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Extra-abdominal desmoid tumors

A follow-up study of 53 patients with extra-abdominal desmoid tumor is presented.
Fine-needle aspiration biopsy seemed to be valuable in the differentiation between a desmoid tumor and a truly malignant soft tissue tumor. The angiographic pictures did not differ from those of other soft tissue tumors, but valuable topographic information was obtained from the angiography in the preoperative planning. The overall local recurrence rate was 17/44, and 15 of the recurrences occurred within the first 2 years. Complete removal of the affected muscle or muscle group is recommended.
Local control can be achieved by non-ablative surgery even after multiple recurrences. Only one out of 53 patients died of the tumor disease.
Radiotherapy can be attempted in extensive tumors, and amputation should be reserved for tumors which have failed to respond to this treatment.

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Desmoid tumors are fibrous, infiltrating, and non-metastasizing tumors, usually involving skeletal muscles. Desmoids appearing in the abdominal wall have been recognized as a separate entity. Extra-abdominal desmoid tumors arising from musculo-aponeurotic structures in the extremities and the limb-girdles present a difficult diagnostic and therapeutic problem. The microscopic picture is uniform from one tumor to another, but the clinical behaviour can be very different.

The aim of this paper is to describe the clinical behavior of extra-abdominal desmoids and to study the role of biopsy and aspiration cytology and angiography in the preoperative work-up. Based on a series of 53 patients, some principles for surgical treatment are defined and different surgical procedures are related to the final outcome of the tumor disease.

Patients and methods

From 1966 to 1978, 20 patients were treated for an extra-abdominal desmoid tumor at the Department of Orthopaedic Surgery II, Sahlgren Hospital, Göteborg. In addition, the material includes 33 patients whose tumor had been sent for diagnostic consultation to the Department of Pathology II, Sahlgren Hospital, Göteborg.

Two patients were lost to clinical follow-up. Six patients died from intercurrent diseases.

Fourty-seven patients were followed up for at least 5 years and the mean follow-up time was 11 (5–34) years.

Histological specimens were available form all 53 patients. Patients with a shorter follow-up time than 8 years since the last operation were examined with respect to recurrence. Patients with a follow-up time longer than 8 years were only interviewed by telephone. All available histologic sections were reviewed.

The chi-square test was used for testing rate differences between groups in a contingency table.

Diagnostic procedures

Sixteen patients were operated on without an adequate preoperative work-up and all these underwent an *inadequate* operation.

In 14 patients, an *adequate* or *probably adequate* operation, was carried out without preoperative, cytological or histological diagnosis.

Plain radiography and *angiography* were carried out preoperatively in 11 patients (Table 1). In two patients, the tumor was detectable already on plain radiograms and in one patient bony destruction was found.

Angiograms with only one projection were carried out in two patients. In nine patients, the angiograms

Table 1. Angiographic appearance of 11 extra-abdominal desmoid tumors

Case	Vascularity in relation to the surrounding tissue ("macrovascularity")	Tumor vessels	Signs of rapid circulation		Opacification ("microvascularity")	Wide veins draining the tumor	Dislocation of surrounding vessels
			Early opacification	Early single vein			
1	More	Normal stretched	+	+	Positive, central radiolucency	+	–
2	More	Normal	+	+	Positive	+	–
3	More	Normal stretched	+	Doubtful	Positive	+	–
4	More	Normal	–	–	Slightly positive	–	+
5	Equal	None	–	–	Equal	–	+
6	Less	None	–	–	Equal	–	+
7	More	Normal stretched	–	–	Positive in the periphery	–	+
8	Equal	Normal	–	–	Slightly positive	–	–
9	More	Normal	+	+	Positive	+	–
10	High	Pathological in a small area	+	+	Positive	+	+
11	High	Pathological	+	+	Positive	+	+

were taken in at least two different projections and the film series covered even the venous phase. The angiographic features are described in Table 1, using principally the same method of evaluation as Kindblom (1975) used for liposarcomas.

The anatomical location of the tumor (usually the muscle in which the tumor had developed) could be determined from the angiograms in seven patients (Figures 1 and 2).

