CHONDROSARCOMA DEVELOPING IN SYNOVIAL CHONDROMATOSIS

A CASE REPORT

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Synovial chondromatosis is a rare condition in which osteocartilaginous nodules are formed by synovial metaplasia and become intra-articular loose bodies. It is usually monarticular, most commonly affecting the knee and is regarded as invariably benign. There are reports of malignant change, but only the two case studies of Mullins, Berard and Eisenberg (1965) and of Dunn et al. (1974) fully document the development of chondrosarcoma in this condition.

We report a patient with a long history of histologically proven synovial chondromatosis in whom the condition became clinically aggressive and underwent malignant transformation to chondrosarcoma.

CASE REPORT

In April 1959, a 25-year-old man presented with a swollen stiff left knee after minor trauma. Radiographs showed loose bodies in the joint, and at operation the synovium was found to be thickened, inflamed and dotted with multiple cartilaginous nodules. Numerous loose bodies were removed but some were inaccessible in the posterior compartment. Some swelling in the popliteal space persisted after operation, but the joint became painless with a range of movement of over 90°. In December 1962, after a flexion injury, a painful mass was palpable in the posterior joint space, but the pain settled with physiotherapy.

In April 1968, an increase in swelling in the popliteal fossa followed an episode of minor trauma. The joint was held 50° flexed and could move through only 10°. Radiographs showed chondromatosis extending through a large part of the joint both anteriorly and posteriorly. An anterior synovectomy was carried out, and although only slight improvement in the range of movement was achieved the patient was able to resume his employment as a lorry driver. In 1975, he sustained fractures of the left femur and tibia in a tractor accident. These healed unevenly and the knee remained quiescent during treatment of the leg despite an increase in the radiographic size of the calcified masses.

The patient was lost to review for three years from 1978. Pain recurred in September 1981 and at this stage the skin over greatly enlarged posterior swellings showed pressure changes and a small calcified mass had developed on the femoral shaft at mid-thigh level. The knee was again explored, abundant calcified material was removed, but some was left adherent to the upper tibia. There was considerable symptomatic improvement. In January 1982, the calcified mass in the thigh was biopsied, showing heavily calcified cartilaginous tissue in keeping with synovial chondromatosis.

In September 1984, there was rapid clinical deterioration with pain, swelling and local heat. A soft-tissue tumour had developed over the lower femur, eroding both femoral condyles. Chest radiography, an ultrasound scan of the abdomen and a CT scan were all normal. In April 1985 the lobulated cartilaginous mass was excised and debris was cleared from the joint. Histology showed transformation to a chondrosarcoma and radiologically there was a tumour in the medullary cavity of the femur (Figs 1 and 2). In view of this the leg was disarticulated through the hip joint in May 1985. Postoperatively the patient was mobilised successfully with a prosthesis. One year later, there has been no evidence of metastases.

Pathology. The initial biopsy in 1959 showed hyperplastic synovium containing round osteocartilaginous nodules typical of synovial chondromatosis (Fig. 3). Further specimens taken in 1968 showed similar histolo-
Radiographs taken in April 1985 showing a large extra-articular soft tissue mass in front of the knee, calcified masses along the shaft of the femur and invasion of the medullary cavity distally.

gy, but the loose bodies were larger and more heavily calcified with peripheral bone formation (Fig. 4). Sections of the 280 g of heavily calcified cartilaginous material excised from the joint in 1981 showed some increase in cellular activity and nuclear pleomorphism. Endochondral bone formation with heavy calcification was still present, and the appearance was within the spectrum of that seen in synovial chondromatosis (Fig. 5). The nodule excised from the mid-thigh in 1982 was almost entirely calcified, with only a few residual foci of chondrocytes and was considered to be due to tenosynovial involvement. There was no evidence of malignant change.

The mass excised in 1985 weighed 550 g and had superficially eroded both femoral condyles. This tumour showed a myxoid stroma, with a striking increase in cellularity and nuclear pleomorphism, especially around the margins of the lobules. Calcification was sparse and bone formation was lacking. Malignant transformation to a chondrosarcoma had occurred (Fig. 6).

Histology in 1959, showing hyperplastic synovium with two osteocartilaginous nodules (H & E × 34).

Appearance in 1968 showing calcification of an osteocartilaginous nodule covered by synovium (H & E × 34).
Histology in 1982, showing heavily calcified cartilaginous tissue and some increase in cellularity and pleomorphism (H & E × 34).

