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Case Report

Malignant Fibroxanthoma (Malignant Histiocytoma): an Ill-Defined Histologic Entity with Unpredictable Biologic Behavior

Report of a Case with Long-Term Survival

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Summary. The case of a 45-year-old woman with malignant fibroxanthoma (malignant histiocytoma) of the abdominal wall is reported. A local recurrence five years after surgery and radiation therapy required removal. Subsequent surgical intervention for radionecrosis and a fecal fistula failed to reveal residual tumor; the patient is free of tumor twenty-five years after the initial excision. The differential diagnosis of atypical and malignant fibroxanthoma is discussed. Studies of the small number of malignant fibroxanthomas reported in the literature suggest a poor correlation of the histopathologic characteristics of these lesions with their relatively benign biologic behavior.

Recently we encountered the case of a 70-year-old woman who had a supposedly malignant tumor of the abdominal wall removed twenty-five years ago. The original pathologic diagnosis was recorded as pleomorphic sarcoma of unknown type (probably fascial sarcoma). Because the patient is still alive, this history prompted us to review the old sections which were now interpreted as malignant fibroxanthoma (malignant histiocytoma) in accord with the nomenclature recommended by the World Health Organization (Enzinger, Lattes and Torloni, 1969). This is a rare tumor whose nature still remains obscure to many pathologists and whose growth characteristics are still poorly understood. This is due to the paucity of statistical data as well as to a confusing terminology.

Case Report

Clinical Abstract

A 45-year-old woman was admitted to the hospital in November, 1945, for surgical removal of a painless deep-seated nonmovable and very firm mass of the midabdominal wall immediately below the umbilicus. On surgery the hen's egg-sized tumor was firmly adherent to the fascia in the midline. A wide resection of the fascia with the tumor was done. Invasion of the peritoneum was not demonstrated. The histologic picture of H & E-stained sections of the tumor was interpreted as pleomorphic sarcoma (probably fascial sarcoma). The patient received a postoperative course of radiation therapy. Five years later a recurrence of the tumor led to a second admission in November, 1950. Pertinent physical finding was a double fist-size tumor mass which had irregularly invaded the entire abdominal wall. Microscopic examination of H & E-stained biopsy sections revealed the same histologic picture as seen five years previously. A second course of radiation after surgical removal of the recurrent tumor was administered. The patient returned to the hospital in July,

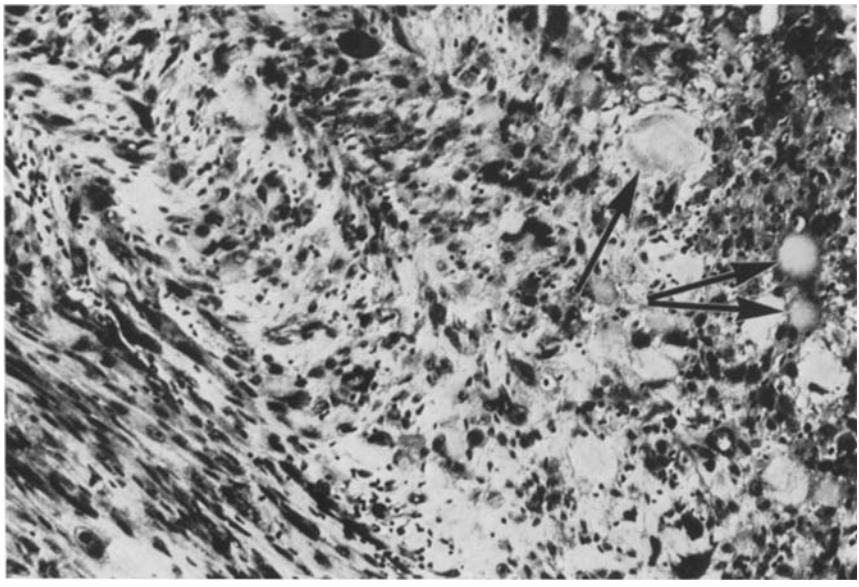

Fig. 1. Malignant fibroxanthoma containing neoplastic histiocytes with abundant cytoplasm (arrows). Hematoxylin and eosin. ×175