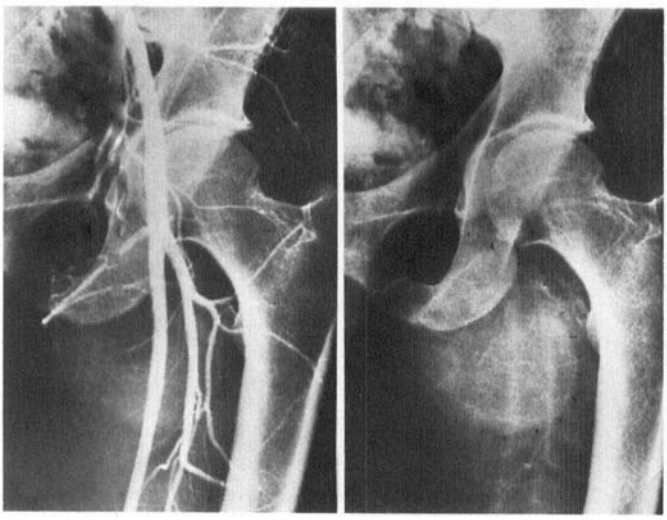


Figure 1. (Case 1). Desmoid tumor in the pectineus muscle. The long axis of the tumor has the same direction as the muscle. The tumor is more vascular than the surrounding tissue and the vessels are of normal configuration but stretched in the periphery of the tumor.

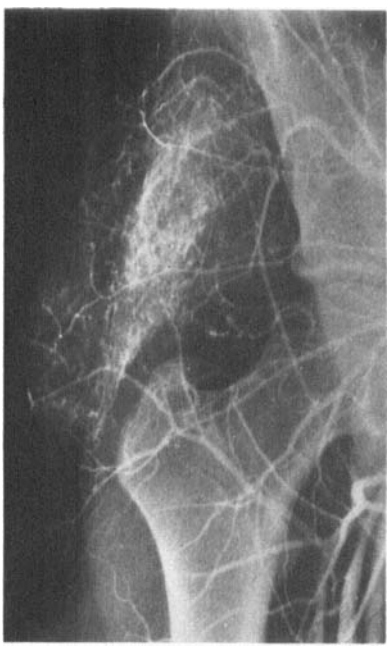


Figure 2. (Case 11). A poorly demarcated desmoid tumor within the gluteus medius muscle. The tumor does not involve the tendon of this muscle seen as the poorly vascularized zone outside the greater trochanter. The tumor is highly vascular with abnormal vessels simulating a malignant soft tissue tumor.

Fine-needle aspiration biopsy for cytologic diagnosis was carried out in 11 patients. In four of them, the diagnosis was a benign lesion and in five, a "semi-malignant" lesion; in the remaining two patients the material was insufficient for diagnosis. In no case did the cytologist classify the lesion as fully malignant.

Incisional biopsy was carried out in 14 patients (in two of them also fine-needle aspiration biopsy). In nine of them, the diagnosis was desmoid tumor or a locally aggressive fibromatous tumor, in two, an unspecified benign mesenchymal lesion, in two, a fibrosarcoma, and in one, an inflammatory process.

Surgical procedures

All but one patient underwent surgical treatment. From the medical records it was possible to make an estimation of the adequacy of the surgical procedure as defined below. In this retrospective study we found it difficult to follow strictly the definitions proposed by Enneking et al. (1980).

Inadequate excision was carried out in 16 patients. This means that the knife had cut too close to the tumor in one or several places with risk of microscopic tumor tissue having been left in the surgical field.

Probably adequate excision was carried out in 10 patients. This means that the tumor had been removed within the anatomical compartment in which is was located, but with a probably adequate margin of healthy tissue surrounding it.

Adequate excision was carried out in 24 patients. This means that the anatomical compartment in which the tumor was located had been removed unopened along with its fibrous boundary (sometimes a single muscle enclosed by its fascia). In four of these patients, the operation was done within a few weeks after an initial inadequate operation, with no signs of local recurrence.

Amputation was carried out in two patients. Both underwent an interscapulothoracic amputation.

Postoperative radiation therapy was given after inadequate excisions in three patients (30–40 Gy). Furthermore, one patient with multiple tumors in the right leg received radiation therapy against both recurrences and a new tumor.

Results

Descriptive data. There were twice as many females as males. The mean age at operation for men was 40 (18–75) years and for women 45 (21–81) years. The tumor was less than 5 cm in 13 patients, and more than 10 cm in 12. The most common locations were the shoulder region, the thoracic wall, and neck (Table 2). The duration of symptoms was less than 6 months in 28 patients, and more than 1 year in 13 patients.

