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Carcinoma of the Breast in Children

A Case Report and Review of the Literature

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SUMMARY

The case of a 10-year-old girl with juvenile carcinoma of the breast is presented. The literature is reviewed and current thoughts relating to the treatment of breast carcinoma in children are discussed.

The major controversy seems to revolve around the choice between radical and local excision in cases of juvenile or secretory breast cancer.

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A review of the literature attests to the rarity of mammary carcinoma in children below the age of 14 years. According to Cholnoky[1] the earliest report of breast cancer in a child was published in 1851, and in 1913 Bryan[2] reported a case of carcinoma of the breast in a boy aged 15 years. In 1917 Levings[3] published the case report of a 12-year-old child who presented with carcinoma of the breast, and 4 months later Simmons[4] reported an additional case. In 1943 Sears and Schlesinger[5] published what appears to be the fifth report of breast cancer in a child.

In 1943 Cholnoky[1] reviewed the literature and concluded that in the age group 0 - 1-year there had been 2 deaths and in the 1 - 5-year group 3 deaths from cancer of the breast. In 1944 Smithy[6] reported on a 'sarcoma' and carcinoma which occurred simultaneously in the breast of a 10-year-old girl. In 1956 Haagensen[7] stated that only 6 cases had been described in sufficient detail to enable acceptance of the diagnosis, and Cutler[8] expressed the view in 1961 that cancer of the breast in children below 15 years of age is practically non-existent.

In a more recent review McDivitt and Stewart[9] published 7 cases of juvenile carcinoma of the breast seen at the Memorial Hospital for Cancer, New York, over a 15-year period. The youngest child was 3 years and the oldest 15 years old and all were females. The patient reported by Hartman and Magrish[10] in 1955 presented with a single axillary metastasis, as in the case reported by Byrne *et al.*[11] in 1973. In 1972 Nichini *et al.*[12] published what appears to be the first case report on inflammatory breast cancer in a child. Heidenreich's 1976 review[13] confirmed the rarity of the disease. He collected a total of 131 cases of mammary carcinoma leading to death in children below 14 years of age.

The present case is published not only because of its rarity but because histological findings in the lymph nodes indicated that local mastectomy would have been inadequate therapy, since early metastatic dissemination to the axillary lymph nodes had already occurred. Furthermore, it appears that the benign appearance at operation can be misleading.

CASE REPORT

A 10-year-old Coloured girl was referred to our clinic because the school nurse had noticed a lump in the child's right breast. Apart from mild dental caries she

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enjoyed good health and her development was well within normal limits for age and sex. There was no evidence of anaemia or generalized lymphadenopathy. Abdominal examination revealed no hepatosplenomegaly, ascites, precocious puberty or other endocrine abnormality.

Specific interrogation failed to produce evidence of an injury to the breast, nor could it be established how long the mass had been present.

The mass was clearly visible, occupied the subareolar area, was non-tender and measured 3,5 × 3 cm. Its consistency was that of soft rubber, and it was smooth, not fixed to the chest wall, and freely mobile in all directions but fixed to the areola. Both nipples were of the infantile type but not retracted. There was no skin oedema or nipple discharge. The edge of the mass was rounded and it resembled a lipoma or tensely cystic swelling.

Numerous small lymph nodes were felt in the axillae, but there was no clinical evidence of malignancy. The axillary lymph nodes on the affected side were smaller than those on the contralateral side. All the side-room tests as well as the special investigations were negative.

Because malignancy could not be excluded, the mass and a rim of normal breast tissue which was stretched over the periphery of the tumour were removed in one block and submitted for histological examination. On incision the main mass was found to contain mostly semi-liquid necrotic material which was easily expressed, and the macroscopic appearance was that of a benign cyst.

The wound was sutured with 4/0 Dermalon and a Portavac underwater drainage apparatus was applied.

