A LONG-TERM FOLLOW-UP OF SYNOVIAL SARCOMA

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Synovial sarcoma is the name given to malignant tumours that arise from pre-existing synovium, or to mesenchymal tumours that arise elsewhere but show a synovial pattern of differentiation.

Although descriptions of malignant tumours arising from joints, tendons and bursae had long been known (Hardie 1894, Marsh 1898, Lejars and Rubens-Duval 1910), it was not until 1927 that certain common features were recognised and the disease emerged as a clinical and pathological entity (Smith 1927). At that time the name synovioma was applied to the tumour. However, the term synovial sarcoma is now usually employed because synovioma is an ambiguous term that may erroneously be applied to a benign synovial tumour.

As synovial sarcoma is relatively rare a surgeon may see only one case in a lifetime. This review is based on the combined experience of the Toronto teaching hospitals over the last twenty years; its object is to lay down guidelines in prognosis and treatment. Thirty-nine cases have been collected and analysed. If any doubt existed about the diagnosis the case was excluded from the series.

CLINICAL PICTURE

There is little in the early manifestation of synovial sarcoma to indicate its lethal potential. The patients are usually young adults (Fig. 1), but no age group is exempt; the youngest patient in this series was aged ten weeks and the oldest ninety-two years. Males were slightly more susceptible than females in the ratio of three to two.

Half the patients presented with a painless mass near a joint, one-quarter had a painful mass, and one-quarter complained only of pain. Four out of ten patients had noticed a painless mass for an average of four years before diagnosis; the others had had symptoms for less than
three months. A history of recent injury was obtained in about a quarter of the cases, as is common in bone and joint tumours.

The principal sites of involvement are the extremities (Table I), usually adjacent to a joint, although it is most unusual for the tumour to arise from the synovial lining of a joint. It commonly occurs in intermuscular spaces and has also been reported in the chest wall (Eisenberg and Horn 1950), in the hypopharynx (Martens 1955) and in the retroperitoneal tissue (Ariel and Pack 1963).

**TABLE I**

**Principal Sites of Thirty-nine Synovial Sarcomata**

<table>
<thead>
<tr>
<th>Site</th>
<th>Percentage</th>
</tr>
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<tbody>
<tr>
<td>Knee</td>
<td>25</td>
</tr>
<tr>
<td>Thigh</td>
<td>17</td>
</tr>
<tr>
<td>Foot</td>
<td>17</td>
</tr>
<tr>
<td>Hand</td>
<td>15</td>
</tr>
</tbody>
</table>

Other sites include elbow, forearm and ankle.

The size of the tumour may vary from 2 to 20 centimetres in diameter. It may be firm but feel cystic; hence the occasional tragic case in which a “ganglion” has been removed and not examined histologically. The overlying skin may be normal, or it may be red and inflamed. One patient in this series developed multiple cutaneous metastases in the forearm.

Radiographs may be of assistance in making the diagnosis. A discrete mass, usually homogeneous in density and oval or lobulated in form, may be seen. Flecks of calcification are present in about 40 per cent of the cases. Rarely, there is local bone destruction or invasion. As a rule, patients present for treatment before obvious metastases are present. Metastases are most frequently found in the lungs, and regional lymph nodes are not uncommonly involved, but no tissue is exempt and in this series metastases have occurred in bone, brain, skin and viscera.

**Pathological Features**

There is no diagnostic feature in the gross appearance of synovial sarcoma. The tumour may be firm and fibrous or soft and mucoid. It is often white, but the colour may be modified by necrosis and haemorrhage. Cyst formation and spots of calcification are not uncommon (Fig. 2). Deposits of calcium salts are not specific to synovial sarcomata; they occur also in fibrosarcomata. A false appearance of encapsulation is often seen.

**Histology**—The classical diagnostic feature is the presence of two neoplastic cellular elements, the fibrosarcomatous spindle cell and the pseudo-epithelial cell (Fig. 3). The diagnosis cannot be sustained in the absence of one of these characteristic features. In addition, tissue spaces or clefts within the tumour suggestive of a synovial cavity are usually found (Fig. 4). The clefts are lined by cuboidal or columnar-shaped cells with the long axis radial to the lumen of the cleft. If any material is present in the cleft it is acellular and basophilic. Vascular spaces, which occur frequently in all types of sarcoma, must not be confused with synovial clefts. Special stains such as mucicarmine are of little value in diagnosis.

**Survival Rate and Recurrence**

In this series of thirty-nine cases the five-year survival rate was 45 per cent. This compares favourably with previous series (Table II). However, as metastases can appear more than ten years after apparent local cure, the five-year survival rate does not really reflect the influence
FIG. 2
Undecalcified specimen. Flecks of calcification within the tumour mass are seen on the right. Two vascular clefts are seen on the left of the field. (Haematoxylin and eosin, ×200.)