In six women, the tumor was first noticed during pregnancy. Three patients had systemic connective tissue diseases (ankylosing spondylitis, rheumatoid arthritis, periarthritis nodosa), and one patient had liver metastases from an adeno-carcinoma of the colon.

Multicentric appearance of the tumor was seen in one patient, and spontaneous regression was seen in one patient. Malignant transformation of the tumor did not occur.

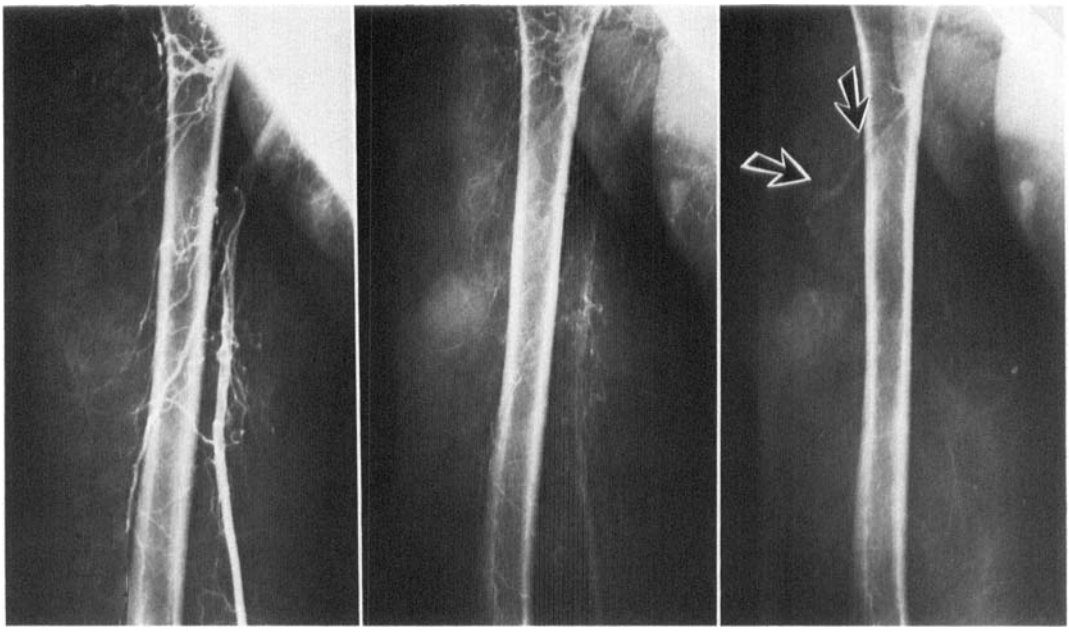
Gross appearance. The lesions presented as firm, unencapsulated, poorly demarcated masses, often invading muscles and muscle fascia. In one case, it invaded bone. The cut surfaces showed a grey-white, whirled pattern.

Microscopic appearance. The tumors characteristically were moderately cellular, composed of spindle cells of fibroblast type and abundant collagen arranged in sweeping bundles. Mitotic figures were infrequent.

Usually there was a uniform distribution of spindle cells and collagen throughout the tumors, although occasionally hyalinized areas were seen. In some tumors there were small

Table 2. Location of 53 extra-abdominal desmoids

	No. of tumors
Neck	9
Shoulder	18
Upper arm	3
Below elbow	2
Thoracic wall	10
Mediastinum	1
Hip-buttock	5
Thigh	4
Foot	1



A B C

Figure 3. (Case 3). A desmoid tumor in the triceps brachii muscle. Note the stretched artery in the periphery of the tumor (A and B), the opacification, and the wide vein draining the tumor (C).

areas with a prominent mucoid matrix. The tumors usually infiltrated, often diffusely, the involved muscle (Figure 3).

The vessels were typically slender and elongated with their long axis parallel to the direction of the tumor fascicles. They were fairly uniformly distributed throughout the lesions and with a diameter ranging from 10 to 100 μm. They appeared as open primitive vessels lined with a single row of prominent, sometimes protruding endothelium, enclosed by a thin rim of fibrous tissue. In the periphery as well as in bands of more regular fascia-like connective tissue, interwoven within the tumors, larger vessels could also be seen: mature arteries and veins, many of which measured 1–2 mm in diameter, and vessels of arteriol, and venol type. Staining for elastin brought out the elastic components of the arteries and veins. The dominating primitive tumor vessels contained no elastin.

