

**DESMOID TUMOURS—A REVIEW**S. BHATTACHARYA, B. K. ACHARYA, M. S. D. JAISWAL, D. C. SRIVASTAVA  
A. CHATURVEDI N. C. MISRA AND R. P. SAHJ**SUMMARY**

*Seventeen cases of Desmoid tumours, 7 abdominal and 10 extraabdominal were chosen for this study and managed in the last five years. A wide local excision and one stage reconstruction was attempted in them. Simple reconstructive measures viz. primary closure aided by internal fasciotomies and split skin grafting were found adequate for extra abdominal sites whereas abdominal desmoids after ablation usually left a defect requiring prolene mesh and local skin flaps.*

Desmoid tumour is a fibroblastic proliferative lesion, derived primarily from fascial sheaths and musculoaponeurotic structures and in terms of anaplasia falls somewhere between a hyperplastic connective tissue and a low grade fibrosarcoma. Because of its tendency to infiltrate exclusively into surrounding structures a wide resection is attempted to achieve tumour free margins. This in turn results in a large soft tissue defect which is a challenge to the ingenuity of a plastic surgeon. Local recurrences are still not uncommon and a strict follow up and repeated surgical interventions are often required to achieve long term survival.

**Material and Method**

17 cases of desmoid tumours, 7 abdominal and 10 extra-abdominal are being reviewed in this study. The site of tumours is mentioned in (Tab. 1). 13 patients were females and 4 males. They were between 28 to 56 years of age. Six patients were operated once only. Eight patients were operated once more for recurrence after the first surgery. Two patients were operated 3 times and one on 5 occasions. No other modality of treatment except surgery was tried in these patients.

The virgin cases were first biopsied and on confirmation of the diagnosis a wide tumour excision was done. In the recurrent cases a wide tumour excision was repeated whenever

**Table 1.**

Abdominal	Extra Abdominal
1. Epigastrium —2	1. Upper arm — 1
2. Hypogastrium—5	2. Shoulder — 2
	3. Lateral chest wall — 2
	4. Upper arm, shoulder and lateral chest wall — 2
	5. Knee & Leg (Lt) — 1
	6. Neck and sub-mandibular region — 1
	7. Scapular region — 1

**Table 2. Mode of repair (Extra abdominal desmoids)**

Total No. of cases	— 10
Total No. of operations	— 23
Primary closure with internal fasciotomies	— 10
Split skin grafting	— 9
Local skin flaps	— 2
Fasciocutaneous flap	— 1
Myocutaneous flap	— 1

**Table 3. Mode of repair (Abdominal desmoids)**

Total No. of cases	--	7
Total No. of operations	--	10
Primary anatomical closure	--	3
Closure by prolene mesh and local skin flaps	--	7

the first recurrence was observed on palpation. Upto this stage the surgery was same for both abdominal and extra-abdominal desmoids. Hereafter the management of the resultant defect varied.

In the abdominal desmoids an anatomical repair of the abdominal wall was successful in 4 patients after the first operation but 3 of them recurred and had to undergo wider excisions. This time, however, a prolene mesh was used to substitute the missing rectus assembly and adjoining muscles and local skin flaps were designed to close the skin defects (Fig. 1 A-F). In the remaining 3 abdominal desmoids the first resection was large enough to demand a prolene mesh repair.

Of the extra abdominal tumours the lesions in the upper arm (Fig. 2), shoulder, lateral and posterior chest wall were closed primarily. The peripheral extensions of the tumour, beyond the limits of skin fixity acted as a tissue expander and aided in primary closure. The lesions in the lateral chest wall despite their large size could be closed primarily by undermining the anterior and posterior fasciocutaneous units upto the midline and longitudinal internal fasciotomies (Fig. 3) The desmoid in the scapular region was operated thrice and on the last occasion a lateral thoracic region flap (Bhattacharya et al., 1988) was used to cover the defect. The one in the neck and submandibular region after wide tumour clearance resulted in a large tissue defect which was covered by a pectoralis major myocutaneous flap (Ariyan et al., 1979). Interestingly, though the neck mass looked like a lobular lymph node mass, no lymph nodes in the cervical

chain were found involved on exploration. As there is paucity of skin in the leg the desmoid in this region was excised 4 times and a split skin graft was applied on each occasion. The last recurrence was over the knee joint (Fig. 5) which too was treated similarly.