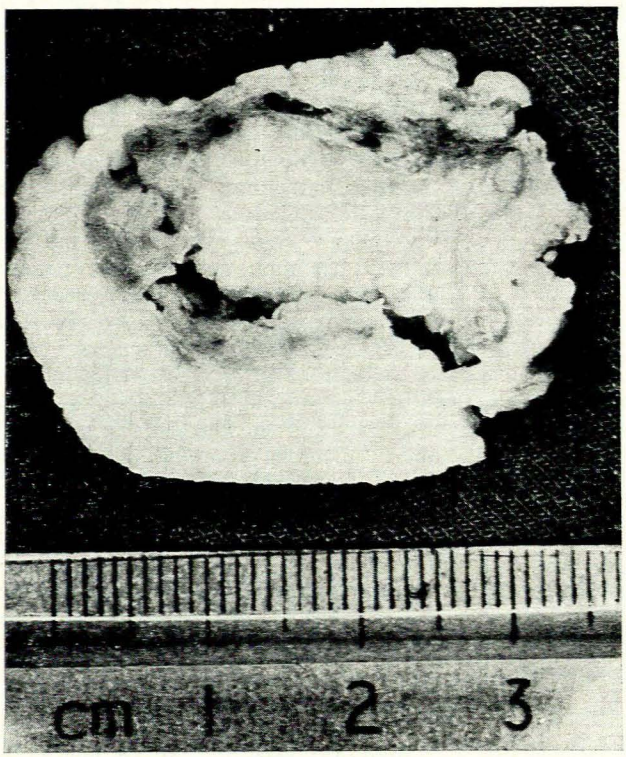

Fig. 1. The cut surface of the circumscribed tumour. The dark peripheral outline is due to haemosiderin deposition.

Pathological Findings

The macroscopic appearance was that of a well-circumscribed oval nodule (measuring 3 × 2 × 2 cm) with surrounding fibrofatty tissue. The colour of the tumour varied from grey-white to yellow and red-brown in some areas (Fig. 1). The consistency was soft in the centre and firmer at the periphery.

Microscopic examination showed the tumour to consist of epithelial cells growing in a papillary and in some areas in a solid fashion, forming duct-like structures and acini (Fig. 2). The cells had pale cytoplasm varying from granular to clear. The nuclei were large and vesicular with prominent nucleoli. Pleomorphism and hyperchromatism of the nuclei were present but not marked. Occasional mitoses, normal and abnormal, were seen. Stains for epithelial mucin revealed the presence of intracellular as well as extracellular accumulations of mucus. Necrosis of tumour tissue was seen only in small areas where the tumour grew more solidly.

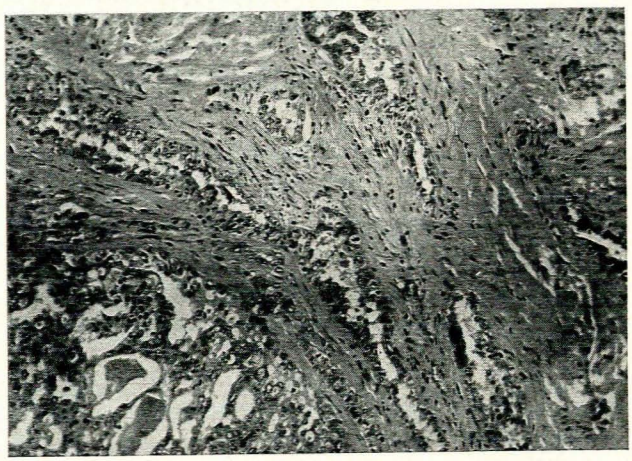

Fig. 2. Juvenile or secretory breast carcinoma is seen to be infiltrating the stroma at the periphery of the tumour (middle). **The tubular pattern is evident in the lower left-hand corner (H and E × 100).**

No true capsule was present, apart from partial compression of the surrounding tissue. The tumour was clearly invasive around normal breast ducts, which in places showed epithelial proliferations but without significant atypia. Heavy deposits of haemosiderin were found in both the connective tissue and epithelial components.

Samples for electron microscopic examination were taken from the formalin-fixed specimen. Ultra-thin sections were studied with a Zeiss electron microscope 9S2. The epithelial cells of the tumour showed mucin secretion in relation to the microvilli and within an acinus. Prominent junctional complexes were evident, and scattered liposomes were present in the cytoplasm (Fig. 3).

The diagnosis of a juvenile secretory breast carcinoma (infiltrating duct type) was made.

With this information at hand, the problem was re-evaluated. Because the disease was clinically in stage I, it was decided to proceed with conventional radical mastectomy, which we believe to be the treatment of choice for