Survival. One patient died from the tumor disease; a large recurrent tumor in the mediastinum compressed the trachea and caused suffocation. Another patient who had a tumor in the supraclavicular fossa died 1 month postoperatively with respiratory complications.

Local recurrence. Of the 47 patients who were followed up for at least 5 years, two did not survive this observation period and one received only radiation therapy. The local recurrence rate was studied for the remaining 44 patients. Seventeen patients had a local recurrence after the first surgical attempt. The adequacy of the surgical procedure was of great importance for achieving local control of the primary tumor.

Only three patients out of 21 had a recurrence after an adequate local excision, whereas after an inadequate excision 10 out of 14 patients had a recurrence. Four out of the seven patients who had undergone a probably adequate excision had a recurrence but neither of the two patients who had undergone a primary interscapulothoracic amputation had a recurrence. The size of the tumor, the age of the patient, and the duration of symptoms preoperatively did not correlate to the local recurrence rate. Tumors located in the upper arm and the thoracic wall tended to recur more often than tumors located elsewhere.

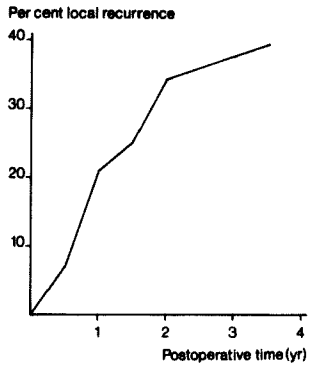


Figure 4. Timing of local recurrence in 17 patients with extra-abdominal desmoid tumor. No recurrence appeared later than 40 months postoperatively.

Two of the three patients treated with postoperative radiation after an initially inadequate operation were free from recurrence. One woman with multiple occurrence of desmoids was irradiated against an extensive recurrence in the groin and the foot. The tumor diminished slowly during 5 years and she is still alive with this tumor and a new tumor in the hamstrings.

The time-interval from operation to recurrence is shown in Figure 4. Fifteen of the 17 recurrences were within 2 years. The latest recurrence was seen 40 months postoperatively.

Treatment and final outcome of local recurrence. There was local recurrence in 17 patients, once in seven, twice in six, three times in two, and four times in two. There was no further recurrence in 10 patients after an adequate excision of the tumor-bearing region, in two patients after an interscapulothoracic amputation, and in one patient after a below-knee amputation. In two patients the first recurrence was successfully treated by a new inadequate excision. In one patient with four local recurrences, the first and second were treated by a probably adequate excision and the third and fourth with an inadequate excision. The patient was free from tumor at follow-up 27 years later. One patient is still alive with a local recurrence treated only by radiation.

Discussion

Extra-abdominal desmoid tumors are rare. In 1948, Musgrove & McDonald reviewed the literature, established criteria for histologic diagnosis, based on 34 cases, and gave some fundamentals for treatment. Very few series with long-term follow-up have been published (Hunt et al. 1960, Dahn et al. 1963, Enzinger & Shiraki 1967, Das Gupta et al. 1969, Rock et al. 1984).

All authors emphasize the discrepancy between the harmless microscopic appearance of the tumor and its potential to attain large size and the pronounced tendency to recur locally. The tumor arises from musculo-aponeurotic structures predominantly in the shoulder and the neck region. Typically the tumors are oblong and directed in the axis of contraction of the involved muscle. Several authors have noticed the lack of pseudo-encapsulation seen in malignant soft tissue tumors (Musgrove & McDonald 1948, Hunt et al. 1960). The tumor often has a thickened central portion with flattened irregular and non-palpable extensions along the muscle bundles throughout the muscle. Radical removal of the tumor must therefore include the tendinous origin and insertion of the muscle.

Among etiologic factors attention has been focused on trauma and hormonal factors. Rock et al. (1984) reported an episode of trauma in 19 per cent, and in our series we found a history of relevant trauma in nine patients. Hormonal influence on tumor growth with spontaneous regression of desmoids at the menopause and the menarche has been reported (Strode 1954, Dahn et al. 1963). In our series, the tumor occurred during pregnancy in five women, and in one woman the tumor increased in size during pregnancy.

