EXTRA-ABDOMINAL DESMOID TUMOURS

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Desmoid tumours are not common but have a distinct resemblance to fibrosarcomata. Their clinical appearance and progress should be recognised since failure to distinguish them from sarcomata could result in extensive and unnecessarily mutilating operations.

Three cases of extra-abdominal desmoid tumours are reported. Two of the patients had tumours arising from multicentric foci in the same limb. The disappearance of the tumours in two patients after the menopause, and the variation in the tumours during the menstrual cycle in the third patient, add weight to the theories about endocrine control.

The term desmoid tumour is used to describe a curious neoplastic proliferation of fibrous tissue with some features of both a benign fibroma and a malignant fibrosarcoma (Robbins 1962). It was first mentioned by Dr J. McFarlane of Glasgow in 1832 when he recorded the histories of two patients with organising tumours between the muscular layers of the abdominal wall. The desmoid tumour found in the abdominal wall of some women who have borne children is well recognised and described in textbooks of surgery and pathology. It has been stated that the majority of desmoid tumours occur in women, and their frequency after pregnancy has been attributed to the violent muscular contractions of parturition. Desmoid tumours are said to be most common in the third to fifth decades but they may present at any age and in either sex. Apart from the more common abdominal tumours, histologically identical tumours occur in other parts of the body, particularly in the shoulder girdle and arm, in the thigh and in the buttock where they present considerable diagnostic problems.

The features of the desmoid tumour are that it is a proliferation of fibroblasts; it is locally invasive but does not metastasise; it does not arise from muscle but rather from the musculo-aponeurotic structures of the body; it is, properly speaking, a tumour of the supporting tissues of muscles. The presenting sign is a firm, rapidly growing swelling. Pain is not a marked feature unless there is pressure on surrounding structures. The overlying skin is freely movable but the tumour is fixed to the involved muscles. A histologically similar tumour is found in bone (Jaffe 1958).

Microscopically the lesions are grey-white, firm, non-encapsulated and poorly demarcated, varying in size from 1 to 20 centimetres. The tumours are firm and rubbery, invading muscles and muscle bundles; in two of our cases the tumour had invaded the trabecular spaces of adjacent bones. Histologically they resemble cellular fibromata, the individual cells being uniform in size and shape; the absence of mitotic figures and giant cells helps to distinguish desmoid tumours from fibrosarcoma (Figs. 1 and 2).

We report three cases of extra-abdominal desmoid tumours; two of them featured multicentric foci in a single limb, a characteristic first noted by Barber, Galasko and Woods (1973).

CASE REPORTS

Case 1. A woman aged twenty-five years was referred to our clinic in 1949 on account of a swelling of her right foot. It consisted of a hard mass involving the forefoot, palpable both on the dorsum and on the sole and fixed to the metatarsals. The patient could not say how long the swelling had been present, but for several weeks before her attendance it had increased rapidly in size and she had been unable to put on her shoe.

Radiographs showed involvement of the metatarsals (Fig. 3). Because of the history and radiographic appearance, the question of malignancy was discussed and biopsy undertaken. The Pathology Department reported an “active” fibroma but did not consider it to be malignant. As much of the tumour mass as possible was excised, but it was recognised that excision was far from complete. The patient had informed us at the time of her admission that she was three months pregnant. Physical examination was normal in all other respects, as were the biochemical investigations.

A recurrence of the growth was excised one year later and this patient had no further trouble until 1953, when a lump appeared over the right abdominal wall. At this time she was again three months pregnant so she was referred to the Gynaecology Department where a classical abdominal desmoid tumour was removed.

In 1955 a rapidly growing lump, about the size of a closed fist and causing discomfort when she sat down, appeared in the upper posterior third of her right thigh. It had invaded the hamstring muscles but was mobile in the transverse plane, and its removal did not present a great problem though the bellies of the involved muscles had to be sacrificed. The histological findings were as before, but the pathologist felt that we had to consider the possibility of fibrosarcoma. After discussion, however, it was agreed that the tumour was a desmoid and that a further extensive operation was not indicated.

In 1961 a lump appeared in the right thigh proximal to the
Fig. 1
Photomicrograph showing the main histological features of a desmoid tumour. The picture is that of a fibroma with benign-looking cells in a fibrous matrix. The absence of mitotic figures and giant cells helps to distinguish these tumours from fibrosarcomata. (Haematoxylin and eosin, ×125.)

Fig. 2
Photomicrograph showing the interface between desmoid tumour and muscle. The close apposition between the tissues is seen with an inflammatory cell exudate at the interface. (Haematoxylin and eosin, ×125.)
previous tumour. It was fixed deeply to the ischium and produced symptoms from pressure on the sciatic nerve. At operation the tumour was more extensive than had been apparent. It had infiltrated the semitendinosus and semimembranosus, extended under the gluteus maximus involving it and the pyrififormis, enveloping the sciatic nerve, which was dissected free with great difficulty. The greater part of the involved muscles had to be sacrificed in its removal. The pathologist who studied the specimen said that he would regard it as a low-grade fibrosarcoma; once again after discussion the section was accepted as that of a desmoid tumour.

