MULTICENTRIC EXTRA-ABDOMINAL DESMOID TUMOURS

Report of Two Cases

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Desmoid tumours consist of locally invasive, non-metastasising proliferations of fibroblasts, which may arise from any musculo-aponeurotic tissue, and apparently infiltrate between adjacent muscle fibres. The term includes those histologically identical tumours occurring in the abdominal parietes and elsewhere—that is, the extra-abdominal desmoid tumours. All the previously reported cases of purely extra-abdominal desmoid tumour, without associated pathology, have been of unicentric origin. This report is of extra-abdominal tumours arising from multicentric foci in a single limb.

CASE REPORTS

Case 1—A twenty-five-year-old foreman-carpenter was admitted to the Nuffield Orthopaedic Centre in December 1967 with a two-year history of a constant dull ache in the left shoulder. There was limitation of movement of the shoulder and there was a hard mass, apparently

FIG. 1

Case 1—First resection of lesion in supraspinatus muscle. Bisected specimen showing the interlacing arrangement of fibrous bundles.

attached to the humerus, at the deltoid insertion. Physical examination was otherwise normal. Haemoglobin was 17 grammes per 100 millilitres, white cells 7,000/cubic millimetre, sedimentation rate 1 millimetre/hour (Westergren), alkaline phosphatase 8.6 King-Armstrong

units, and acid phosphatase 1 King-Armstrong unit. Radiographs showed no abnormality of the humerus. The mass was excised: histological examination showed poorly cellular collagenous fibrous tissue with some muscle fibres of normal appearance. A specimen of the underlying bony cortex appeared microscopically normal apart from the insertion of poorly cellular fibrous tissue on the external surface. There was no evidence of neoplasia.

Symptoms persisted and the swelling recurred six months later. In October 1968 a swelling was also noticed in the left supraspinous fossa. There was no biochemical or haematological

FIG. 2

Case 1—Histological section of tumour showing uniform cellularity and abundant collagen production characteristic of a desmoid tumour. Blood vascular channels, clearly distinguishable from the connective tissue of the lesion, are present throughout. (Haematoxylin and eosin, $\times 150$.)

abnormality. Local excision of this “well encapsulated” mass (Fig. 1) from the supraspinatus muscle was carried out: it was a fibroblastic, moderately cellular and well defined lesion, with uniform cellular morphology and without significant mitotic activity (Fig. 2). The probability of a “desmoid tumour” was suggested. This was confirmed in January 1969 after partial excision of a further recurrence of the deltoid tumour and of a third mass, arising in the posterior axillary wall.

Pain and stiffness continued; the deltoid tumour recurred within three months and that in the supraspinatus in eighteen months. Extensive local resection of both tumours was performed in November 1970 and invasion of the infraspinatus was noted. The deltoid tumour again recurred within six months and by November 1971 there was a recurrence of tumour in the supraspinatus and infraspinatus muscles and in the deltoid muscle, and a new tumour appeared in the triceps, close to the old scar. A new tumour, not related to previous incisions, appeared in the latissimus dorsi muscle. Urinary 17-oxysteroids were 25.1 milligrams/24 hours and 17-hydroxycorticoids 19.2 milligrams/24 hours. Because it had proved impossible to control the lesion by local excision, forequarter amputation was carried out in November 1971.

There has been no recurrence of tumour since amputation.

Case 2—The patient, a boy, initially presented in 1956, aged two and a half years, with unilateral bowing of the left fibula which was treated by resection of a segment of bone (Fig. 3). The histological appearance was normal. Six months later elongation of the left calcaneal tendon was performed for persistent equinus deformity, but this recurred soon after operation.

At the age of four the patient fell from a tricycle and sustained a fracture through the shaft of the same femur: this united satisfactorily with the limb immobilised in a Thomas splint.

At the age of seven the boy developed a swelling in the left buttock. This swelling was excised. Histological examination was said to show a "low grade fibrosarcoma", and excision was followed by irradiation. The swelling recurred and increased in size despite further radiotherapy, and two years later, in 1963, a hindquarter amputation was carried out for severe sciatic pain.

Dissection of the disarticulated limb revealed three separate foci of abnormal tissue in the glutei, in the gastrocnemius and in the tendo calcaneus. The sciatic nerve was surrounded by the proximal lesion but was clearly separated from it. Histological examination of the lesions showed well differentiated fibroblasts with abundant collagen and with the internal arrangement and relationship to adjacent tissue of desmoid tumours (Figs. 4 and 5). There has been no recurrence of tumour after amputation.

DISCUSSION

Extra-abdominal desmoid tumours have been regarded as a distinct clinical entity since 1923 (Nichols), although they were initially described by Bennett in 1849.

The incidence of these tumours is unknown but, in their series of desmoid tumours seen in the Mayo Clinic between 1908 and 1945, Musgrove and McDonald (1948) reported eighty-five abdominal desmoids and thirty-four desmoids occurring in extra-abdominal sites.

Extra-abdominal desmoid tumours are classified within the spectrum of benign fibromatoses. These occur predominantly in adults and range from palmar and plantar fibromatoses to the more invasive forms such as mesenteric fibromatosis, Gardner's syndrome and desmoids. The importance of accurate diagnosis of desmoid tumours lies primarily in their differentiation from fibrosarcomata. Clinically they present as radio-resistant, locally invasive neoplasms with a recurrence rate of up to 50 per cent after inadequate excision (Enzinger and Shiraki 1967). Although desmoids are essentially benign tumours, death may result from pressure on and invasion of vital structures, as in the neck or mediastinum (Stout 1954). They do not metastasise.

