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 [1], 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 [5]. 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

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Table I: Patient Data

No.	Age/Sex	Onset of symptoms to definitive surgery (duration)	Initial Diagnosis	Previous Surgical Procedure	Swelling Duratiion (months)	Pain	Site	Size (Widest Diam) (cm)	Mobility	Lymph Nodes	Surgery	F/U	Status
1	34/M	3 months	Giant cell tumour	-	3	Yes	Lat malleolus	10	fixed	Nil	BKA	55	Well
2	57/M	10 years	Lipoma	Excision 10 yrs	15	Yes	Lat thigh	19	Fixed	Nil	Wide excision	21	Lung Mets
3	10/M	10 months	Abscess	Excision 9 months	9	No	Groin	5	fixed	Inguinal	Wide excision	29	Died
4	67/F	11 years	Bursitis	Excision 4 years Re-excision 3 years	3	Yes	Medial malleolus	3	fixed	Nil	Wide Excision	44	Well
5	29/M	6 years	Chondroma	Excision 6 years	36	No	Dorsum foot	5	fixed	Inguinal	Wide Excision BKA	93	Died
6	15/M	4 years	Popliteal Cyst/ Pigmented Villonodular synovitis	Desloughing 9 months	17	Yes	Popliteal fosssa	4	Fixed	Nil	AKA	42	Well
7	72/F	12 years	Ganglion Lipoma	Aspiration Local Excision 5 years Re-excision 3 ears	12	Yes	Dorsum Hand/wrist	7	Fixed	Nil	Forearm amputation	16	Died
8	67/M	6 months	Pseudo-Aneurysm	-	4	Yes	Dorsum Foot	10	fixed	Nil	Syme amputation	10	Lost to F/U
9	58/F	5 years	Soft tissue sarcoma	Excision	12	Yes	Thigh	14	mobile	Nil	Wide excision	5	Lost to F/U
10	35/M	3 years	Calcified bursa	Excision/ curettage 3 years Re-excision 2years	12	Yes	knee	8	fixed	nil	Wide Excision	83	Well

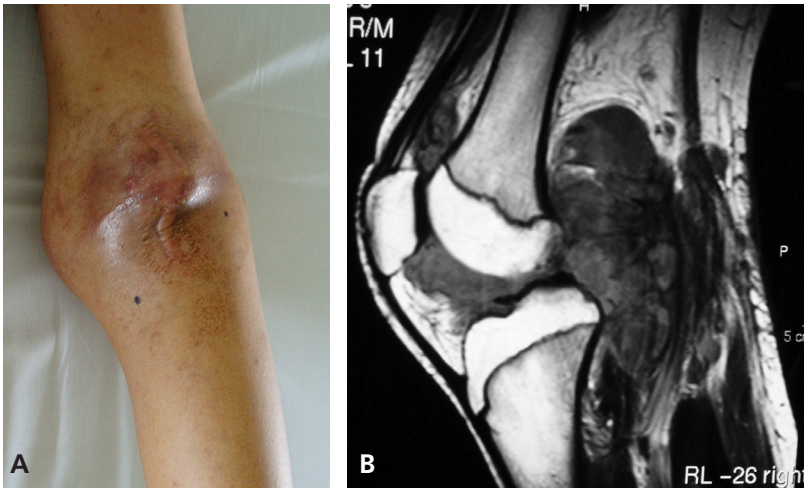

Fig. 1: 15-year-old male with popliteal swelling diagnosed initially as villonodular pigmented synovitis. 1A: Photograph of the swelling and scar from the initial surgery. 1B: MRI of the swelling.

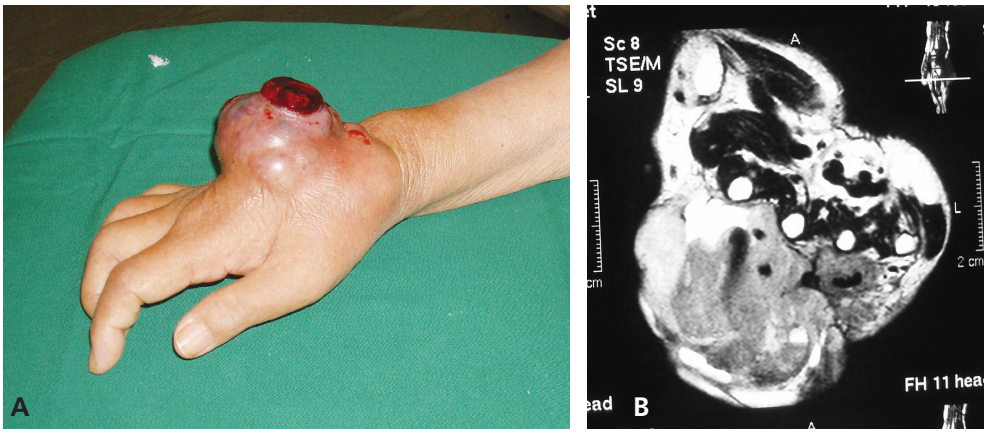

Fig. 2: 72-year-old female with a recurrent swelling over the dorsum of the hand after two previous excisions. 2A: Photograph of the recurrent swelling. 2B: MRI of the swelling.

multiple visits to general practitioners where malignancy had not been suspected.

Two features were noted in these patients that were different from patients with other types of soft tissue sarcomas. Most patients with soft tissue sarcomas do not have pain until the later stages. Eight of our patients had associated pain, while 9 patients had fixed swelling. Most of the tumours were mobile clinically even quite large. The fixity of the swelling was more likely due to the fact that there was little intervening muscle tissue between the swelling and the bone. In accordance with their aggressive nature, the tumours quickly invaded the underlying tendon or bone, causing pain and decreased mobility on physical examination.

Another anomaly in this series was the long history of swelling (up to 12 years) and the absence of traceable metastases when they presented at our unit. Perhaps the prior excisions, albeit with non-oncological margins, played a role in this clinical finding. Another possibility is that the tight soft tissue compartments around the joints made it less likely for the tumour cells to detach and find their way into the circulation. However, it does not mean that synovial sarcomas are innocuous, slow-growing malignancies. Once they reach a certain size/stage, they will manifest their aggressive potential, as shown by the recurrences and survival rates [6,7,8,9].

CONCLUSION

Synovial sarcoma often affects younger adults and tends to occur around joints. Most patients have associated pain and the swelling has a propensity to be fixed on examination. Misdiagnosis is common and often contributed towards delay in final diagnosis and definitive treatment. The high rate of local recurrence may be related to inadequate margin of excision in these patients.

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