### Observation and Results

17 cases of desmoid tumours between 28 and 56 years of age were treated in the last 5 years. Of the 7 patients who were treated for their abdominal desmoids, 3 recurred within a period of 1½ to 4 years. In all of them an anatomical repair was attempted previously but subsequently a prolene mesh and local skin flaps had to be used to close the defect left after the repeat surgery.

Only 2 extra-abdominal desmoids, the one in the neck and submandibular region and the one in the scapular region have remained tumour free for 3 years now. The remaining 8 patients recurred in between 8 months to 3 years. A twenty eight year old girl who had a desmoid in the upper arm was excised and primarily closed. She had a recurrence after 1½ years. It was again excised and closed primarily but it again recurred after 2 years and 10 months of tumour free interval (Fig. 2).

In the 2 patients who had desmoids over the shoulder, the tumour was excised and primarily closed in the first operation but the recurrence in both of them demanded the sacrifice of a large area of skin and the resultant defect was split skin grafted. They are now tumour free for the last 2 years and 8 months respectively.

The lateral chest wall desmoids were excised with a wide skin defect. (15×11 cm). They were primarily closed by widely undermining the anterior and posterior flaps upto the midline and doing internal fasciotomies longitudinally. We detected recurrences in both these patients after 16 months and 24 months, but as these were less than 4 cm in size in transverse diameter, a repeat excision and primary closure could be achieved in one

and a Z plasty had to be fashioned in another one (Fig. 3A & B).

A 36 years old lady had an upper arm desmoid which was excised and the defect primarily closed. She, however, developed a recurrence in the posterior axillary fold after 8 months (Fig. 4A) and this required a second wide excision mobilization of the anterior and posterior skin flaps longitudinal internal fasciotomies and primary closure (Fig. 4B). Yet

another female, 38 years old, had a horse shoe shaped lobular desmoid mounting the deltoid, acromion, and posterior axillary region. This was excised and closed by multiple Z plasties primarily. 3 years later she had a recurrence which was excised with tumour free margins and the raw area was split skin grafted.

The desmoid over the tibia and knee was seen as a 2.5 cm lobular mass over the upper medial aspect of left leg. This was excised

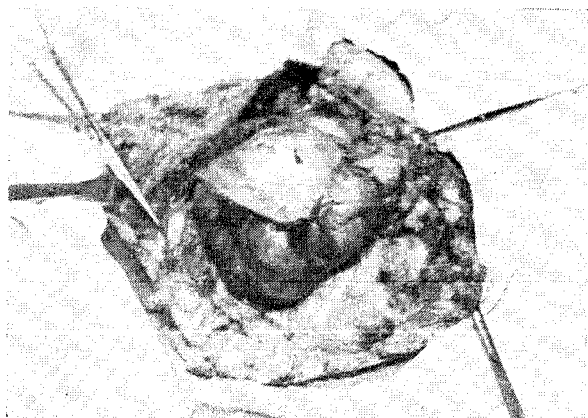


Fig. 1A. An abdominal desmoid in the epigastrium. Note the encapsulated nature and the wide area of skin fixity. 16 x 14 cm skin was excised.

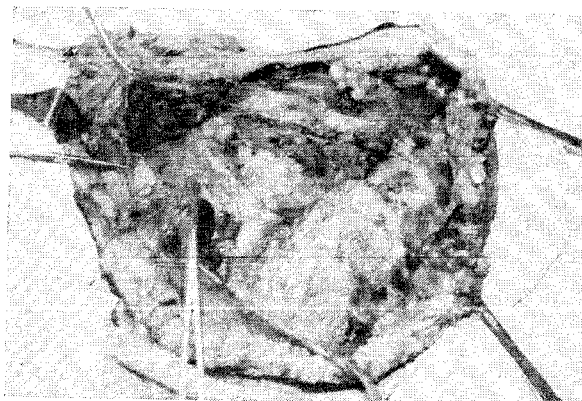


Fig. 1B. The cut margins of the rectus assembly and anterolateral muscles of abdomen in view-held up by haemostats, showing the defect.

