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CYSTOSARCOMA PHYLLODES*

A malignant cystosarcoma phyllodes in a 14-year-old girl

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Cystosarcoma phyllodes is a rare type of mammary neoplasm. It is a combined fibroepithelial tumor in which the mesenchymal or stromal component is more hypercellular than in the usual fibroadenoma. The most frequent clinical presentation is that of a freely movable, bulky, occasionally painful tumor. The mass is of variable size and generally presents in middle-aged women[1-5].

The majority of tumors behave in a benign manner, but the correlation of clinical behavior with pathologic findings has not been entirely optimal[1,4,5].

Several authors have advocated subdivision of cystosarcomas into the histologically benign and malignant types. Nowadays morphologic criteria are used for the determination of type, prognosis and appropriate therapy[6].

In this article we present the case of a 14-year-old girl with a rapidly growing and metastasizing malignant cystosarcoma phyllodes.

Case Report

A 14-year-old girl was admitted to the hospital with a painless left mammary mass. She had first noticed the mass four months previously when it was approximately 2 cm in diameter.

On clinical examination a firm, ill-defined, uncircumscribed, shapeless, infiltrating lesion, approximately 10 cm in diameter, was found. The diameter had increased from 2 cm to 10 cm in four months indicating that the lesion was indeed growing rapidly. A mammography revealed a large, invasive lesion. During left axillary examination a hard, immobile lymph node was palpated.

Routine laboratory and X-ray examinations were all within normal limits.

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A left simple subcutaneous mastectomy and axillary dissection were performed. During the operation a frozen section was taken; the diagnosis was malignant cystosarcoma phyllodes. The postoperative period was uneventful.

Following the operation the patient was given combined radiotherapy and chemotherapy (adrioblastin + cyclophosphamide + 5-fluorouracil). Two months later lung, bone and liver metastases were detected.

Pathological findings. Macroscopically, the mastectomy, together with the axillary dissection material, measured 10 x 7 x 6 cm. The tumor mass was poorly demarcated and revealed no capsule. It was a solid infiltrating mass, gray-white in color, in texture rubbery and fibrous and in shape spherical with rather smooth external contours.

Grossly metastatic lymph nodes were also determined and dissected from axillary material.

Microscopically, the term cystosarcoma phyllodes is used to describe a general morphological configuration of a fibroadenoma with a rather cellular and hyperplastic stroma.

In our case the epithelial lining of the canaliculi was largely cuboidal and generally resembled the duct epithelium of the breast; there were no malignant epithelial changes (Figure 1). Stromal cells were not uniform, but consisted predominantly

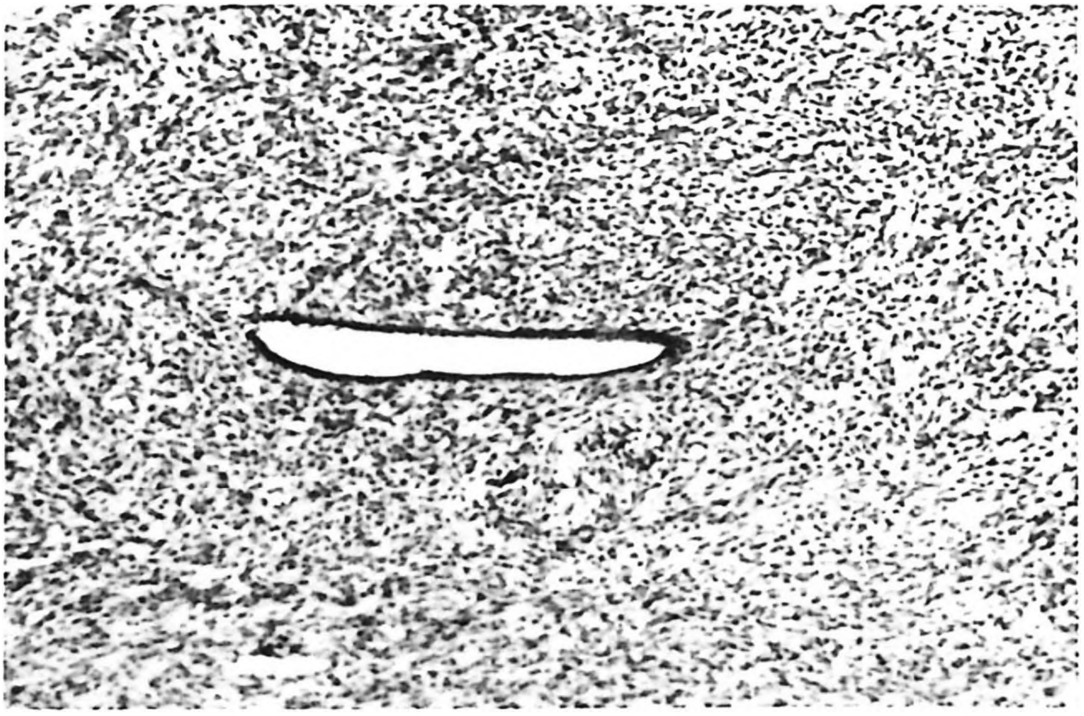

Figure 1

The epithelial lining of the canaliculi resembles the duct epithelium (HE x 75).

of irregular strands and solid patternless masses, similar to those of fibrosarcoma. Since the tumor revealed typical proliferated, connective tissue compressing the epithelial-lined spaces and canaliculi it was regarded, histologically, as malignant (Figure 2).