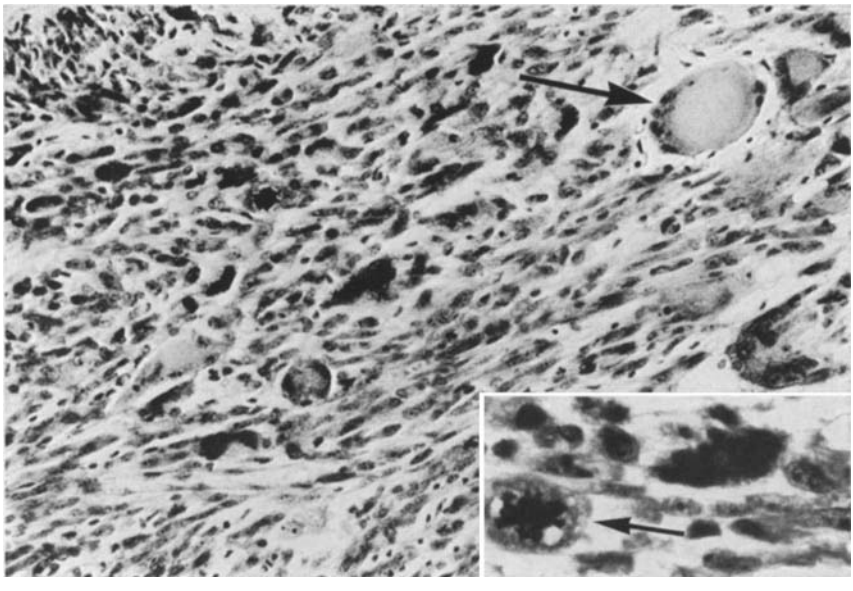

Fig. 2. Note cellular pleomorphism, large neoplastic histiocyte (large arrow) and atypical mitosis (small arrow, inset). Hematoxylin and eosin. ×175. Inset: ×440

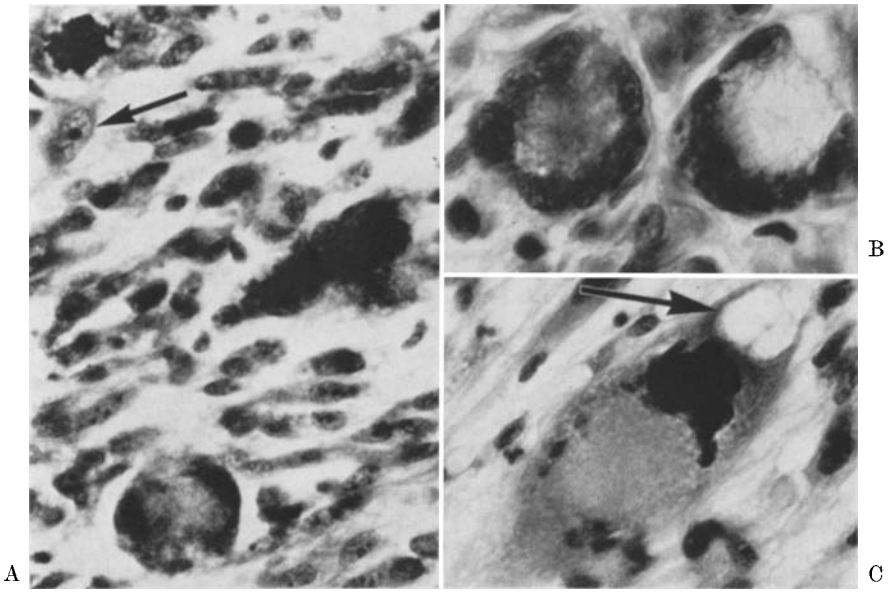

Fig. 3 A—C. Atypical mitosis (A, upper left corner), prominent nucleoli (A, small arrow) and considerable nuclear atypia (A, center, and C) are evident. Touton giant cells are depicted in B and vacuolization is obvious (C, large arrow). Hematoxylin and eosin. A and C: ×440. B: ×700

1951, with radionecrosis of the abdominal wall in association with a fecal fistula of the transverse colon. Several biopsies confirmed the absence of any residual tumor. In February, 1953, portions of the colon and of the small intestine were resected for closure of the fecal fistula and for removal of intestinal adhesions. A plastic reconstruction of the abdominal wall was performed. On her fifth and last admission, February 10, 1970, the patient was cachectic and dehydrated. Several hernias were evident in the abdominal wall. The tentative diagnosis was partial obstruction of the small intestine secondary to adhesions. However, a gastrointestinal series did not reveal the suspected obstruction. The patient is currently being treated by symptomatical measures because surgery does not seem to be indicated.

