CASE REPORT

Breast carcinoma in a prepubertal girl

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SUMMARY

Breast carcinoma is a very rare disease in children. We present a rare case of breast cancer in an 11-year-old prepubertal girl. Clinically, it was a case of locally advanced breast cancer (T4bN1M0). The core biopsy report showed adenocarcinoma of the not otherwise specified (NOS) variety (oestrogen receptor-negative, progesterone receptor-negative and human epidermal growth factor receptor 2-negative). Diagnosis was late in this case due to lack of suspicion. A modified radical mastectomy was considered to be adequate treatment. Histologically, it was adenocarcinoma NOS, which is rare in young girls (the secretory type being more common). Incidence, differential diagnoses, investigation and management of breast carcinoma in young girls are discussed. The purpose of reporting this case is to highlight that prevention and early detection of breast carcinoma in children is very important.

BACKGROUND

Breast carcinoma is a very rare disease in children. Though most of the breast carcinoma cases are found in females, male breast cancer cases have also been reported.

We present a case of carcinoma of the breast in an 11-year-old prepubertal female child. It was a case of adenocarcinoma not otherwise specified (NOS). Clinical features, investigations, differential diagnoses and treatment modalities have been discussed here.

By reporting this case, we would like to bring to light that breast carcinoma can also occur in the paediatric age group, so it should be kept in mind in the differential diagnosis of a breast lump in a prepubertal girl. We would like to emphasise that early diagnosis and treatment of paediatric breast cancer results in favourable prognosis. We would also like to stress that to standardise the management protocol of paediatric breast carcinoma, more research needs to be performed.

CASE PRESENTATION

An 11-year-old prepubertal girl presented with a lump in the right breast of 8-month duration (figure 1). The lump was painless and gradually increasing in size. There was no sign of any swelling in the axilla or the neck. There was no history of nipple discharge. The opposite breast was normal. There were no other systemic symptoms such as back pain, headache, seizure, haemoptysis or jaundice. There was no family history of breast or ovarian cancer. There was no history of exposure to radiation.

On general survey, the patient was thinly built. No secondary sexual characteristics had yet developed.

On local examination, the lump was occupying the whole of the right breast. Its size was 5×6 cm. The surface was lobulated. The skin appeared tense and dusky with prominent dilated veins overlying the mass. On palpation, the lump was hard in consistency. It was fixed to the overlying skin and the underlying pectoralis major muscle. There was a single 2 cm mobile non-tender lymph node palpable in the anterior group of lymph nodes of the right axilla. The left breast and left axilla were normal. The abdomen, chest, cranium and spine examinations were normal.

The provisional diagnosis made was carcinoma of the right breast T4bN1M0.

INVESTIGATIONS

Sonomammography showed no other lesion in the same breast and axilla. Sonomammography of the opposite breast and axilla was normal (figure 2). Core biopsy of the lump showed infiltrating ductal carcinoma of the NOS variety. Immunohistochemistry was used to evaluate expression of the oestrogen receptor (ER), progesterone receptor (PR) and human epidermal growth factor receptor 2 (HER2). The tumour was ER-negative, PR-negative and HER2-negative. Ultrasonography of the whole abdomen was within normal limits (figure 3).

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included invasive intraductal carcinoma, non-Hodgkin's lymphoma, phyllodes tumours, fibroadenoma, fibrocystic breast disease, low-grade phyllodes tumour, lipoma, adenoma and sebaceous cyst.

Figure 1 Preoperative right-sided breast carcinoma in a prepubertal girl.

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Figure 2 Ultrasonography showing a normal left breast.

TREATMENT

Modified radical mastectomy (Patey modification) with level III axillary lymph node clearance was performed. Part of the pectoralis major muscle was removed as it was infiltrated by the growth (figure 4).

