Radical Treatment of Recurrent Cystosarcoma Phylloides

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Malignant cystosarcoma phylloides is a rare breast cancer which frequently recurs locally before metastasizing. Local recurrence of cystosarcoma phylloides is unlike adenocarcinoma of the breast since it does not necessarily indicate systemic metastases; and, therefore, aggressive surgery can cure a number of patients with locally recurrent tumor. The present report is an extreme example of a patient with locally recurrent malignant cystosarcoma phylloides, and it illustrates the value of radical surgery in controlling this disease which had previously recurred 21 times. Fifteen months following sternal resection, the patient enjoys her longest tumor-free interval in 16 years.

CYSTOSARCOMA PHYLLOIDES is a rare tumor comprising 0.3% to 0.9% of all breast tumors. The majority (70%) of these appear benign on histological examination. However, a significant number of "benign" tumors will recur and some have been reported to metastasize.[11,21] Even more confusing is the frequent propensity for the malignant variant of cystosarcoma phylloides to repeatedly recur locally before metastasizing. When the surgeon encounters such local recurrence, he often performs a conservative operation because of presumed coexistent metastatic disease. The result of such therapy is further local recurrences in addition to distant spread. The purpose of this report is to describe a radical surgical procedure to locally control a malignant cystosarcoma phylloides which had recurred 21 times.

Case Report

In 1958, a 50-year-old white female (NIH #09-51-49-3) underwent biopsy of a lump in the left breast which was reported to be cystosarcoma phylloides. Approximately one year later, the mass recurred and she underwent a simple mastectomy. The pathologic report was again cystosarcoma phylloides. She subsequently had 19 more recurrences in the anterior chest wall; each treated with local excisions. By 1970, the pathologic diagnosis was reported as malignant cystosarcoma phylloides. At that time, the patient received 2000 R to the anterior chest wall, following which the lesions recurred at an even faster rate with three recurrences developing in the 12 months prior to our examination.

In December 1972, she was referred to the Surgery Branch of the National Cancer Institute. Examination revealed a 4 × 5 cm mass on the anterior chest wall overlying the sternum at the level of the fourth rib, and this non-tender mass was fixed to the underlying tissue at the medial margin of the mastectomy scar (Fig. 1). There were no palpable axillary, supraclavicular or cervical lymph nodes, and the remainder of the physical examination was negative. Workup for metastatic disease (including chest tomograms, IVP, upper GI series, barium enema, and bone, brain, liver and gallium scans) was negative.

On the 21st hospital day, the patient underwent sternectomy from the level of the third rib to the xyphoid process, partial resection of ribs 4, 5, 6 and 7 on the left; excision of the previous mastectomy scar; and left axillary and bilateral internal mammary lymph node dissection. The sternal defect was closed with lyophilized fascia lata homograft, which was covered with a skin flap from the opposite breast. The remainder of the exposed chest wall was covered with split thickness skin graft. Postoperatively, she did well except for retraction of the breast flap, leaving a portion of the fascial homograft exposed. This was treated with saline soaks and procine heterograft coverage to stimulate granulation. Finally, the area was covered successfully with autogenous skin grafts. Pathologic diagnosis of the specimen, as well as all 20 previous recurrences, was malignant cystosarcoma phylloides. The tumor penetrated between the ribs, but did not invade the bone or pleura. Axillary and internal mammary lymph nodes were negative for tumor. The patient was discharged in good condition and at this time, 15 months following surgery, enjoys the longest recurrence-free interval she has experienced in the past 15 years (Fig. 2).

Discussion

Histologically, cystosarcoma phylloides is characterized by proliferation of both the stromal connective tissue and epthelial components. The malignant variety exhibits marked hypercellularity of the connective tissue stroma, with cellular atypia and frequent mitoses.[12] All of these features were present in the case reported (Fig. 3).

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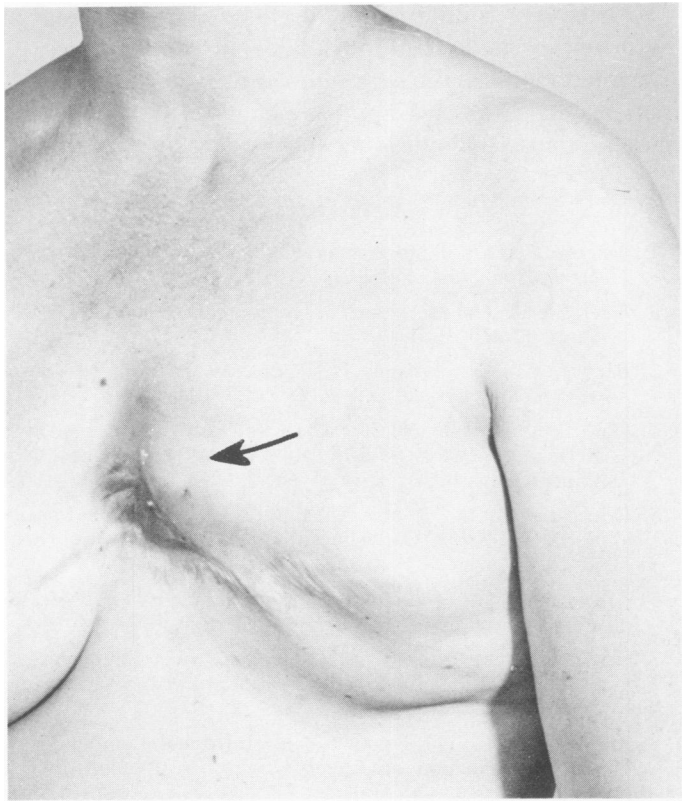