Case 2. A woman aged thirty-seven on first attendance presented at the Orthopaedic Clinic in 1971 with intermittent backache and left-sided sciatica of several years duration. She had also noted a firm swelling on the posteromedial aspect of the thigh which was occasionally painful.

Examination revealed a firm swelling about the size of a large apple lying between the hamstrings and the adductor muscles. The swelling was smooth and rounded and attached proximally to the muscle origins around the ischiopubic ramus; it was not painful. Radiographs of the lumbar spine and pelvis revealed no bony abnormality, and the history of backache and sciatica was not thought to be related to this swelling in the thigh. Routine physical examination and laboratory tests were negative.

Exploration and excision of the tumour was undertaken in April 1971. A firm tumour was encountered beneath the semimembranosus and adductor magnus; the latter muscle was infiltrated by strands of tumour tissue particularly near its origin. The tumour was densely adherent to the sciatic nerve at the upper border of the adductors. The bulk of the tumour mass was excised but it was realised that excision was incomplete. Histological examination of the specimen showed what was thought to be a well-differentiated fibrosarcoma; on further study the diagnosis was revised to that of a musculo-aponeurotic fibromatosis. The pathologist added that while the tumour was unlikely to metastasise it was likely to recur locally and to spread, an opinion which events proved to be correct.

Between June 1972 and April 1974 the patient underwent five further explorations of the posterior aspect of the thigh for excision of the recurrent tumour. On each occasion a marked feature was the involvement of the sciatic nerve and after the third operation the patient developed a foot drop which has persisted. At the sixth operation the tumour was found to have entered the pelvis through the greater sciatic notch in company with the sciatic nerve. At this point the opinion of a radiotherapist was sought, to determine the possibility of treatment since reports indicated that the tumour became quiescent after the onset of the menopause (Musgrove and McDonald 1948). Endometrial curettings were normal so an induction of the menopause was carried out by irradiation when the patient was aged forty-one years.

In the four years since the treatment there has been no local recurrence in the thigh and the patient has been in good health, the only remaining feature being the foot drop.

Case 3. A girl aged sixteen years came to the Orthopaedic Department in December 1974 complaining of a swelling above the inner aspect of the right elbow. She had noticed the swelling over a period of nine months, but it had recently become larger and slightly painful. There was a diffuse, firm swelling extending from the medial epicondyle for a distance of five centimetres up the arm; it was firmly adherent to the underlying structures. Radiology revealed erosion of the humerus at the site of the tumour (Fig. 4). Physical examination was otherwise normal. At operation a firm fibrous tumour was found arising from the common flexor origin, infiltrating the muscles, the joint capsule and the underlying bone; the ulnar nerve was in the substance of the growth and great difficulty was experienced in dissecting it free. The pathologist who examined the sections said that the possibility of a low-grade fibrosarcoma had to be considered, but he obtained a second opinion and it was agreed that it was fibromatosis. The patient returned to work but was seen in August 1976 with a local and extensive recurrence of the tumour; movements of the elbow were restricted and the joint was fixed in flexion. Radiography revealed increased erosion of the lower shaft of the humerus (Fig. 5). The tumour was excised with great difficulty; again the ulnar nerve was completely enveloped and spared only by tedious dissection.

Professor Mackenzie of the Department of Histopathology, Westminster Medical School, saw a section of the tumour and diagnosed musculo-aponeurotic fibromatosis; his report concluded, "There is no doubt this will recur and will continue to do so until the lesion has been completely excised." His prediction was correct: within three months there was evidence of return of the growth on the
inner aspect of the elbow. It was decided to temporise and to offer gratification the tumour began to diminish in size.

In April 1977, however, a second tumour appeared over the deltoid region on the same arm and grew rapidly, involving the deltoid and part of the triceps. It was fixed deeply to the humerus, though there was no radiological evidence of involvement of the bone. By this time the tumour on the inner side of the arm had resolved and could not be palpated (Fig. 6). The two tumours were discrete and had no connection. In view of the behaviour of the first tumour a policy of waiting was adopted, but in January 1978 the patient complained of pain in the deltoid region. A red fluctuant area in the middle of the tumour was explored and found to be a haematoma, probably due to injury at work. A large section of the tumour was excised, part of which was sent for histological examination and part for hormonal assay. It had been noted that pain was a variable feature, and when questioned the patient stated that her pain was worse premenstrually; she also mentioned that her periods were irregular. Both the cytoplasm and the nucleus from the tumour cell contained a significant level of oestrogen receptor, and a trial of tamoxifen was recommended. 10 milligrams three times daily. This drug was administered for two months without any obvious effect on the tumour. As the patient was a young and premenopausal girl it was decided on ethical grounds to discontinue the drug in the absence of clinical improvement.

