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Rhabdomyosarcoma of the Breast in an Eight-month old Girl

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Summary

Akenzua GI, Evbuomwan I and Guirguis MN. Rhabdomyosarcoma of the Breast in an Eight-month old Girl. *Nigerian Journal of Paediatrics* **1985; 12:129.** An 8-month old girl with rhabdomyosarcoma of the breast had local recurrence and axillary node metastasis after excision biopsy. Subsequent radical mastectomy was followed by local recurrence and extra-nodal metastasis. A combination of vincristine, cyclophosphamide, methotrexate and prednisone had no significant effect on its rapidly fatal course. Rhabdomyosarcoma occurring as a pure stromal sarcoma may be more dangerous than when it is associated with a fibroadenoma.

Introduction

PRIMARY mammary sarcomas are a rare, heterogenous group of lesions with poorly defined clinicopathologic features and variable prognosis. These tumours may therefore, pose problems in diagnosis and treatment[1]. They constitute less than 1% of all malignant breast neoplasms, with an incidence of one case occurring per year per 1.5 million population as reported in one study[2]. Although they may occur in either sex and at any age usually after puberty, they are infrequent in children and in infancy. Reported cases and published series show a wide range of histologic types in breast sarcomas[3–5]. Rhabdomyosarcoma is infrequently reported although it has been described in association with fibroadenoma[6] and in cystosarcoma phylloides[7]. However, in none of these cases has it occurred in a young child.

Recently, we saw rhabdomyosarcoma of the breast in a girl under the age of one year. The tumour in this case lacked the usual features of rhabdomyosarcoma on light microscopy and it posed significant problems in diagnosis and treatment.

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Case Report

CO was admitted to the University of Benin Teaching Hospital (UBTH) on 20 April, 1982 because of a mass over the left breast. She was aged 8 months and was the first child of a young married couple with no family history of cancer. She had been delivered normally after a 36-week normal gestation, weighing 2kg. There was no history of mastitis in the newborn period.

the primary and metastatic sites. The parents refused further treatment and removed the child from hospital on 18 September, 1982. She died at home on 3 December, 1982 after a one-day history of vomiting. Unfortunately, autopsy could not be performed.

Pathological findings

Grossly, the tumour removed was a lobulated mass 7cm x 6.5cm x 4cm attached to, but not infiltrating the skin. On cut-section, it had no defined capsule; the appearance was that of an infiltrating carcinoma of the mature breast. There were areas of brownish discoloration suggesting haemorrhage.

The microscopic appearance was puzzling; it was initially felt that the appearances favoured the diagnosis of an infiltrating duct carcinoma (medullary type). It was highly vascular and hypercellular (Fig 2). It consisted of malignant cells arranged in large and small masses, sheets and thick long bands which anastomosed to form a crude interrupted network (Fig 3). The cells were separated by strands of vascular connective tissue and were provided with ample eosinophilic cytoplasm and vesicular nuclei with one or more nucleoli. Cellular pleomorphism was marked; mitotic figures conspicuous, amounting to 8 figures per high power field (hpf). Sarcoplasmic cross-striation and typical "racquet" and "strap" cells were not evident. The normal architecture of two axillary lymph nodes was completely replaced by sheets of malignant cells.

Paraffin blocks from the tumour were sent to Birmingham Children's Hospital from where the following report was received:

"The tumour is formed by closely packed cells usually in broad trabeculae or lobules but sometimes, in sheets or narrow trabeculae with surrounding fibrosis. The trabeculae are separated by fibro-vascular septa and often show extensive necrosis away from the septa, sometimes producing central cavitation. The surviving cells often form a radiating cuff around fibro-vascular

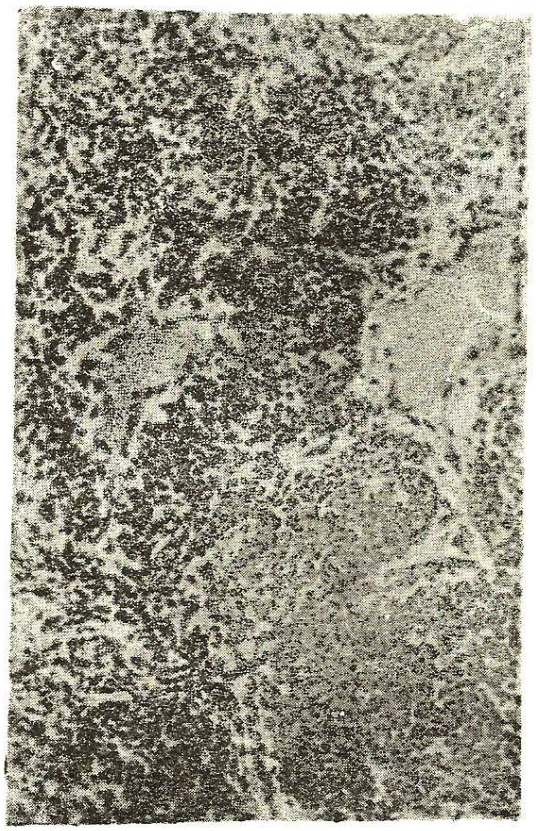

Fig 2. H and E Section of the tumour. Note hypercellularity and vascularity. (magnification x 100).

septa. The cells vary considerably in the amount of cytoplasm but this is usually fairly abundant and eosinophilic, sometimes polygonal but very often cigar shaped or elongated. The nuclei are sometimes large and vesicular and sometimes, smaller and relatively dense. Mitotic figures are frequent, often 2 and sometimes 4 or 5 per hpf. The appearances are those of an embryonic rhabdomyosarcoma with an alveolar pattern.