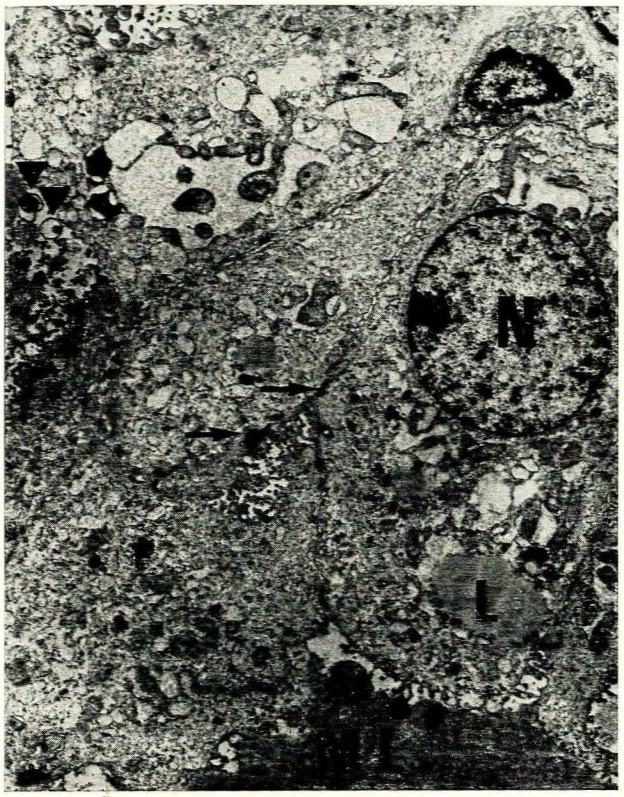

Fig. 3. Electron micrograph of cells of a well-differentiated secretory carcinoma of the female breast. Junctional complexes (long arrows), **mucin granules** (arrowheads), **a mucinous lake (ML), the nucleus (N) and liposomes (L) are well preserved (× 8 300).**

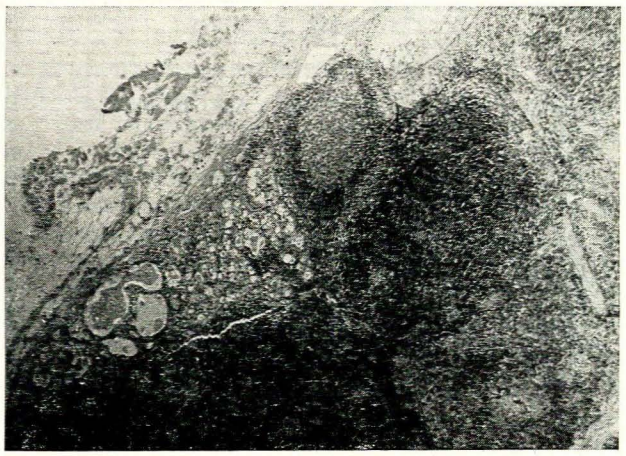

Fig. 4. Tubular carcinoma metastases in the subcapsular sinus of an axillary lymph node (H and E × 100).

early cancer of the breast. A standard Halsted's skin incision was made which incorporated the nipple and areola. The axillary lymph nodes and pectoralis muscles were removed in continuity.

At operation one of the lymph nodes in the anterior axillary group was noticed to be slightly larger than the rest. Histological examination of this node confirmed that the enlargement was due to well-differentiated metastatic spread (Fig. 4); the rest of the axillary nodes were found to be free of disease on histological examination.

DISCUSSION

The symptomatology of carcinoma of the breast in children does not differ from that of the adult counterpart. In the 7 cases reviewed by McDivitt and Stewart[9] each patient presented with an asymptomatic lump in the breast. All the tumours were discovered incidentally by a parent or doctor on routine examination, as in the present case.

Shackelford, cited by Hartman and Magrish,[10] described a case in a schoolboy who had had asymptomatic enlargement of the breast for 7 years before consulting a surgeon. Only when he had developed ecchymosis because of trauma was consent for surgical treatment obtained. At operation he was found to have a pea-sized lesion beneath the nipple, and he died 7 weeks later with widespread metastases — this case illustrates the fact that the size of the tumour bears no relation to its potential for dissemination. Our patient had a large tumour but no clear-cut clinical signs of lymphatic spread.

McDivitt and Stewart[9] believe that the biological behaviour of cancer of the breast in children is less aggressive than in the adult. This view is substantiated by the fact that only 1 of their 7 patients initially treated by excision biopsy had a recurrence of the disease. They could find no correlation between the age at presentation and delay in obtaining immediate biopsies. Four patients were biopsied immediately, and the remaining 3 after periods of 8 months, 1 year and 5 years respectively. Only 1 patient developed a recurrence of the tumour, and all 7 patients eventually remained free of disease for periods of up to 15 years. In one girl who was treated by excision biopsy the tumour recurred locally after 2 and again after 8 years; 2 years after the second re-excision she was still free of disease. The 5-year survival rate for this group of 7 patients was 100%. However, McDivitt and Stewart's[9] view is not supported by that of Byrne *et al.*[11]

Only 1 of the 13 patients studied by Haagensen[7] developed metastases to the axillary lymph nodes. For this reason mammary cancer in the young is often referred to as 'juvenile or secretory carcinoma', a term which implies that breast cancer in children could be treated adequately by local excision without adversely affecting the prognosis.

