EFORT13-1646

Synovial chondrosarcoma arising in primary synovial chondromatosis: A tertiary unit experience

S. Evans [1,*], M. Boffano [1], S. Chaudhry [1], L. Jeys [1], R. Grimer [1]
[1]Orthopaedics, NHS, Birmingham, United Kingdom

INTRODUCTION: Malignant transformation of primary synovial chondromatosis to synovial chondrosarcoma is recognized to be a rare event.
OBJECTIVES: Identify patients treated at our centre with primary synovial chondromatosis that subsequently developed into a synovial chondrosarcoma
METHODS: Retrospective review of a prospectively collected database. Patient demographics were recorded along with the site of primary synovial chondromatosis. From this details of those patients who subsequently developed synovial chondrosarcoma were collected.
RESULTS: 78 patients with primary synovial chondromatosis identified. All had been diagnosed following analysis of their radiological and histological findings between 1980-2011. 33 females (42.3%), 45 males (57.7%). Mean age at presentation with primary synovial chondromatosis 28 years. The site of primary synovial chondromatosis was; knee in 30 patients, hip in 22, hand in 7, shoulder in 6, elbow in 5, foot in 5, wrist in 2, and the ankle in 1. Of the 78 patients, 5 (6.4%) went on to develop synovial chondrosarcoma; 4 around the hip, 1 around the knee. Median time from original diagnosis of synovial chondromatosis to malignant transformation was 240 months (range 32-468). The single knee patient was managed with an above knee amputation, but developed lung metastases 32 months post-operatively and died 18 months later. Of the 4 hip patients, 2 were managed with a hind-quarter amputation. At last follow-up (one 12 years, the other 12 months) post-surgery, both were doing well with no local recurrence or lung metastases. Unfortunately, the 2 other patients who developed synovial chondrosarcoma around the hip had large, aggressive tumours. Both had debulking prior to developing lung metastases within 18 months of these palliative procedures.
As with other chondrosarcomas, radio/ chemotherapy are not typically effective treatments and, therefore, our cases did not receive adjuvant therapy. In summary, 78 patients were identified with primary synovial chondromatosis, with 5 developing synovial chondrosarcoma. This represents transformation to malignancy in 6.4% of cases. Furthermore, it is apparent from performing Kaplan-Meier survival analysis that the risk of malignant transformation increases as time elapses.

CONCLUSION: Multiple cases have been reported of chondrosarcoma arising in primary synovial chondromatosis but to our knowledge the case series we present is the largest single centre review in the literature. Our series has shown the prevalence of chondrosarcoma in primary synovial chondromatosis to be 6.4% thereby questioning the dictum that malignant transformation in primary synovial chondromatosis is rare.
The local recurrence of primary synovial chondromatosis has been reported to be as high as 23% despite adequate surgical debridement. The difficulty lies with identifying those recurrences that are benign. At our centre we view multiple recurrences with the development of marrow invasion as highly suspicious of malignant transformation and we would recommend that any rapid deterioration in the patient's clinical course should be regarded as suspicious and treated at, or referred appropriately to, a tertiary hospital familiar with managing these often complex cases.

Disclosure of Interest: None Declared

Keywords: synovial chondromatosis, synovial chondrosarcoma