FIG. 3
This shows the pseudo-epithelial cells lining a synovial cleft. The supporting cells form the fibrosarcomatous element of the tumour. (Haematoxylin and eosin, ×200.)

FIG. 4
A synovial cleft is present on the left side of the field. The lumen of the cleft contains acellular basophilic material. A vascular space is seen to the right of the field. (Haematoxylin and eosin, ×200.)
of treatment (Table III). The longest survival in this series was for nineteen years. Survival was not related to the length of time the tumour had been present before treatment, nor to the histological appearances.

Survival related to method of treatment—Three methods of treatment are in current use: primary amputation, local excision and radiotherapy. The results of these are summarised in Table IV.

**TABLE II**
**REPORTED RATES OF FIVE-YEAR SURVIVAL IN SYNOVIAL SARCOMA**

<table>
<thead>
<tr>
<th>Author</th>
<th>Number of cases</th>
<th>Five-year survival (per cent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haagensen and Stout (1944)</td>
<td>104</td>
<td>3</td>
</tr>
<tr>
<td>Pack and Ariel (1950)</td>
<td>.</td>
<td>60</td>
</tr>
<tr>
<td>Wright (1952)</td>
<td>.</td>
<td>47</td>
</tr>
<tr>
<td>Mackenzie (1966)</td>
<td>.</td>
<td>49</td>
</tr>
<tr>
<td>Toronto series (1974)</td>
<td>.</td>
<td>39</td>
</tr>
</tbody>
</table>

**TABLE III**
**SURVIVAL RATES IN THIRTY-NINE CASES OF SYNOVIAL SARCOMA**

<table>
<thead>
<tr>
<th>5 years</th>
<th>10 years</th>
<th>Over 10 years</th>
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</thead>
<tbody>
<tr>
<td>13 out of 29 (45 per cent)</td>
<td>7 out of 24 (30 per cent)</td>
<td>3 out of 24 (12 per cent)</td>
</tr>
</tbody>
</table>

Two patients died of metastases after ten years. Longest survival nineteen years.

**TABLE IV**
**RESULTS OF VARIOUS TREATMENTS**

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Number</th>
<th>Local recurrence</th>
<th>5-year survival</th>
<th>10-year survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary amputation</td>
<td>11</td>
<td>-</td>
<td>4 out of 7 (57 per cent)</td>
<td>3 out of 6 (50 per cent)</td>
</tr>
<tr>
<td>Local excision</td>
<td>20</td>
<td>13</td>
<td>7 out of 18 (38 per cent)</td>
<td>3 out of 14 (21 per cent)</td>
</tr>
<tr>
<td>Secondary amputation</td>
<td>7</td>
<td>1</td>
<td>3 out of 7 (43 per cent)</td>
<td>Nil</td>
</tr>
<tr>
<td>Radiotherapy</td>
<td>5</td>
<td>No clearance</td>
<td>Nil</td>
<td>Nil</td>
</tr>
</tbody>
</table>

The incidence of recurrence after local excision was 65 per cent. Contrary to Mackenzie’s (1966) statement that recurrence is not necessarily a disaster, of the nine patients in this series who had more than one recurrence, two who are children are alive and well after five years, but only one adult (subjected to secondary amputation) has survived for more than four years.

From the figures in Table IV it is clear that amputation must be the treatment of choice. If there are special factors such as the rare tumour not sited on a limb, or a minute tumour...
in a very old patient, or a patient who refuses amputation, then local excision followed by radiotherapy is the best alternative. It is difficult to be dogmatic about radiotherapy, which is never curative, because most patients treated by local excision were exposed to it and no definite advantage could be shown in terms of lessened local recurrence. Of the five patients treated by radiotherapy alone, all died in less than five years. Nevertheless, radiotherapy can cause local regression, and it may check the growth of the tumour for months, or occasionally for a few years.

The numbers treated by other methods are too small to allow any definite conclusions to be drawn. One patient had intra-arterial infusion with cytotoxic drugs without benefit. The exposure to the cytotoxic agents was, however, inadequate: it is known that continuous intra-arterial infusion for solid tumours of the head and neck should be carried out for several months if there is to be any hope of cure. Immunological control was attempted in one hopeless case, but this was a clear example of "too little, too late".

It is obvious that the results of treatment have not improved over the last ten years. The limits of operation and of radiotherapy appear to have been reached. However, the long survival in many cases emphasises the need for further immunological studies. It is to be hoped that through work of this nature advances in the treatment of synovial sarcomata may be made.

SUMMARY

1. Thirty-nine patients with synovial sarcoma have been reviewed.
2. The average rate of five-year survival was 45 per cent; of ten-year survival, 30 per cent; and of survival for more than ten years, 10 per cent.
3. The only important factor influencing the long-term results was the method of treatment; primary amputation was by far the best.

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REFERENCES