Ultrastructural findings: The electron microscopic material was prepared from portions of paraffin blocks. All the prints showed that most of the cells contain abundant microfilaments, usually irregularly arranged, forming more or

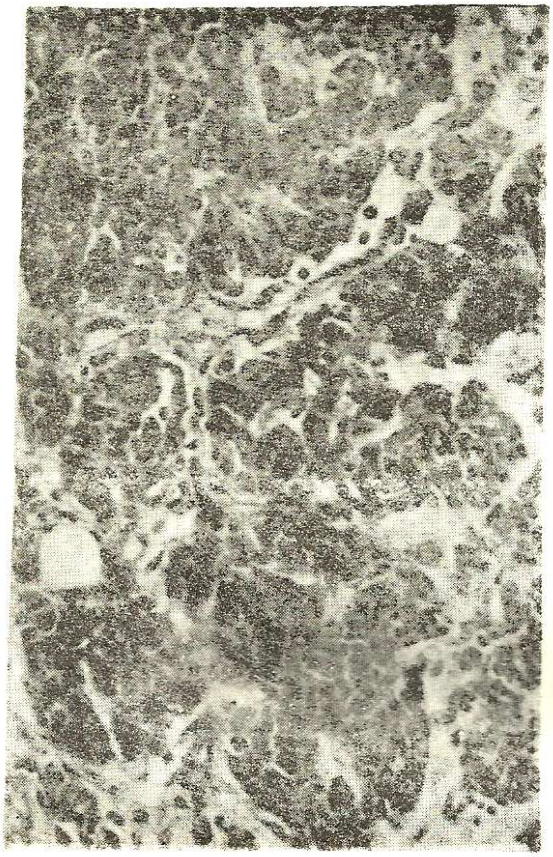

Fig 3. H and E Section of the tumour. Note frequent mitotic figures and cellular pleomorphism. (magnification x 200).

less central mass. The periphery of the cells are occupied by abundant free ribosomes, polyribosomes and rough endoplasmic reticulum. The microfilaments probably less than 10 nm in diameter are probably actin. In several areas, there are broad bands of more or less parallel microfilaments which appear to be thicker, measuring over 10 nm in diameter and probably corresponding to myosin. Some of these show poorly formed condensations which might indicate an attempt to form Z–bands. It is difficult to be sure that some of the broad bands are not extra-cellular but their constituent filaments do not have the wider diameter of collagen or reticulin nor do they have any characteristic banding. Intracellular condensations of the tonofilaments type are not seen. Basement membrane material is absent and there appears to be only a little intercellular collagen in a few places. There are occasional tight junctions between adjacent cells. Pinocytotic vesicles are not seen but preservation of plasma membrane is not good. The appearances are consistent with rhabdomyosarcoma."

Discussion

We believe this to be the first report of rhabdomyosarcoma of the breast in an infant. The case is of interest because of the rarity of rhabdomyosarcoma in this location, and of breast sarcoma at this age. After searching available literature, the youngest patient with rhabdomyosarcoma of the breast we found was the 16–year old girl reported by Woodard *et al*[8], in whom the tumour was associated with a fibroadenoma. Unlike the case described here, that patient was alive 11 years after a simple mastectomy.

It is significant that the tumour in the present case had all the features characteristic of mesenchymal breast neoplams with unfavourable prognosis, including infiltrating margin, cellular atypia and high mitotic count[4 9]. The rapidity of growth of the tumour, its cellular pleomorphism, mitotic activity and focal necrosis were analogous to those described in two of the three rhabdomyosarcomas in the series reported by Oberman[1], as was the infiltration of pectoral muscles. Although two rhabdomyosarcomas in the latter series and all four in the series reported by Botham *et al*[10] lacked sarcoplasmic cross-striations on light microscopy, they had typical 'racquet' and 'strap cells'. By contrast, all three features were lacking in the present case; nevertheless, rhabdomyosarcoma was confirmed by electron microscopy.

It might be argued that rhabdomyosarcoma in this case could have originated from tissues on the thoracic wall other than mammary stroma. But, we think it most pertinent that it presented as a primary mammary tumour. Similarly, others[1] have argued that such distinction is unnecessary. Since we could not find any epithelial elements after meticulous examination of several sections, we think this is a true stromal sarcoma although specimen from initial excision biopsy was not available for examination.

Barnes and Pietruszka[9] have summarised the recent literature on 'stroma sarcomas'. They noted that from the few reported cases, rhabdomyosarcoma of the breast appeared to behave in an ominous fashion. They agreed with Norris *et al*[4] that tumours with the features such as were found in this patient should be considered potential threat to the patients' survival. Hence, faced with a disease with such a grave outlook and whose natural history in infancy is unknown, we chose the combination of drugs described above empirically. However, the result was discouraging and lead us to suspect that rhabdomyosarcoma occurring as a pure stroma sarcoma may pose a greater danger than those associated with fibroadenomas.

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