FIG. 1. Preoperative photograph of the patient showing the large 4 × 5 cm recurrence on the left anterior chest wall (arrow).

The clinical behavior of malignant cystosarcoma phylloides is well documented.[1,6,9,14,16] Metastases have been reported to occur in as few as 6.6%[20] of cases to as high as 70%.[11] The overall metastatic rate is approximately 25%. Most metastases are blood-borne but spread to axillary lymph nodes has been reported to comprise about 10% of all metastases.[10,13,15,19] Occasionally, direct invasion of the chest wall and lungs occurs.[17] Local recurrence is the single factor most associated with metastases. Kessinger[7] in her review of 59 patients with metastatic cystosarcoma phylloides found that 26 of these patients (44%) had a minimum of one local recurrence prior to metastasizing. However, local recurrence of cystosarcoma phylloides does not always indicate systemic metastases. Norris and Taylor reviewed 20 cases of malignant cystosarcoma phylloides and noted that while eight of these patients (40%) recurred locally, only five of the 20 (25%) metastasized.

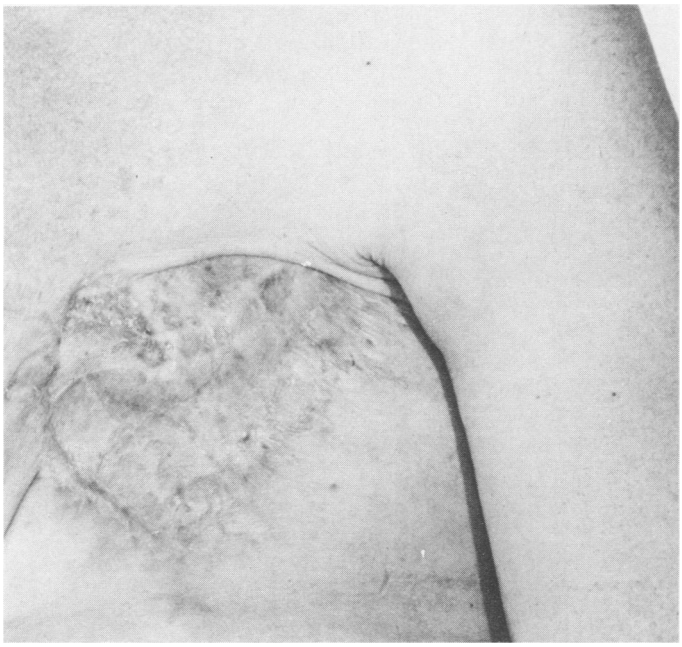

FIG. 2. Photograph of the patient nine months following surgery demonstrating the well-healed operative site.

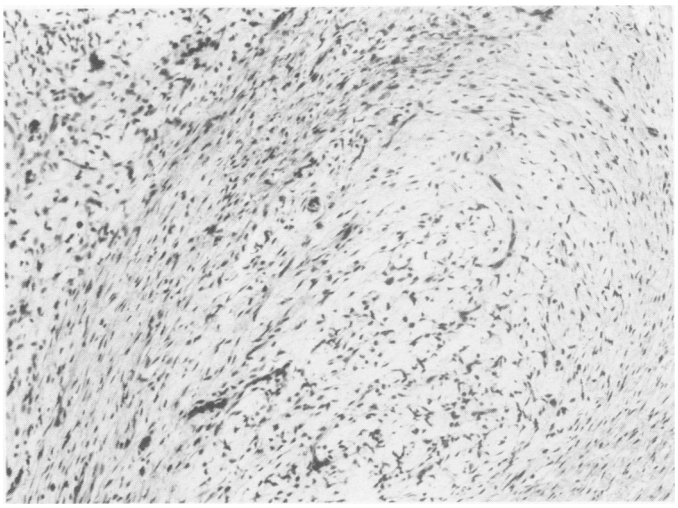

FIG. 3a. Photomicrograph of the original tumor removed in 1958. Note the marked hypercellularity of the connective tissue stroma and cellular atypia. (H & E ×140).

