Synovial Sarcoma of the Extremities. A Diagnosis that is Easily Missed

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ABSTRACT
Synovial sarcoma of the extremities is an uncommon type of soft tissue sarcoma occurring predominantly in young adults at the para-articular regions. We present a series of 10 patients with an average age of 44 years and include a follow-up of 39 months. Eight patients had a surgical procedure for a mistaken benign lesion. In contrast to other soft tissue sarcomas, the swellings were associated with pain and most were fixed to the underlying structures. Five patients had a local recurrence after many years, stressing the necessity for close and long term follow-up in these patients.

INTRODUCTION
Synovial sarcoma is an uncommon neoplasm that can occur in many parts of the body, but mainly in the extremities. It occurs more often in the younger adult age group and in para-articular regions. As such, it can be easily misdiagnosed and can thus be treated inappropriately (i.e. where an excision is performed with inadequate margins). We reviewed 10 patients with synovial sarcoma of the extremities and sought to elucidate the peculiarities of this condition so as to better understand the condition and to improve treatment.

MATERIALS AND METHODS
Between 2001 and 2006, 10 patients with a histologically proven diagnosis of synovial sarcoma involving either the upper or lower limbs were treated at our hospital. A detailed history was obtained from each patient, with documentation of symptoms and events that took place. This was supplemented by the patients’ referral letters and information from the patients’ outpatient cards where visits to government clinics were documented. On admission, a plain radiograph, an MRI of the lesion, a chest radiograph, a CT thorax and a bone scan were taken. After confirmation of the diagnosis with a biopsy, definitive surgery was carried out, followed by radiotherapy and chemotherapy in selected patients. Patients were subsequently followed up at regular intervals.

RESULTS
There were 6 male and 4 female patients with a mean age of 44 years (range 10-72 years). Nine tumours occurred in the lower limb (2 at the foot, 2 at the ankle, 2 at the knee joint, 2 in the thigh and one at the groin); one lesion involved the upper limb (hand). Tumours arising near large joints did not actually involve the joints but were in the para-articular regions. The majority of the patients (7) gave a long history of the presence of a swelling, ranging from 3 to 12 years. Before being seen at our unit, 9 patients were initially diagnosed with a benign lesion, and surgery had been performed (seven excisions and one desloughing) on 8 patients. All excised tumours recurred; 5 recurred more than two years post excision. A second excision was performed in 3 of these patients with subsequent recurrence before referral to us. (Table I)

The average size of the tumour swelling was 8.5 cm [range 3cm – 9cm]. (Figs. 1 and 2). Nine tumours were fixed to the underlying structures; 8 were associated with pain. There was regional lymph node spread in two patients. No lung or bone metastases were detected. Six patients were treated with wide excision. Four patients required amputations (forearm, above knee, below knee and Syme’s), which were performed when it was felt that better function would be achieved than with limb with amputation salvage surgery. Based upon the oncologists’ recommendation, 4 patients received adjuvant chemotherapy and 6 patients underwent radiotherapy. At the last follow-up, 4 patients were disease-free, 3 had died and one had lung metastases.

DISCUSSION
Synovial sarcoma is an aggressive tumour that affects patients in their prime. Because it occurs in the young and around the joint, it is often mistaken for a benign lesion such as a ganglion or bursitis. Nine of our patients were previously misdiagnosed. Eight patients had undergone surgical procedures before referral. We had to restage each patient before proceeding to do a wide excision or amputation. Patients were first seen at our clinic an average of 5.3 years from the onset of the symptoms. Some had had