

DESMOID TUMORS PARTICULARLY AS RELATED TO THEIR SURGICAL REMOVAL*

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DESMOID TUMORS ARE hard, fibrous, infiltrating tumors, usually found in the anterior abdominal wall and arise from the fibroblast of the deep fascia and aponeurotic structures. They do not metastasize; consequently, their complete removal results in cure. While such tumors have been described as occurring in various parts of the body wherever skeletal muscles are found, the fascia connected with the recti and internal and external oblique muscles are by far the most frequent areas involved.

Such tumors may occur in any age group, but by some observers, particularly Pack,¹ are thought to be related to pregnancy, because in his experience they are more commonly found in females in the child bearing period. They give few, if any, symptoms; therefore, their presence is usually discovered on routine physical examination or accidentally by the patient. They grow slowly, infiltrate and destroy the muscle fibers and tend to follow the direction of least resistance; hence, follow the course of muscle bundles and fascia planes. While beginning unilaterally, they slowly infiltrate the abdominal wall and may ultimately invade this structure widely, making their removal a major or impossible task. They have been found adherent to the periosteum of the pelvic bones, attached to Poupart's ligament, and in two of the cases being reported, the tumor had infiltrated the entire thickness of the abdominal

wall and had become intimately adherent to the sigmoid colon and omentum. Whether or not visceral organs may be invaded by these tumors seems to be a point as yet undetermined. In one of our cases, the attached area of sigmoid was resected with the tumor, but actual invasion of the gut wall could not be demonstrated on microscopic examination. On physical examination, the tumor is usually recognized as lying deeply in the abdominal wall, coming more into prominence when the head is raised and the body flexed in contradistinction to tumors lying within the abdominal cavity.

The histological picture of these tumors resembles that of fibromata (Fig. 1) and they have been confused with neurofibromata, but by the use of special silver stains, differentiation between the two is easily made. It has been the experience of many observers that sarcomatous degeneration of these tumors may occur and under such circumstances, their microscopic picture changes accordingly. This is said to occur more frequently in males than in females. The skin and subcutaneous tissue is rarely involved unless malignant changes take place.

The treatment of desmoid tumors is preferably by complete excision. However, under certain circumstances, namely, in poor operative risks, in individuals where the tumor has invaded the abdominal wall so extensively that removal is impossible, and in individuals refusing surgery, radium

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or roentgen ray therapy has been found of value. From the histological picture, one would not expect such tumors to respond to this type of therapy. Pack and others have suggested, since response to irradiation has occurred most noticeably in females, that

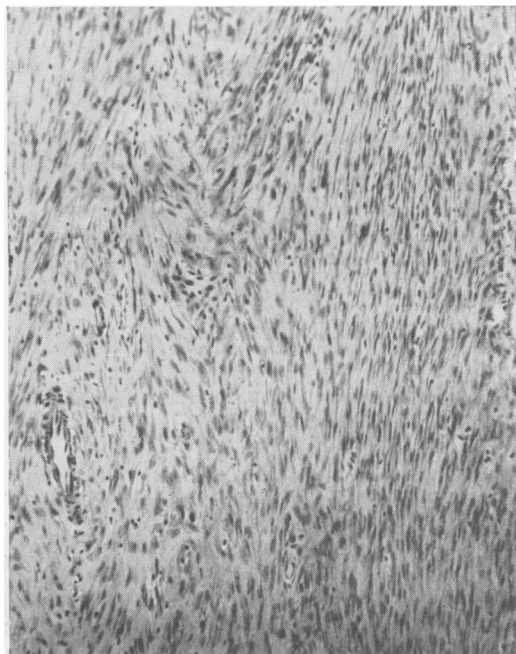


FIG. 1. Typical microscopic picture of desmoid tumors, Case 2.

improvement may result from diminution in ovarian function.

It has been the experience of everyone that desmoid tumors occur rarely. Pack reported that between 1917 and 1943, 17 such tumors were seen at the Memorial Hospital in New York, during which time 50,346 cases of neoplastic disease were admitted to the hospital for the first time. Fifteen were in females, and all but one was associated with recent gestation.

At this Clinic, we have seen four cases during the past 26 years, each of which has been confirmed by microscopic examination of removed tissue.