The specimen from disarticulation of the leg at the hip, performed one month later, showed extensive extra-articular soft tissue with partially calcified masses around the knee joint, filling the popliteal space, and superficially eroding the cortex of the femur and tibia. The medullary cavity of the femur between the condyles was replaced by a cartilaginous tumour with a maximum diameter of 5 cm (Fig. 7). This was a poorly differentiated chondrosarcoma which infiltrated between bone trabeculae and was richly cellular with marked atypical nuclei (Fig. 8). The extra-articular masses showed variable cellularity with calcification which was often heavy, while developing osteocartilaginous nodules were evident in the synovium around the cruciate ligaments. The nodules attached to the femoral shaft showed unchanged histology, interpreted as tenosynovial chondromatosis with no malignant change.

Biopsy appearance in 1985 showing extra-articular chondrosarcoma with marked peripheral cellularity and atypical nuclei (H & E × 56).

**DISCUSSION**

Synovial chondromatosis is an uncommon benign condition characterised by multiple osteocartilaginous nodules in the synovium which detach to form loose bodies. It is usually monarticular, affecting the knee most commonly and sometimes the elbow and the hip. Some series have been reported to show a tendency for local recurrence after synovectomy (Mussey and Henderson 1949; Murphy, Dahlin and Sullivan 1962; Milgram 1977; Villacin, Brigham and Bullough 1979), but despite atypical cellularity they did not describe malignancy.

Care must be exercised in interpreting the histological findings. Cellular activity and atypical appearances have been recognised as consistent with the active growth of metaplastic cartilage (Murphy, Dahlin and Sullivan 1962; Villacin, Brigham and Bullough 1979).

Amputation specimen 1985. The photograph of a hemi-section of the distal femur and upper tibia shows chondrosarcoma involving the medullary cavity and a calcified nodule attached to the femoral shaft. The histological section (Fig. 8) shows poorly differentiated chondrosarcoma infiltrating between trabeculae of bone (H & E × 34).
These features may be difficult to distinguish from chondrosarcoma, and the knowledge that the biopsy has been taken from an extra-osseous site may be necessary to avoid the erroneous diagnosis of malignancy.

The two previously reported cases of chondrosarcoma included satisfactory documentation of antecedent synovial chondromatosis for 13 and 22 years. In our case there was an interval of 26 years between the initial synovial biopsy showing chondromatosis and the transformation to chondrosarcoma. Clinical and radiological examination of the joint are not decisive in the diagnosis of malignant change, since obliteration of the joint space and extra-articular soft tissue masses may occasionally occur late in the course and are not on their own indicative of malignancy (Goldman and Lichtenstein 1964; Sim, Dahlin and Ivins 1977).

However, a deteriorating clinical picture with rapidly enlarging extra-articular masses should arouse suspicion and confirm the need for biopsy. In our case, significant alteration in pathology was seen in the extra-articular masses, and radiographs showed the medullary cavity of the femur to be involved. As in the two other reported cases there had been a long clinical course, but the series reported by Murphy et al. (1962) included follow-up to 32 years without malignant change. The sequential histological findings in our case included an increase in cellular atypia after 22 years, but this, at an extra-osseous site, was considered insufficient evidence for the diagnosis of malignancy. Four years later, the resected tumour mass gave unequivocal evidence of malignancy, which was substantiated by the involvement of the medullary cavity.

Each exacerbation of symptoms in our case was preceded by minor trauma except for the rapid deterioration in the last year. Murphy et al. (1962) found no convincing correlation between trauma and synovial chondromatosis.

The earlier treatment of our patient had never restored a full range of movement but he had been pain-free and able to continue his strenuous work. At this stage complete excision of the affected tissues would probably have required either an arthrodesis or an arthroplasty, and it was felt that radical surgery was not indicated. Following malignant transformation and the involvement of the medullary cavity of the femur, a hip disarticulation was performed in this as in the other two previously reported cases. Our patient shows no signs of distant metastases 12 months after amputation but Dunn et al. (1974) found evidence of metastasis in their patient after 2 years 6 months.

This case of chondrosarcoma secondary to synovial chondromatosis demonstrates the progressive nature of the condition and the difficulty of management as well as the propensity for recurrence when total excision of affected tissues is impossible. Sudden clinical deterioration in long-standing cases should alert the clinician to the possibility of malignant transformation.

REFERENCES