DISCUSSION

Desmoid tumours are unique in their clinical manifestation. They are so locally invasive that they appear malignant but metastatic spread does not occur. Complete surgical excision is usually impossible due to involvement of important nerves and blood vessels, and local recurrence is common.

The desmoid tumour of the abdominal wall is familiar. Extra-abdominal desmoids are not so well known but have been reported in the literature with conflicting theories about their aetiology and the best form of treatment (Musgrove and McDonald 1948; Strode 1954; Ramsey 1955; Hart, Morgan and Ackerman 1960; Enzinger and Shiraki 1967). When these lesions do occur extra-abdominally, they have a predilection for the muscles of the shoulder girdle and upper arm, the buttock and the thigh. In all previous reports except one, the purely extra-abdominal desmoid tumours have been unicentric in origin with direct spread; the exception was the report by Barber et al. (1973) of two cases in which extra-abdominal desmoid tumours arose from multicentric foci in a single limb.

The three cases described in this report are considered to be of interest on three grounds. First, in two of the cases the tumour arose from multicentric origins in a single limb, as described by Barber et al. (1973). Secondly, in two of the reported cases there was involvement of bone which increased the problems of diagnosis and treatment. Thirdly, the course of the tumours has led us to suggest treatment which is at variance with Barber’s recommendations.

These three cases differed greatly from those described by Barber et al. (1973); our patients were female, theirs were male. There was nothing to suggest that injury was an aetiological factor. In two of the cases the tumour regressed after the onset of the menopause, this being therapeutically induced in one patient at the age of forty-one. In the youngest of our patients the
behaviour of her tumours fluctuated during her menstrual cycle, and her periods were irregular. These facts tend to substantiate the theories that there is endocrine influence on the course of this condition, as previously noted by Strode (1954) and Dahn, Jonsson and Lundh (1963).

There have been no previous reports of involvement of bone by a desmoid tumour as occurred in two of our cases. The term desmoplastic fibroma was used by Jaffe (1958) to describe an intra-osseous fibrous tumour, distinct from fibrosarcoma, and various benign fibrous lesions of bone. The term desmoplastic fibroma was used because of histological similarity to the more familiar desmoid tumour of the abdominal wall. These tumours, however, differ from the cases recorded here in that they remain intra-osseous and cause bone expansion, unlike the destruction from outside caused in our patients; also, the desmoplastic fibroma is intra-osseous in its origin and course. Further cases of desmoplastic fibroma have been described by Dahlin and Hoover (1964), Godinho, Chiconelli and Lemos (1967) and Sugiura (1976).

The course of the tumours in our patients incline us to make different recommendations to those made by Barber et al. in 1973, although we must stress the basic differences of sex in the groups reported. We agree with Strode (1954) that desmoid tumours affecting musculo-aponeurotic structures outside the abdominal wall continue to pose problems in interpretation and management. Musgrove and McDonald (1948) said that many surgeons have not realised that only a radical local excision will ablate the lesion. Enzinger and Shiraki (1967) reviewed thirty cases of tumours affecting the upper arm and shoulder girdle, with a follow-up of at least ten years after initial diagnosis. They found that thirteen patients were cured by the initial treatment of wide local excision. Seventeen tumours, however, recurred after local excision and required further operations, one patient having four more operations. All these patients were alive and free of recurrence no less than ten years after initial diagnosis, and these authors concluded that while the chances of survival were excellent the prognosis in regard to recurrence was poor.

While radical amputation will completely ablate the tumour in most cases, a less than radical amputation may leave some of it behind in the stump if the tumour is multicentric in origin. Also, in one of our patients the tumour entered the pelvis via the greater sciatic notch; we feel that even hindquarter amputation would not have totally ablated the tumour in such a case. Accordingly we suggest the following procedure if an extra-abdominal desmoid tumour is suspected.

Biopsy should be carried out to verify the diagnosis and distinguish the tumour from fibrosarcoma. If it grows to such a size as to interfere with function then the tumour should be excised. We agree with Ramsey (1955) that incomplete local excision is preferable to sacrificing major peripheral vascular and neural structures, or sacrificing an extremity unless there is a risk to life.

The place of hormones in the treatment of these tumours remains unclear. In our third case, a girl aged sixteen, study of the tumour by techniques devised for breast cancer did show oestrogen receptor sites. An anti-oestrogen agent, tamoxifen, was prescribed, but administration was not prolonged in view of the lack of response of the tumour and the age of the patient.

Why multicentric lesions should confine themselves to one limb is not known. It is interesting that fibrous dysplasia of bone often affects only one half of the skeletal system and that it is histologically a developmental overgrowth of fibrous tissue in bone.

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