OUTCOME AND FOLLOW-UP

Histopathology of the specimen showed (figure 5)

- ▶ Infiltrating ductal carcinoma NOS—grade II;
- ▶ All surgical margins were clear;
- ▶ Axillary lymph nodes—11 in number, reactive, no metastasis pT3NoMx.

DISCUSSION

Young girls are not considered to be at risk for breast cancer. However, breast cancer can occur at any age. Hence, all women irrespective of age should be aware of their personal risk factors for breast cancer. Less than 0.1% of all breast cancers occur in women under 30 years of age. In children, the average age of primary breast cancer was 11 years, with a range of 3–19 years.¹ In young people, it is usually found in girls, but some cases in boys have also been reported.²

There are several risk factors for developing breast cancer such as:

- ▶ A personal history of breast cancer or some non-cancerous breast diseases;
- ▶ A family history of breast cancer, particularly in a mother, daughter or sister;

Figure 3 Ultrasonography of the abdomen showing a normal liver with no metastasis.

Figure 4 Histopathological examination showing ductal carcinoma.

- ▶ Evidence of a specific genetic defect (BRCA1/BRCA2 mutation);
- ▶ A Gail Index score of at least 1.7% for the development of breast cancer within 5 years, or a 20% lifetime risk;
- ▶ Other risk factors include heavy alcohol use, high intake of red meat, dense breasts, obesity, race and prior radiation to breast.³

Women diagnosed with breast cancer at a younger age are more likely to have a mutation of BRCA1 or BRCA2, p53, PTEN gene, etc. Breast cancer in younger women is more aggressive and less likely to respond to treatment. The most common histological subtype in children is the secretory carcinoma (about 80%).⁴ Other subtypes that have been reported include scirrhous, medullary, anaplastic and papillary adenocarcinoma.²

The most common differential diagnosis of children with a breast mass is fibroadenoma in women and gynaecomastia in men. Other differential diagnoses include fibrocystic breast disease, low-grade phyllodes tumour, lipoma, adenoma, sebaceous cysts, invasive intraductal carcinoma, non-Hodgkin's lymphoma and phyllodes tumours. These are larger in size and tend to present earlier.

Early onset breast cancer has a history of rapid increase in size compared to the above differential diagnosis which has a slower growth.^{5 6}

Diagnosis of breast carcinoma in children is late because of its rarity and painless nature. Excision biopsy affects the growing breast bud, which may lead to deformity and asymmetry of the breast later on; hence, fine-needle aspiration cytology should be

Figure 5 Postoperative picture of the prepubertal girl after modified radical mastectomy.

preferred over excision biopsy.⁷ The triple assessment (palpation, ultrasound examination and core needle biopsy) is currently considered the gold standard for evaluation of breast masses in women younger than 30 years.⁸ Mammography cannot be used as a diagnostic tool in young age groups as the breast has a higher density than in older ages. MRI has been suggested as a diagnostic tool.⁹

Axillary metastasis is rare in children with breast carcinoma. Even if axillary metastasis is present, it rarely involves more than three axillary lymph nodes.¹⁰ Lymphoedema is found in 6–30% of patients after axillary dissection. It is a serious life-long morbidity. Hence, sentinel lymph node biopsy should be used in children just as in adults.¹¹ Breast cancer in younger females is usually ER/PR negative. Even in those with ER-positive tumours, prognosis is poorer than in those with ER-negative tumours.¹²

Treatment protocol for breast carcinoma in adults is now standardised. For early breast carcinoma, upfront surgery in the form of breast conservation (wide local excision followed by radiotherapy to prevent recurrence) or mastectomy is preferred. In locally advanced breast carcinoma, first neoadjuvant chemotherapy followed later by surgery is undertaken. Hormone therapy and chemotherapy are the mainstay of treatment in cases of advance metastatic breast carcinoma.