The etiology is unknown but the two main theories of the causation of these tumours implicate trauma and hormonal imbalance. The evidence offered in favour of trauma is often circumstantial, and microscopic proof is lacking. Musgrove and McDonald (1948) found that fourteen of thirty-four patients related the onset of the tumour to trauma but in only three cases were there microscopic traces of haemosiderin. Occurrence in an operation scar has often been reported in relation to abdominal desmoid but of 122 recently reported extra-abdominal desmoids (Ramsey 1955; Enzinger and Shiraki 1967; Das Gupta, Brasfield and O'Hara 1969; Cole and Guiss 1969) only six occurred in surgical incisions—one after excision of a lipoma (Ramsey 1955); one after excision of an angioliipoma from a child (Das Gupta and colleagues 1969); two after thoracic operations (Enzinger and Shiraki 1967); and two after unspecified operations (Cole and Guiss 1969). The first patient whose case is reported here was a manual worker, but denied specific trauma and claimed to carry loads on his right shoulder. The trauma of operation and later of the fall which produced the femoral fracture suggest that trauma may have been a more significant causal factor in Case 2.

The evidence for an endocrine factor in the etiology is based mainly on the production

FIG. 3

FIG. 4

Case 2. Figure 3—Radiograph showing deformity of left fibula at the time of first attendance. Figure 4—Photomicrograph of gluteal lesion. Well differentiated and poorly cellular fibrous tissue has infiltrated between muscle fibres, which are atrophic and distorted. (Haematoxylin and eosin, $\times 150$.)

FIG. 5

Case 2—Popliteal lesion. Well differentiated fibrous tissue has infiltrated the posterior tibial nerve, two bundles of which are included in this field. There is considerable loss of myelin sheaths but the bundles are structurally intact. (Haematoxylin and eosin, $\times 150$.)

of fibrous tumours in guinea-pigs after oestrogen administration and the prevention of these experimental tumours by the administration of testosterone, progesterone and deoxycorticosterone (Lipschutz 1950). Bioassay of one abdominal desmoid in 1935 by Geschickter and Lewis demonstrated 13,000 rat units of gonadotropic substance per kilogram, but no oestrogen. On the other hand Ober, Velardo, Greene and Taylor (1955) found no gonadotropic, progestational, adrenotropic, thyrotropic or oestrogenic hormones in a popliteal desmoid occurring during pregnancy. Spontaneous regression of desmoids at the menarche and at the menopause have been reported (Strode 1954; Dahn, Jonsson and Lundh 1963). Pack and Ehrlich (1944) reported an inoperable abdominal desmoid which regressed after radiation castration.

Our patients were normal males with no evidence of endocrine abnormality; in Case 1 the 17-oxyteroids and 17-hydroxycorticoids were at the upper end of the normal range.

The age of onset ranges from birth to eighty-two years, but most tumours occur in young adults, females being affected twice as often as males. The most common site is the shoulder girdle and upper arm (Musgrove and McDonald 1948, Ramsey 1955, Enzinger and Shiraki 1967, Das Gupta and colleagues 1969, Cole and Guiss 1969).

The time taken to establish the definitive diagnosis in our cases exemplifies the common clinical and pathological problem. The histological features of extra-abdominal desmoids have been thoroughly discussed by Musgrove and McDonald (1948) and contrasted with those of fibrosarcoma. By definition, a desmoid tumour contains engulfed striated muscle fibres, but the microscopic appearance of tumour adjacent to, and compressed against, periosteum or aponeurosis does not show muscle fibre inclusions, as was found in the first specimen in Case 1.

In Case 1 the clinical encapsulation of the tumour in the supraspinatus muscle was due to the intact and distended epimysium but the presence of a discrete cellular boundary was not confirmed microscopically.

The low degree of cellularity, and the absence of mitotic figures and tumour giant cells were the main evidence for distinguishing our desmoids from fibrosarcomata. Haemosiderin was not looked for in our two cases. Other histological characteristics were not considered by Musgrove and McDonald to be of specific value in borderline fibrogenic tumours.

McAdam and Goligher (1970) stated that "any patient who is found to have a desmoid should be examined for polyposis coli". The maternal grandfather of the patient in Case 1 died from carcinoma of the rectum, but no paternal family history is available. No family history is available in Case 2. Sigmoidoscopy and double contrast barium enema were normal in both cases.

Treatment—In surgically eradicable disease there seems to be no place for radiotherapy; the only case report of malignant change in an extra-abdominal desmoid followed radiotherapy (Soule and Scanlon 1962). Despite the suggested hormonal factors, the value of endocrine therapy is unproven.

Radical local excision is recommended for the solitary extra-abdominal desmoid. It should, if necessary, be repeated for local recurrence (Enzinger and Shiraki 1967) and total excision of the involved muscle is desirable to prevent recurrence (Cole and Guiss 1969). Local operation, without total excision of the involved muscle, was attempted in our cases but was unsuccessful in controlling local recurrence.

Stout (1954) has reported death from invasion of neck and mediastinum by desmoid tumours, and in view of the multicentric origin and the proximity of the lesions to the neck and mediastinum, forequarter amputation was carried out in our Case 1.

The observation that in both patients these multiple tumours occurred in a single limb bud suggests that total limb amputation may be necessary to prevent recurrence in this type of multicentric extra-abdominal desmoid, particularly if the alternative is a useless, mutilated, painful limb.

SUMMARY

1. Two patients with multicentric extra-abdominal desmoid tumours are reported. Multicentric foci have not been described previously.
2. Both cases conform to the usual clinical, macroscopic and microscopic criteria of desmoid tumours in all other regards.
3. Their occurrence in a single limb bud suggests a congenital propensity and leads to the advocacy of radical surgery in such cases.

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