Teasdale and Baum[14] stress the fact that a variety of histological types of breast cancer may occur in children. Their case report described a 12-year-old girl with a painless lump which had been present for 3 weeks. Histological examination of the lesion revealed it to be an invasive intraductal carcinoma; all the axillary nodes, including the apical nodes, showed extensive tumour involvement. In the case described by Nichini *et al.*,[12] in which the lesion was found to be an inflammatory carcinoma, rapid spread to the opposite breast and ovaries was also observed.

A feature peculiar to secretory carcinoma of the breast in children is the presence of a thick capsule resembling

the wall of a benign cyst. This feature was observed by Hartman and Magrish,[10] who believed the capsule to be the result of a desmoplastic reaction to the tumour. Whether or not this capsule contributes to the better prognosis encountered in the case of 'juvenile' carcinoma remains unknown. McDivitt and Stewart[9] found that in all of 7 cases the surrounding breast tissue was penetrated in a truly infiltrative pattern. The presence of a capsule was directly responsible for the misinterpretation of the nature of the tumour in our patient.

Except in the case of inflammatory carcinoma, the preoperative diagnosis of breast cancer in the young may be difficult and confirmatory investigations may be helpful. Some workers[12] have found mammography a valuable adjunct in the diagnosis of breast carcinoma; they believe that thickening of the skin can be better evaluated by means of mammography than by palpation, especially during the course of treatment.

Thermography is recommended, and is believed to be 80 - 90% accurate. A localized area of increased infrared emission of more than 2°C has been taken as indicating underlying carcinoma or inflammatory disease. Cytological examination as an adjunct to diagnosis has not been commented on, but we believe that a negative cytological examination does not exclude carcinoma of the breast, and prefer to excise the whole tumour and submit it for frozen section examination or routine examination of formalin- and glutaraldehyde-fixed specimens. In our clinic a bone scan forms an integral part of the routine work-up of any patient with cancer of the breast — in the case under discussion this investigation could only be carried out 2 weeks after surgery and was interpreted as normal.

There is no unanimity of opinion or specific guideline with regard to treatment of breast cancer in children. Most publications refer to isolated cases, and the clinician is therefore compelled to formulate his own therapeutic policy. Three of the cases reviewed by McDivitt and Stewart[9] were treated by excision biopsy only, 1 by simple mastectomy and postoperative axillary irradiation, and 1 by excision biopsy followed by modified radical mastectomy 8 years later. Only 1 patient in their series was initially treated by radical mastectomy.

According to McDivitt and Stewart[9] the disease tends to run a relatively favourable course and radical therapy is therefore unnecessary. Byrne *et al.*[11] believe that breast cancer in children has the potential for regional metastasis, a point strongly in favour of radical mastectomy. The patient described by him, the one reported by Hartman, and our patient each presented with a single axillary metastasis; this furnishes further support in favour of radical mastectomy as the treatment of choice. Because only one lymph node was found to harbour metastatic carcinoma, radical surgery was regarded as sufficient therapy in all 3 of the abovementioned cases.

Teasdale and Baum[14] believe that local excision alone may be inadequate, and that in children prognosis is related to the pathological features of the tumour rather than the age of the patient.

Hartman and Magrish[10] consider it important to avoid radiotherapy in young children, and recommend radical mastectomy instead. In their experience, radical surgery does not limit physical activity or development in later life. The patient with inflammatory carcinoma reported by Nichini *et al.*[12] received irradiation and chemotherapy as a form of palliative therapy; response to this 'combination' therapy was poor, and her disease remained uncontrolled.

We believe that children find radical mastectomy psychologically less traumatic than do adults.

In our patient radical mastectomy was performed: *(i)* because of the nomadic habits of the patient's parents; *(ii)* because they live too far from hospital for and cannot afford the financial burden of regular clinic attendance; and *(iii)* because we know that the long-term prognosis (15 - 20 years) for stage I cancer of the breast treated by radical mastectomy is excellent. One month after surgery the child had full movement of the arm with no difficulty in combing her hair without having to flex her head. The present case has proved that a lump in the breast in a child must be approached with the same degree of suspicion as in an adult.

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