Figure 2

Sarcomatous stroma and compressed epithelium-lined spaces (HE x 75).

The cells forming the strands had an ovoid, round and fusiform pleomorphic nucleus with coarsely clumped chromatin. Their cytoplasmic borders were not well defined.

There were abundant atypical mitoses (at least 10 mitoses per 10 high-power-field). Less commonly the tumor had an undifferentiated type growth pattern and necrosis and hemorrhage were easily noticed (Figure 3). During the examination of axillary material two metastatic and eight reactive hyperplastic lymph nodes in follicular pattern were observed.

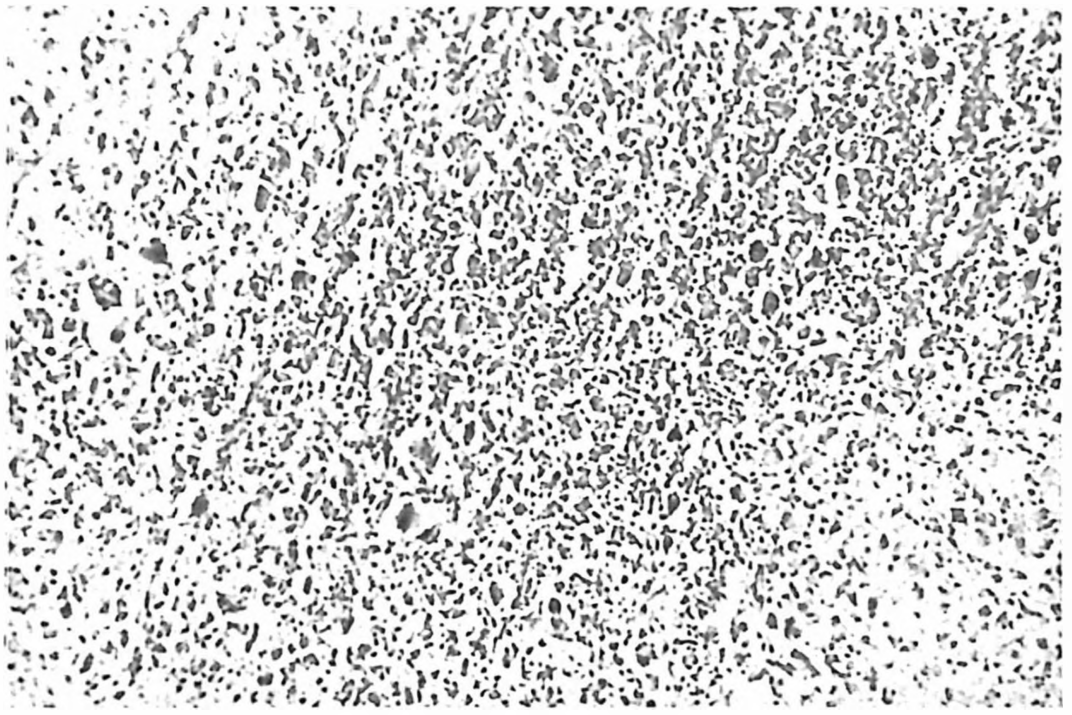

Figure 3

Pleomorphic tumor cells and necrosis (HE x 75).

Discussion

Cystosarcomas are regarded as histologically malignant solely on the basis of the microscopic evaluation of the stromal component. Histological malignancy criteria are similar to those generally used for soft-tissue tumors[7].

In our case, as sometimes happens, there was some significant stromal over-growth at the expense of the ductal component. This produced a focal pattern resembling pure stromal sarcoma of the breast. Only the presence of a few distorted ducts indicated that the underlying lesion was a cystosarcoma[7].

Local recurrences of cystosarcoma have been observed. Hajdu and associates[8] reported tumor recurrences in 18% of 150 women who had benign cystosarcomas and in 8% of 49 patients with malignant cystosarcoma.

Practically all metastases of cystosarcoma result from hematogenous dissemination. The metastases were pure sarcoma in our patient. Metastases of the epithelial component have rarely been reported[9].

Cystosarcomas were not categorized as benign or malignant by Norrin and Taylor[10]. They indicated that no single histologic feature was wholly reliable in predicting clinical behavior and emphasized the low risk of recurrence or of death for patients with small sized tumors (less than 4 cm in diameter), slight mitotic activity (fewer than three mitotic figures per ten high-power-fields), pushing margins and minimal cystologic atypism.

In the evaluation of the pathologic features, tumor size, mitotic-figure-count, stromal atypia and tumor contour are important. Malignant cystosarcoma tend to have a larger than average diameter (median 6.5 cm). In benign tumors 0-4 mitoses per 10 high-power-field, predominantly pushing margins, and slight stromal atypia predominate, whereas in malignant tumors, 10 or more mitoses per 10 high-power-field, predominantly infiltrating margins and prominent stromal atypia are to be seen[6]. Between these figures, there are borderline malignant cystosarcomas[6].

Cystosarcomas are usually seen in middle-aged women, so we thought this case of metastasizing malignant cystosarcoma in a 14-year-old girl was of interest.

Summary

The case of a 14-year-old girl with a malignant cystosarcoma phyllodes is presented. The lesion had grown rapidly and metastasized to the axillary lymph nodes, lung, bone and liver.

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