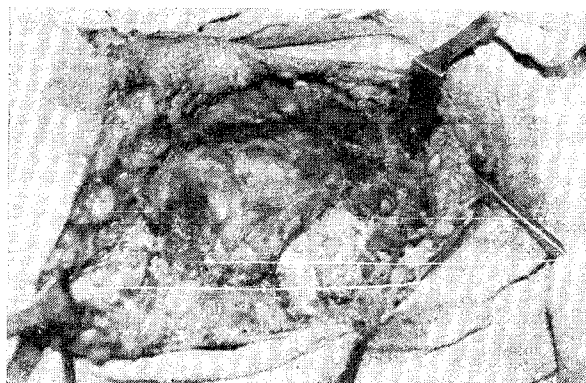


Fig. 1C. The defect in the musculoaponeurotic plane bridged by a prolene mesh.

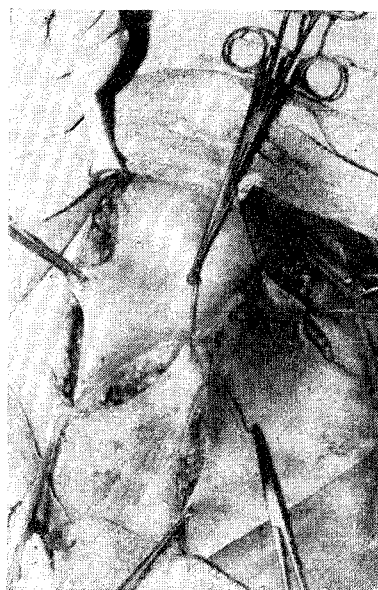


Fig. 1D. Local skin flaps fashioned and planning of closure done by stay sutures.

and skin grafting of the defect was done. This tumour has recurred 4 times since and when last seen had assumed a 24 × 18 cm large lobular appearance over the knee and lower thigh and had ulcerated as well (Fig. 5A). This too was excised with wide tumour free margins and the defect was again split skin grafted (Fig. 5B). This patient had recurred 4 times in 5 years and the longest tumour free occasion was that of 15 months after the first surgery.

We have encountered no systemic spread in any of our patients and by a monthly follow up for the first year after every surgery and two-monthly follow up thereafter we try to

pick up local recurrences at the earliest and treat them effectively.

**Discussion**

Desmoid tumours (or aggressive fibromatosis or musculo-aponeurotic fibromatosis) is a unique tumour of the musculo aponeurotic supporting tissues which is unencapsulated, non-metastasizing but infiltrating extensively into the surrounding local tissues (Mac Kenzie, 1970; Benninghoff and Robbins, 1964). A great majority of these are "Abdominal desmoid tumours" being present in the anterior abdominal wall (Brasfield and Das Gupta, 1969, Kirchmer and Woma, 1977) but extra abdomi-

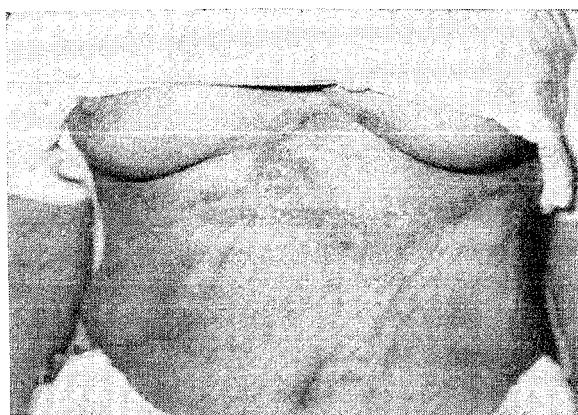


Fig. 1E. The abdominal closure after 3 months.

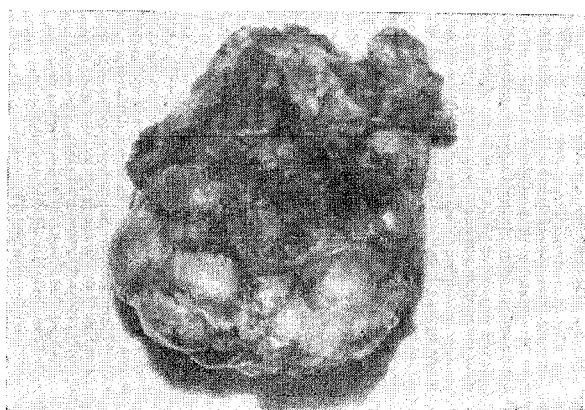


Fig. 1 F(a)