CASE REPORTS

The first case occurred in a male of 62 and was successfully operated upon by Dr. George F. Straub.² The second case, a Chinese female, was first seen on November 3, 1938, at the age of seven, with an extensive tumor of the abdominal wall (Fig. 2A). The tumor had been first noted as a small nodule arising in the suprapubic area when the child was one year of age. On November 18, 1938, an attempt was made to remove the tumor surgically, but it had invaded the abdominal wall so extensively that this could not be accomplished. A large amount of the tumor was resected followed by irradiation, but little of value was accomplished. On July 30, 1943, resection of a portion of the tumor was again done, followed by irradiation with the hope of palliation. By November 24, 1943, the tumor had enlarged considerably (Fig. 2B), and at that time, it was our belief that probably the lesion was undergoing sarcomatous degeneration and that life expectancy was correspondingly short. The tumor became ulcerated and the patient was confined to bed for a period of six months. The patient was lost sight of until recently, when much to our surprise, it was learned that she was not only living, but was well, had been at work for the past five years, and was contemplating marriage. Figure 2C is a photograph of the abdomen taken on September 11, 1953. The physician, under whose care she has been for the past ten years, stated that nothing specific had been used—no roentgen ray or radium treatment in particular. The patient's father ascribes the improvement in part to Chinese herbs, the nature of which has not been determined. Examination of the patient's abdomen reveals areas of thickening and induration of the abdominal wall, in all probability due to residual areas of tumor, though there has been a marked regression in the tumor since 1943. The patient states that the tumor is slowly becoming smaller; there is no ulceration. Since clinical improvement began about the time of onset of menstruation, it would seem logical to conclude that in all probability, ovarian function played a part in the patient's marked improvement. It is interesting to speculate what effect pregnancy might have on this tumor, and also the ultimate cessation of ovarian function.

The third case was a Japanese male, age 29, who first came to the Straub Clinic on July 14, 1952, because of a tumor in the left lower quadrant. On examination, there was a firm, somewhat tender mass in the left lower quadrant of the abdomen that seemed to be a part of the abdominal wall, indefinite in outline but roughly about 6 cm. in diameter. The patient had been aware of this tumor for about one month.

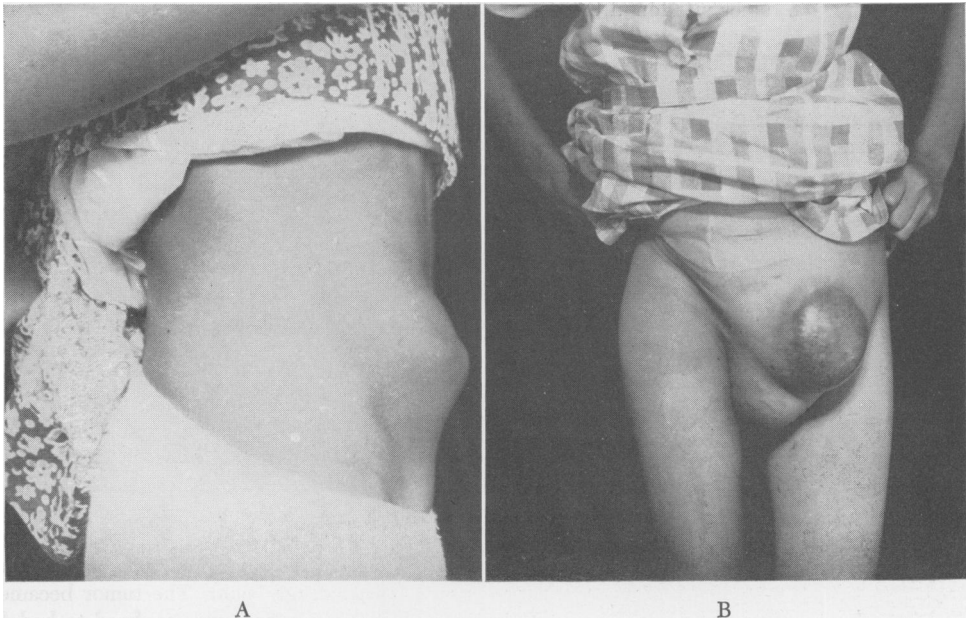


FIG. 2 (A). Photograph of abdominal wall, Case 2, in 1938, and (B) in 1943 when tumor was thought to have undergone sarcomatous degeneration.



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FIG. 2

(C) September, 1953, 21 years after onset, showing marked regression which began ten years previously at onset of menstruation. No ulceration is present.