Spontaneous regression has been observed and reported among others by Enzinger & Shiraki (1967). Their long-term follow-up (10 or more years) suggests that the tumor growth rate often decreases after an initial period of rapid growth and eventually cease to grow. Also Rock et al. (1984) reported that out of 68 patients with residual tumors the tumor activity decreased in 60 patients after an average follow-up of 6 years. They also found a more

aggressive growth in younger people, and a higher incidence of recurrence. In our series, complete spontaneous regression was noted in a woman at follow-up 18 years after a partial removal of the tumor performed at the age of 28. A 62-year-old woman lived 12 years after diagnosis of a desmoid tumor in the supraclavicular region with no sign of tumor growth after the first year.

Multiple desmoids occurring in the same extremity have been reported by Rock et al. (1984) in 19 and by Allen (1977) in three cases. Desmoids, sometimes multiple, have also been reported to occur in patients with familial polyposis coli (Gardner's syndrome) (Dahn et al. 1963).

A 25-year-old woman in our series had multiple occurrence of desmoids in her right lower limb. She is alive 8 years after the first operation but with persisting tumors in the groin and hamstrings which have ceased to grow during recent years.

The histologic diagnosis from an incisional biopsy has been a problem to pathologists. In our series the diagnosis was missed in five out of 14 biopsies, probably because several pathologists were involved and the majority of them had no special training in the pathology of soft tissue tumors. This problem has diminished in recent years with centralized treatment of soft tissue tumors and close collaboration by a team of trained pathologists and surgeons.

Angiography and, in recent years, CT are of great value in the preoperative work-up, especially for determining the anatomical location and extension of the tumor (Figures 1 and 2). In seven of the patients in whom angiography was performed, the anatomical location of the tumor could be determined. The location is usually a muscle or a muscle group. If a soft tissue tumor can be radically removed without any significant loss of function, the operation can be carried out without preoperative histologic diagnosis.

Radiographic bone changes with erosion and a periosteal reaction of spicules of bone radiating into a soft tissue mass have been described by Abramowitz et al. (1983). These changes are secondary to the local pressure of a deep-seated tumor resulting in a cortical defect or a periosteal reaction. Our case demonstrated another kind of bone erosion. The tumor invaded the bone and caused a total destruction of the fifth metatarsal head.

We have found only sporadic reports of angiographic findings in extra-abdominal desmoids in the literature (Yaghmai 1977, Tötterman & Reitamo 1979, Skaane et al. 1980). A normal vascular pattern with normal vessels, although sometimes with hypervascularity, is usually described. We found essentially the same angiographic features although pathological vessels were found in 2 of our 11 angiograms.

Although the natural history is not fully known and there are suggestions that the tumor ceases to grow with time, the recommended treatment is surgery. The fundamental principles in the management are the same as for a malignant soft tissue tumor. Unless the tumor is completely removed there is a high risk for recurrence, especially in younger patients (Enzinger & Shiraki 1967, Rock et al. 1984). It is even possible that inadequate surgery may stimulate the growth of desmoids (Allen 1977). When first diagnosed the desmoid should preferably be treated by complete removal of the affected muscle or muscle group. The local recurrence rate in our series was 39 per cent. In other reported series, the recurrence rate has varied between 19 and 68 per cent (Hunt et al. 1960, Enzinger & Shiraki 1967, Das Gupta et al. 1969, Rock et al. 1984). After adequate excision three out of 21 tumors recurred, whereas after inadequate excision 10 out of 14 recurred. The recurrence should be treated according to the same principles as primary tumors.

In our series, the recurrence was successfully treated by local surgery in 13 out of 17 patients. In three patients, the tumor ceased to recur after an inadequate operation. Two patients underwent interscapulothoracic amputation and one a below-knee amputation to control the recurrence. However, amputation for this non-metastasizing tumor should be seriously questioned. In our series, one patient lived with a tumor for 12 years and died of an unrelated disease. In another patient, the tumor regressed spontaneously, and one patient is alive with multiple tumors 8 years after di-

agnosis with no sign of progression. We think it justified to follow the recommendation of Rock et al. (1984), that a recurrent lesion that appears inactive should be followed by sequential assessments, unless the tumor can be removed without significant compromise of function.

The value of radiation therapy for desmoid tumors has been disputed. Most authors have been unable to observe any significant effect, but the small number of patients treated with irradiation have usually been included in surgical series. Conclusions about the value of radiotherapy cannot be drawn from our study.

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