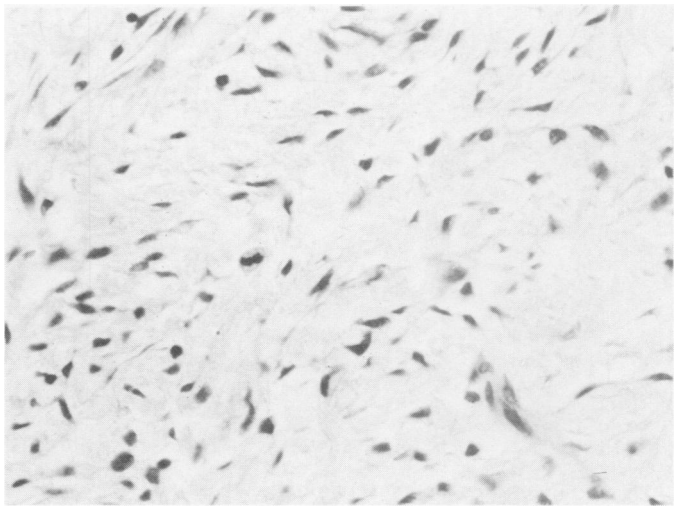

FIG. 3b. High power photomicrograph of the primary tumor showing bizarre cells with frequent mitoses characteristic of the malignant form of cystosarcoma phylloides. (H & E ×560).

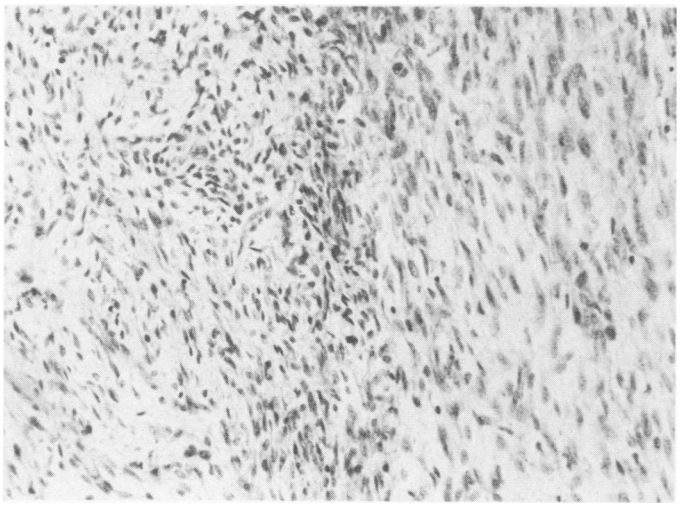

FIG. 3c. Low power photomicrograph of the 21st recurrence which demonstrates the proliferation of epithelial and stromal elements and cellular atypia previously seen in the primary tumor. (H & E 225×).

It appears, therefore, that control of the primary tumor is of paramount importance in successfully treating malignant cystosarcoma phylloides. Most authors[2,3,17,19] have recommended a minimum of simple mastectomy as the initial procedure for this disease. However, in marked contrast to adenocarcinoma of the breast, where local recurrence is metastatic spread,[4,5,8,18] recurrence in cystosarcoma phylloides is not necessarily a manifestation of systemic disease. Therefore, the patient presenting with local recurrence deserves aggressive surgery to control the local disease.

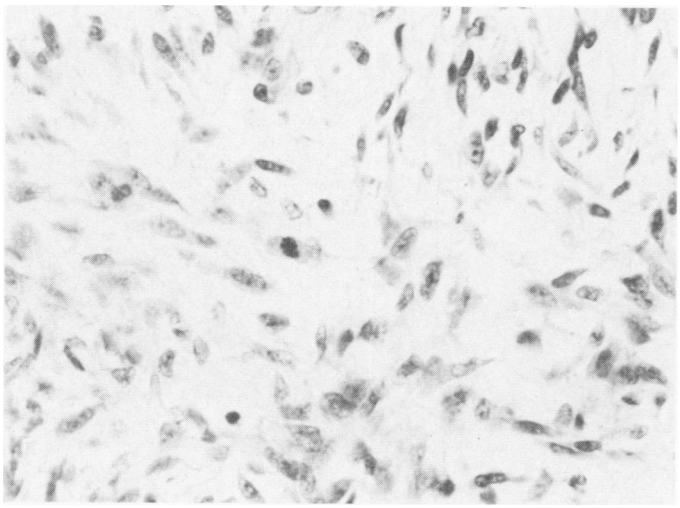

FIG. 3d. High power photomicrograph of the 21st recurrence. Immature nuclei and frequent mitoses are prominent features of this malignant tumor. (H & E 560×).

Our patient is an example of effective management of recurrent cystosarcoma phylloides by radical surgery. Despite the unusually high number of recurrences in this patient, an aggressive approach to recurrent malignant cystosarcoma phylloides was successful.

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