ABSTRACT
The purpose of this report is to highlight the clinicopathological features, diagnosis, management and prognosis of rare childhood breast cancer. We herein report a case of slowly growing, locally advanced secretory carcinoma of the breast in an 11-year-old boy with metastatic axillary lymph nodes, 12 of them were totally replaced by the tumor cells. No evidence of distant metastases was detected. Modified radical mastectomy with axillary clearance was done, with uneventful postoperative course. On searching the Sudan medical literature, no previous report of similar case was found. In conclusion, breast carcinoma, though a rare entity, yet should be borne in mind if a firm breast lump was found in a child. Fine needle aspiration cytology is effective to start with as a diagnostic tool.

Key words: Breast cancer, secretory carcinoma, child metastasis.

INTRODUCTION
Breast cancer is exceptionally unusual in children, accounting for less than 1% of all childhood malignancies, and less than 0.1% of breast tumours occur in children. Secretory breast carcinoma (SBC) was first described in 1966 [1-4]. It is a variant of duct carcinoma, was described originally in children although subsequent reports revealed that it may occur in all age groups, mainly during the second and third decade [5].

In fact, published literature reports only a few cases with axillary lymph node metastases and only four cases with distant metastases [6]. The mean age at diagnosis was approximately 11.5 years [7]. Breast tumors in children are classified as being either carcinomas or sarcomas. The majority of sarcomatous lesions are phyllodes tumors, whereas most carcinomas are of a ductal etiology. Although uncommon, malignant disease must be considered in the differential diagnosis of the paediatric patient with a breast mass [8]. Modified radical mastectomy followed by irradiation and chemotherapy for axillary metastasis, is a widely accepted policy.

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CASE REPORT

An eleven years old boy presented to our surgical unit in Omdurman teaching hospital, with a 5 years history of painless lump in the right breast, which increased gradually in size. Recently the patient experienced bloody nipple discharge and his breast started to hurt him. No history of trauma or symptoms suggestive of any system involvement. No features of precocious puberty.

On examination the patient was found to be pale, with 8x7 cm firm irregular breast mass, skin changes and fixation to the nipple-areola complex. Palpation of the right axilla revealed firm matted significant lymphadenopathy. The other breast, axilla and both supraclavicular lymph nodes were not involved. The rest of body exam was unremarkable.

Investigations done preoperatively included, chest radiography and abdominal ultrasound scan, both showed no abnormalities or evidence of metastasis. His haemoglobin was nine gram/dl. Fine needle aspiration cytology (FNAC), yielded bloody material. Microscopically, the smears showed numerous sheets of highly atypical cells with increased Nuclear: Cytoplasmic ratio and hyperchromatic nuclei with presence of heavy mixed inflammatory cells in RBCs background. A suspicious of malignancy was suggested.

After a discussion of the patient condition and a blood transfusion to correct his anemia, a decision for modified radical mastectomy and axillary clearance was made (Figure 1). Post-operatively, the specimen was sent to histopathology.

The report showed, in the gross appearance, a formalin fixed single hemispheric piece of breast tissues measuring 8x7x3 cm partially covered by skin measuring 7 x8 cm. The sample was previously excised into two halves with attached deep margin. The nipple showed fungating mass. Cut sections shows firm whitish homogeneous mass located near the areola. It is 0.5 cm from the closer deep margin, which is ink. Block 1: nipple. Block 2-7: deep margin and representative sections. Block 8-9: Re-gross from the deep margin. The histological sections show a tumor with microcystic “honeycomb” and tubular growth pattern. The small cysts often merge into large spaces filled with eosinophilic secretion closely simulates thyroid follicles. The tubular spaces are surrounded by sclerotic struma and contain micropapillary structures. The neoplastic cells have abundant eosinophilic cytoplasm, vesicular ovoid nuclei, rare mitosis, and intercytoplasmic lumina. The tumor cells are seen at the deep surgical margin. Axillary lymph nodes were found to be matted together. About 12/15 identified axillary lymph nodes are involved or totally replaced by the tumor cells. Interpretation of the histopathology revealed, SBC with involved deep surgical margin and axillary lymph nodes metastasis. Immunohistochemical for ER/PR and HER is not available. The patient run smooth post-operative course without complications and was then referred for chemo-radiation.