Microscopic Findings

The tumor displays a high degree of cellular pleomorphism. Even casual inspection at low power has convinced several experienced pathologists that they were dealing with a malignant mesenchymal tumor. Divergent opinions, however, were offered as to the precise classification. The most frequent diagnoses were liposarcoma and rhabdomyosarcoma. This is probably due to the bizarre, hyperchromatic, large and giant histiocytic cells having abundant irregular deeply acidophilic cytoplasm varying from homogeneous to vacuolated appearance. Such cells may resemble neoplastic lipoblasts or cells seen in pleomorphic rhabdomyosarcoma. The lesions contain multinucleated giant cells, predominantly of the Touton type with peripherally arranged nuclei. Prominent acidophilic nucleoli are common. The number of mitoses is variable from several to occasional per high power field. Nuclear atypia is striking.

Discussion

Especially during the past decade these lesions have puzzled surgical pathologists. A number of authors has tried to clarify the problems of differential

diagnosis and nomenclature (see Kempson and McGavran, 1964; O'Brien and Stout, 1964; Stout and Lattes, 1967; Enzinger, Lattes and Torloni, 1969). A detailed review of the problems concerned with a uniform classification of histiocytic tumors is beyond the scope of this article. The reader is referred to the discussions by Stout and Lattes (1967) and to the recent classification as adopted by the World Health Organization (Enzinger, Lattes and Torloni, 1969).

It is now well known that a group of cutaneous lesions histologically similar to the malignant fibroxanthoma does not behave in a malignant fashion in spite of very atypical malignant appearing histological features. These lesions are called atypical fibroxanthomas (see Kempson and McGavran, 1964; Kroe and Pitcock, 1969). Difficulties arise in connection with distinction of these atypical fibroxanthomas from malignant fibroxanthomas. Even experts in the field of soft tissue tumors admit the lack of reliable histopathologic criteria for distinction of the two lesions (Enzinger, 1970) which is of paramount importance for proper therapeutic measures.

According to Kempson and McGavran (1964) and Kroe and Pitcock (1969) Helwig is rightly credited with the introduction of the entity of atypical fibroxanthoma into the pathological literature. The latter designation has also been adopted by the World Health Organization (see Enzinger, Lattes and Torloni, 1969) and is now probably in general usage. Gordon (1964) called these tumors pseudosarcomatous reticulohistiocytomas and already gave a good account of the discrepancy between the malignant histologic characteristics and the benign biologic behavior of these lesions, which, according to the view held by the WHO, may not even be a neoplasm. A similar experience was recorded by Kempson and McGavran (1964) and by Kroe and Pitcock (1969). Careful evaluation of the clinicopathologic features of these atypical fibroxanthomas furnishes criteria which justify the separation of a malignant fibroxanthoma from the above entity. The histological appearance of both lesions may be very similar or almost identical. However, all lesions reported by the foregoing authors were located in the dermis in the older age group. They are characteristically found in the exposed areas of the skin, such as head, neck and upper extremity. In the above cases there is not a single exception to this rule. The lesions are generally small, mostly less than 3 cm in diameter (Enzinger, Lattes and Torloni, 1969). Kempson and McGavran (1964) admit that there are lesions histologically similar to the atypical fibroxanthoma which occur in the soft tissues in younger persons, tend to recur locally and behave in a locally aggressive manner. They consider them malignant fibroxanthomas which have also been reported by O'Brien and Stout (1964). Because pathologists are slow in recognizing this entity its real incidence cannot be estimated. An unknown number of cases is almost certainly misinterpreted because areas of this tumor may resemble rhabdomyosarcoma, liposarcoma or fibrosarcoma. According to Enzinger (1970) Kempson has recently confirmed the difficulty of predicting the clinical course from the histologic presentation of these lesions. Enzinger admits that malignant fibrous xanthomas are not as aggressive as the histologic picture would suggest if one applies the usual criteria of malignancy.

The justifications of placing our case in the malignant category despite the absence of metastasis are the unusual location in the deeper parts of the ab-

dominal wall, the size of the primary and particularly of the recurrent tumor and the locally aggressive behavior. These features set the case apart from the atypical fibroxanthoma. The clinical course underlines the relatively benign behavior of at least some of these lesions. Kobak and Perlow (1949) already adhered to the concept that there may be gradual histologic transitions between xanthomatous giant cell tumors and less well differentiated variants, the most malignant resembling fibrosarcoma. This concept may be valid for histiocytic tumors in general. The fact that some of these tumors fail to metastasize does not disprove their malignant potentialities. It is hoped that additional studies of larger series which are currently in progress at several institutions (Enzinger, 1970) will shed further light on these neoplasms and provide better guidelines for therapy.

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