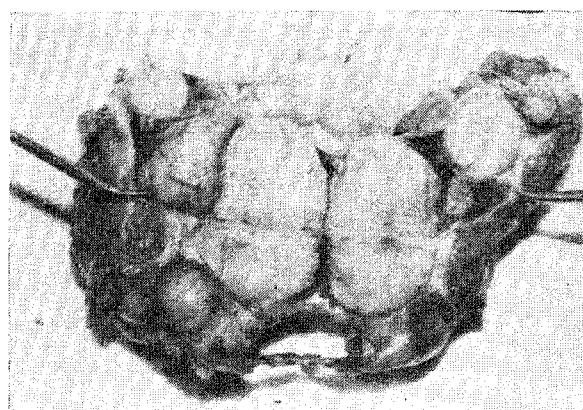


Fig. 1 F(b)

Fig. 1F. The gross tumour and its cut section.

nal desmoids, originating in the extremities, and in relation to virtually every other muscle present in the body are also not uncommon (Das Gupta et al., 1969; Musgrove, 1948). The tumour is seen between 30-50 years of age

and about 70% of the patients are women. Actiologically they are either due to fibrous overgrowth to trauma or because of some endocrinal disturbances. The frequency with which the abdominal desmoids occur following



Fig. 2. Desmoid in the upper arm recurring in the deltoid region after two previous excisions and primary closures.



Fig. 3A

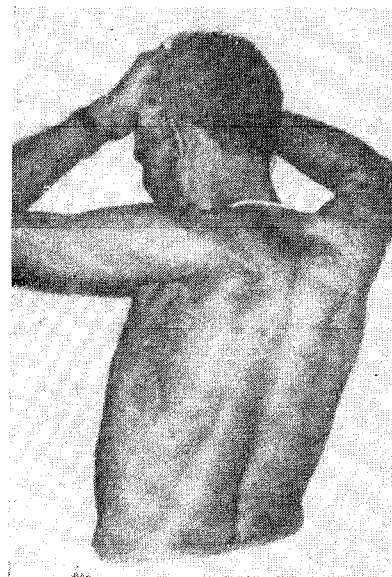


Fig. 3B

Fig. 3A&B. 2½ year tumour free follow up of a lateral chest wall desmoid. Note the primary closure with a small Z-plasty at the lower end.

the violent muscular contractions of parturition and its frequent origin on a healing scar testifies the former theory of origin while the high concentration of estrogenic and pituitary gonado-trophic hormones in the bioassay of the tumour tissue suggests the later theory.

The main aim of reconstruction of the abdominal wall following the ablative surgery for an abdominal desmoid is to reestablish the inte-

grity and support of the fascial components and to provide a skin coverage. To meet this goal musculofascial flaps (Wangensteen, 1934, Lesnick and Davids, 1953, Hershey and Butcher, 1964, Gerber, 1965, Ye, Devine and Kirklin, 1953), fascial grafts (Mc Peak and Miller, 1960) and prosthetic supports like metal and plastic have been used from time to time.

Extra abdominal desmoids, however, do not



Fig. 4A. An upper arm desmoid was excised and primarily closed 8 months back. Now she has a recurrence in the posterior axillary fold.

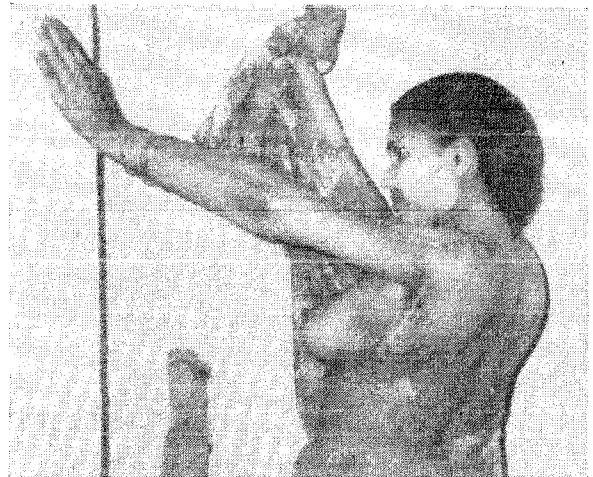


Fig. 4B. The recurrence has been excised and this defect too was primarily closed.