DISCUSSION

The age of children with SBC ranges from 3 to 18 years with a male to female ratio of 1: 6 [3, 9]. There is usually a delay of months to years in diagnosis and this is possibly due to the painless nature of the disease. Our patient in fact presented himself to his rural health facility after three years from onset, but was reassured without being investigated [3]. On average, most patients with benign breast lesions presented within 3-5 months of their symptoms, with usually a palpable lump detected by either the child or the mother [8].

Despite the fact that maxillary lymph node involvement in SBC is rare and practically no distant metastases are usually observed [1], our patient actually exhibited significant lymph node involvement. The most common benign breast lesions include
fibroadenoma, fibrocystic breast disease, low grade phyllodes tumor, lipoma, adenoma or sebaceous cyst. The malignant lesions include invasive intraductal carcinoma, a non-Hodgkin lymphoma, or phyllodes tumors, which were typically larger in size and tend to present earlier [6,8]. Phyllodes tumor may be associated with juvenile papillomatosis [10].

FNAC is preferable to excision biopsy to start with, because if the growing breast bud is disturbed, this will lead latter to deformity and asymmetry of the breast. FNAC is a very important diagnostic tool in the paediatric population because it provides a fast, minimally invasive, low risk, and well-tolerated procedure [5]. The morphologic spectrum seen by FNAC was inflammatory lesions, benign ductal cells, ductal hyperplasia, papillary lesions, benign neoplasm, suspicious cytology (0.3%) and cancer (0.3%). Only 3% of the male breast aspirates provide a diagnostic challenge [7]. Vesoulis Z et al reported a case that exhibits cytological changes resembling “lactating adenoma” [11].

SBC has a distinctive pathologic characteristic of large amounts of intracellular and extracellular secretion and granular eosinophilic cytoplasm of neoplastic cells. This neoplasm was initially thought to be a malignant neoplasm of children and called juvenile carcinoma. The term secretory is based on the distinctive histological features and is therefore preferable to the term juvenile [9]. Pathologic variants include intraductal, lobular, medullary, inflammatory, and Secretory carcinoma [2,12,13]. SBC accounted for 84% of these malignant breast tumors in children [3, 14].

SBC has immunohistochemical and molecular characteristics that distinguish them from intraductal carcinoma [15]. Most tumors stain positive for S100 and negative for estrogen receptor, progesterone receptor, and HER2/neu (i.e., triple negative) [16].

However, because SBC is so rare, the methods of surgical treatment and the role of adjuvant therapy, particularly for young patients, remain controversial and no form of therapy is unanimously recommended and accepted [1,16]. Three patients out of 22 were found to have axillary node metastases, local recurrences were reported, but neither systemic metastasis nor disease associated death has been reported in children [9]. This makes our patient as number four to be reported with extensive axillary lymph node involvement. The most recent review of this subject recommended modified radical mastectomy and axillary node dissection as treatment [3,12]. Irradiation and chemotherapy were given in our patient because of axillary metastasis.

SBC tends to have a low-grade clinical course and rare events of axillary lymph node or distant metastases. This results generally in a favourable excellent prognosis, particularly in paediatric group [9]. Tixier H et al reported a child who developed a local relapse and lymph node metastasis after 16 years from the initial diagnosis of SBC [17-18]. In conclusion, breast carcinoma though a rare entity yet should be borne in mind, if a firm breast lump is found in a child. Fine needle aspiration cytology is effective, to start with, as a diagnostic tool.

Figure 1- Post-operative photography of the child with breast cancer.

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REFERENCES