitary enchondromas is estimated to be low, about 2-3% (1). In contrast the risk of local malignant transformation in multiple enchondromas in Ollier's disease or Maffucci's syndrome is estimated to be 10-50% (1, 10, 12, 13, 15). However, the risk of secondary malignancy is difficult to assess because most enchondromas show no symptoms



Fig. 6. — AP X-ray of the right forearm of a 39-year-old patient with Ollier's disease and grade I chondrosarcoma in the right radius after resection of the distal radius and reconstruction with a vascularised fibula.

and are therefore never detected. Hence the risk of malignant transformation is likely to be overestimated. There have been no prospective observational studies with a larger group of patients. Secondary chondrosarcomas have been described in cases with recurrence of an enchondroma that was initially diagnosed histologically as a solitary enchondroma. In these patients it is thought that the enchondroma transformed into a secondary chondrosarcoma. However, in some patients the

primary histology may already show a chondrosarcoma in a thorough second review (5, 8, 14).

Surgical treatment of solitary enchondromas is often advised because of the diagnostic difficulties and to prevent malignant transformation. Most often this will be curettage and filling of the defect with autologous bone graft (3, 11). The assumption has been made that the risk of complications is low. This is in contrast to our data. Twenty-three percent of our patients presented complications. Eleven percent are permanently impaired secondary to the surgical treatment. Therefore the indication for surgery should be discussed more thoroughly than is commonly thought.

During the same time period two patients presented with secondary chondrosarcomas. One patient had Ollier's disease and therefore had a known higher risk of malignant transformation. Secondary chondrosarcoma is the most common skeletal tumor in patients with Ollier's disease or Maffucci's syndrome (15). These tumours tend to be histologically low-grade chondrosarcomas; therefore wide resection is successful in almost all of these patients (9). In keeping with this our patient with Ollier's disease had no recurrences or metastases during follow-up of nine years.

The other patient with a secondary chondrosarcoma had a histologically proven pulmonary chondrosarcoma metastasis. The course of the disease, clinical and radiological features raised the suspicion of malignancy in contrast to the initially benign histology. This demonstrates the problem of histological classification between enchondromas and chondrosarcomas. Even if both cases were considered as malignant transformations, the risk of malignant transformation would be less than 3%. The calculated risk for solitary enchondromas would be even less (1 of 68, 1.5%), in Ollier's disease 20% (1 of 5).

Thus one should contemplate a less aggressive non-surgical approach in patients with solitary enchondromas. In solitary enchondromas surgery should be restricted to patients with existing or impending pathological fractures. In all other patients with solitary enchondroma, observation seems to be adequate. If the typical radiological and clinical features are present, a yearly follow-up

is recommended. A high Tc uptake on bone scans alone seems not to be an indication for surgical treatment, because in our series high Tc uptake does not correlate with the findings of a grade I chondrosarcoma or a higher rate of local recurrence. However, in cases with growing lesions or pain, biopsy or a wide resection should be performed. Such a policy would result in a significant reduction of unnecessary operations and their complications.

In cases of recurrences of surgically treated enchondromas the risk of malignant transformation is raised (11). In these cases resection is recommended.

A similar approach should be used in patients with multiple enchondromas. It is not possible to reduce the risk of malignant transformation by resection of one lesion only, because it is not known which of the multiple enchondromas will (if ever) transform into a secondary chondrosarcoma. Therefore a yearly follow-up of the lesions with radiographs seems to be sufficient. Surgical intervention should be restricted to patients with clinical and radiological suspicion of malignant transformation.

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