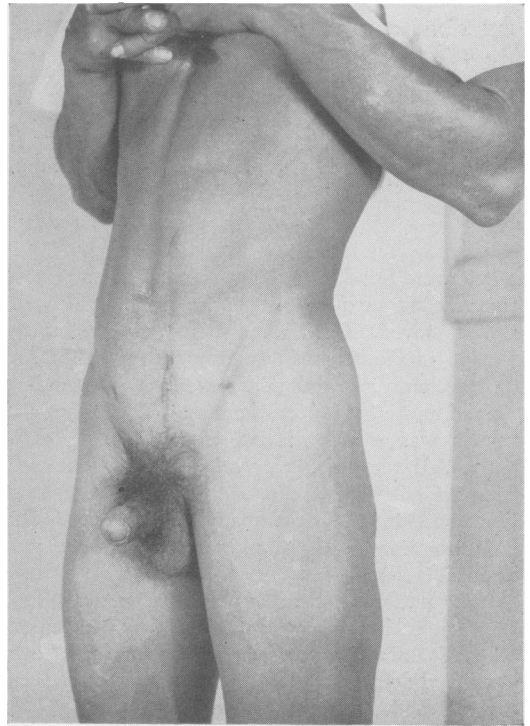


FIG. 3. Photograph, Case 3, showing abdominal wall one year after removal of tumor and its reconstruction by fascia from left thigh.

On exploration, the tumor was found to occupy the entire thickness of the abdominal wall, replacing a portion of the left rectus muscle and the oblique abdominal muscles. The gross appearance and frozen microscopic sections showed the typical picture of a desmoid tumor. Posteriorly, the sigmoid colon and the omentum were adherent to the involved parietal peritoneum. These attach-

ments had noticed heaviness and some slight discomfort on coughing or sneezing, in the left side of the abdomen, and on palpating had felt the tumor. Examination at that time showed a diffuse, non-tender mass in the left side of her abdomen, as shown in the accompanying diagram (Fig. 4). She was the mother of 2 children, her last delivery being on December 19, 1952.

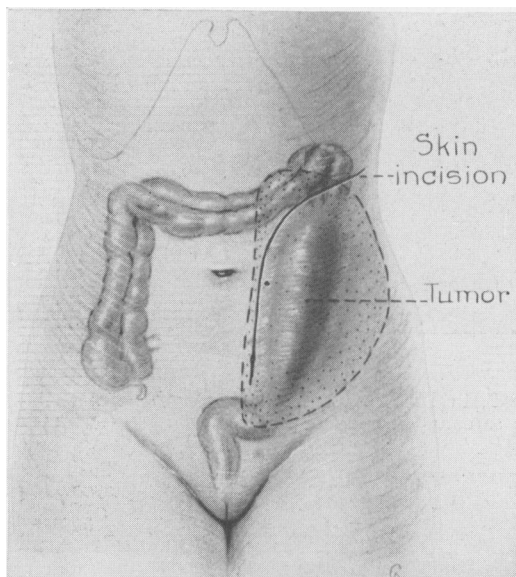


FIG. 4. (Case 4) Diagrammatic drawing to show tumor and extent of removal of abdominal wall.

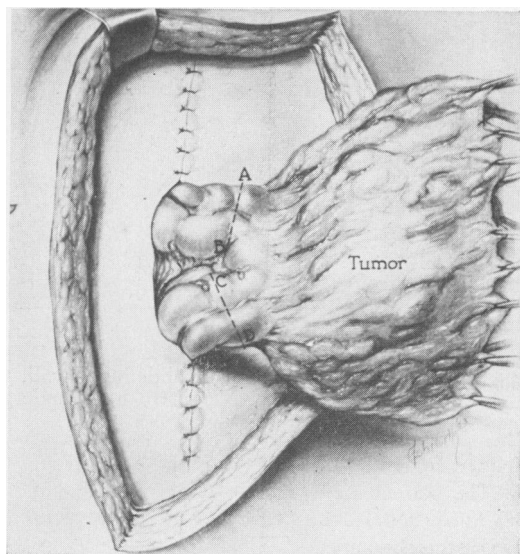


FIG. 5. (Case 4) Showing peritoneum closed. Tumor attached to sigmoid colon and line a, b, c and d where colon was resected.

ments were freed, and the tumor, including a wide surrounding area of the entire abdominal wall, was removed. This left a defect approximately 14 cm. in diameter. The surrounding peritoneum on the anterior abdominal wall and extending around posteriorly over the ureter and spermatic vessels was mobilized and closed without difficulty. A large piece of *fascia lata* was obtained from the left thigh and used to cover the defect as illustrated in the accompanying drawings. The area anterior and posterior to the fascia was drained.

The patient made an uneventful recovery and returned to light duty one month following operation. Examination one year postoperatively shows a solid abdominal wall (Fig. 3), and the patient has returned to his previous occupation.