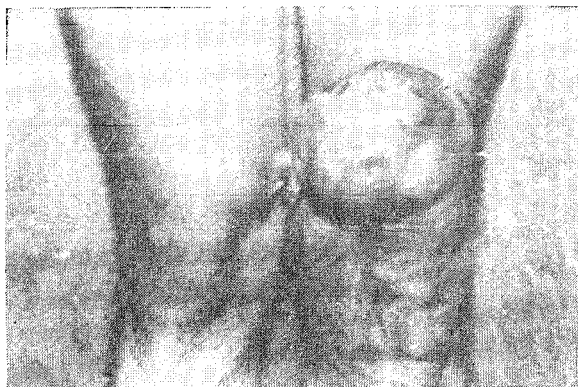


Fig. 5A. Desmoid over the left knee-which had ulcerated. Note the area of split skin grafting done previously over the skin and upper medial thigh.



Fig. 5B. The same patient seen postoperatively.

present a stereo-typed problem. If the raw area left behind does not expose any vital structure and is capable of accepting a split skin graft, this is the choicest method of reconstruction. Post-operative splintage and physiotherapy should be kept in mind whenever grafting is attempted across a joint. On other occasions, however, a fasciocutaneous flap or a myocutaneous flap can be used for one stage reconstruction.

Wangansteen (1934) used a tensor fascia lata myocutaneous unit including the iliotibial tract for abdominal wall defects but realised that this flap was too short for upper abdominal defects and used a musculofascial flap of external oblique aponeurosis and anterior rectus sheath in conjunction with the tensor fascia lata for larger defects. Hershey and Butcher (1964) used an external oblique myocutaneous unit for closure of upper abdominal and lower chest defects borrowing the inframammary skin from chest whenever required. Lesnick and Davids (1953) established the integrity of the anterior abdominal wall by using the musculofascial flaps of external oblique from adjoining area by dividing it laterally and reflecting it medially hinging the flap at the anterior rectus sheath. Such fascial reflections using the anterior rectus sheath as a flap and linea alba as the hinge was also used by Gerber (1961). Open jump flaps carried on wrist carrier have been used by Ye et al. (1953) and Morgan and Zbylski (1972) but as this is a multistaged procedure it is best avoided.

The ready availability of fascia lata, its favourable tensile strength and no foreign body reaction prompted Mc Peak and Miller (1960) in utilizing it to repair the abdominal wall following tumour resection. It presented a smooth and glistening surface, and this, according to them, was responsible for reducing the chances of adhesions and intestinal obstruction.

Furthermore in a case of fibrous or fascial tumour like desmoid the fascia lata was also preferred to a prosthetic mesh because the tumour could grow along the lattice work of the prosthesis making it difficult to differentiate a recurrence from a fibrous scar reaction surrounding the prosthesis.

Metallic prosthesis for the repair of abdominal defects are mentioned only to be condemned because of high infection rate, "work fracture" and fragmentation. Thus silver wire and tantalum mesh fell out of favour (Pearce and Entine 1952; Mc Peak and Miller, 1960). Plastic prosthesis like Nylon, Orlon, Teflon, Dacron, Marlex and Mersilene are now gaining popularity. Marlex mesh shows little foreign body reaction, provides adequate support and has high resistance to infection (Schmitt and Grinnon, 1967). Mersilene net, a Dacron mesh was used by Wilson and Rayner (1974) and is said to become firmly infiltrated with fibrous tissue, remain flexible and is minimally degenerative.

Surgery is not the only modality of therapy described for desmoids. Benninghoff and Robbins (1964) have discussed the role of radiotherapy particularly in cases of incomplete surgery and in recurrences. Radiation alone or radiation and surgery were also tried by Greenberg et al. (1981), Wara et al. (1977) and Suit and Russel (1975). Radiation was advised either immediately after surgery or initiated at the first sign of regrowth.

Hormonal therapy too is known to produce long term remissions (Kinzbrunner et al., 1983) have tried tamoxifen 20 mg four times a day, and Lanari (1983) has used progesterone and have claimed good results. Waddell and Grener (1980) used indomethacin and ascorbic acid to promote similar regression. We have, however, used only surgery as the modality of treatment in our patients.

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