The fourth case, a Japanese female, age 41, was first seen on June 8, 1953, with a diagnosis after biopsy of a Desmoid tumor occupying the left abdominal wall. About 3 months previously,

At operation on June 12, 1953, almost the entire left half of the abdominal wall was removed. The lesion was found intimately adherent to the sigmoid colon (Fig. 5), and it was thought advisable that this area of the gut be resected. The peritoneum was widely mobilized, both along the anterior abdominal wall and posteriorly, the ovarian vessels and the ureter on the left being isolated and protected during this maneuver. It was possible to close the peritoneum up to the region of the attached sigmoid. The sigmoid was resected and an end-to-end open type anastomosis was carried out, the area being soaked with a solution of neomycin, penicillin and streptomycin, being left in the abdominal cavity. After the anastomosis was completed, the gut was dropped back and the remainder of the peritoneum closed. Two large strips of *fascia lata*, practically all the fascia on the lateral aspects of both thighs, were removed and used to replace the abdominal defect, as shown in the accompanying drawings (Fig. 6).

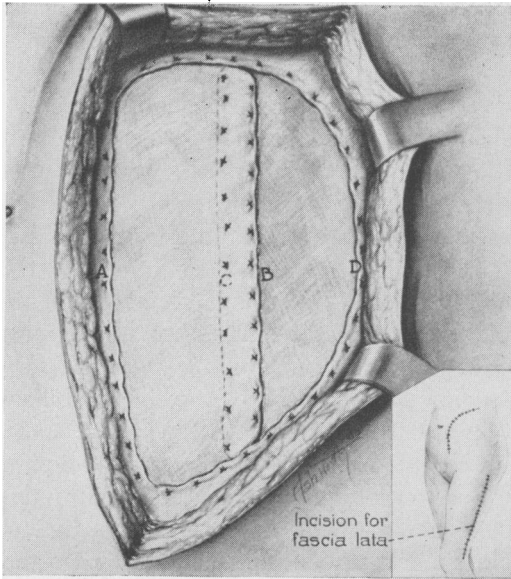


FIG. 6. (Case 4) Showing method of repair of defect in abdominal wall by use of *fascia lata* obtained from left thigh.

The space anterior and posterior to the fascia was drained for 48 hours.

The patient made an uneventful recovery and left the hospital at the end of 10 days. Examination of the abdominal wall one month later showed it to be solid, with no evidence anywhere of weakness (Fig. 7).

DISCUSSION

Desmoid tumors occur rarely; consequently, no one has the opportunity of dealing with the problems of their surgical removal often, and rarely when there is extensive involvement of the abdominal wall necessitating its wide removal. Since complete removal of the tumor is tantamount to a successful outcome (with the apparent exception of the case reported), the surgeon must not be influenced to be conservative, fearful that he will be unable to successfully deal with the defect which he has created. The repair of the abdominal wall has been so successfully accomplished in the two cases in which wide removal was necessary that a description of the method used seems worth recording.

It should be remembered that the removal of skin and subcutaneous tissue is



FIG. 7. Photograph of patient (Case 4) one month postoperative.

rarely necessary because these tissues are rarely involved, unless the lesion is advanced; that the abdominal muscles and peritoneum are the structures to be removed and that this must be carried out well beyond the gross and microscopic limits of the tumor if recurrence is to be prevented. When small, the resulting defect in the abdominal wall may be adequately closed by mobilizing surrounding structures, and this is particularly true of the peritoneum. Large defects may necessitate resorting to the use of dermal grafts, tantalum, or stainless steel mesh, or the method that seems to me preferable, the use of adequate amounts of *fascia lata*.

Whether or not underlying visceral organs attached to the tumor should be resected is, so far as I have been able to determine, a moot point, but judging from

behavior of other neoplasms under similar circumstances, it would seem advisable. When it seems likely that it may be necessary to resect underlying colon, preoperative preparation of this organ with antibiotics is desirable. In the case in which the sigmoid was resected, without such preoperative preparation, the use of the antibiotics as described resulted in primary healing of the wound. However, every precaution against infection should be used because free transplants of fascia, from experience in the preantibiotic era, are known to become infected easily, and such an occurrence under circumstances being described might well be disastrous. It is probably wise to drain the large spaces, both posterior and anterior to the fascia transplants, for 24 to 48 hours in order to prevent collections of blood and serum that invite infection. Further observation in the relationship of desmoid tumors to ovarian function and possibly other hormones may lead to a better understanding of their genesis and methods of therapy.

In conclusion, the author wishes to emphasize the following points: Desmoid tumors are slow growing and do not metas-

tasize, but they may ultimately undergo sarcomatous degeneration. They are curable by complete removal, but continue to grow when operation is incomplete. They should be removed early so as to prevent the necessity of sacrificing a large part of the abdominal wall. However, even though as much as half of the anterior abdominal wall must be removed to effect a cure, the method described, using *fascia lata* to replace this structure, has been found most effective. The surrounding *parietal peritoneum* lends itself to extensive mobilization and for this reason large defects can be completely peritonealized. Attention is called to the regression of a desmoid tumor concomitant with onset of menstruation. No similar case has been found reported in the literature and the significance of this observation has not been determined.

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