There is no accepted guideline for management of breast carcinoma in children. Treatment ranges from excision biopsy to radical mastectomy. Whenever possible, prepubertal girls should be treated initially with wide local excision. Every attempt should be made to preserve the growing breast bud so that normal development is not impaired, but in most of the cases it is not possible. Breast tissue near the margin of the tumour should be sampled generously. It should be examined for any metaplastic changes. These can indicate an increased risk of local recurrence. Mastectomy may be necessary.¹³ For advanced cases, modified radical mastectomy followed by irradiation and chemotherapy for axillary metastasis is a widely accepted policy. Postoperative irradiation reduces local recurrences. The main

disadvantage of postoperative radiotherapy is possible secondary effects such as fibrosis of the lung, rib damage and the consequent asymmetry of the rib cage of the growing child.^{14 15}

The prognosis for children with breast carcinoma is poor. Of all the histological types of breast cancer in the young people, juvenile secretory carcinoma has the best prognosis. Poor prognosis in young patients could be related to reduced screening, more aggressive disease and delayed diagnosis. Even after adjusting for stage, histology and grade, younger females still have a significantly poorer survival.¹⁶

Prevention and early detection of breast carcinoma in these young females is very important. At present, no definitive data on this are available. Future recommendation may be based on some of the ongoing trials on these topics.⁸

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REFERENCES

- Murphy JJ, Morzaria S, Gow KW, *et al.* Breast cancer in a 6-year-old child. *J Pediatr Surg* 2000;35:765–7.
- Dugue G, Bock G, Molho L, *et al.* Breast cancer in the young. *J Natl Med Assoc* 1989;81:1184,1187–8.
- Iglehart JD, Smith BL. Diseases of the breast. In: Townsend CM Jr, Beauchamp RD, Evers BM, Mattox KL. eds. *Sabiston textbook of surgery*. 18th edn. The Saunders 2008:1389–93.
- Tanimura A, Konaka K. Carcinoma of the breast in a 5 years old girl. *Acta Pathol Jpn* 1980;30:157–60.
- Ozumba BC, Nzegwu MA, Anyikam A, *et al.* Breast disease in children and adolescents in eastern Nigeria—a five-year study. *J Pediatr Adolesc Gynecol* 2009;22:169–72.
- Ravichandran D, Naz S. A study of children and adolescents referred to a rapid diagnosis breast clinic. *Eur J Pediatr Surg* 2006;16:303–6.
- Hamza AA, Ngwangki LS, Taha O. Breast carcinoma in a boy with metastatic axillary lymph nodes. *Sudanese J Paediatr* 2012;12:89–92.
- Shannon C, Smith IE. Breast cancer in adolescents and young women. *Eur J Cancer* 2003;39:2632–4.
- Abdul Rashid S, Rahmat K, Jayaprasagam K, *et al.* Medullary carcinoma of the breast: role of contrast-enhanced MRI in the diagnosis of multiple breast lesions. *Biomed Imaging Interv J* 2009;5:e27.
- Rosen PP. *Rosen's breast pathology*. Philadelphia: JB Lippincott 2009;563–70.
- Yorozuya K, Takahashi E, Kousaka J, *et al.* A case of estrogen receptor positive secretory carcinoma in a 9-Year-old girl with ETV6-NTRK3 fusion gene. *Jpn J Clin Oncol* 2012;42:208–11.
- Colleoni M, Rotmensz N, Robertson C, *et al.* Very young women (<35years) with operable breast cancer: features of disease at presentation. *Ann Oncol* 2002;13:273–9.
- Costa NM, Rodrigues H, Pereira H, *et al.* Secretory breast carcinoma—case report and review of the medical literature. *Breast* 2004;13:353–5.
- Szántó J, András C, Tsakiris J, *et al.* Secretory breast cancer in a 7.5-year old boy. *Breast* 2004;13:439–42.
- Ferguson TB Jr, McCarty KS Jr, Filston HC. Juvenile secretory carcinoma and juvenile papillomatosis: diagnosis and treatment. *J Pediatr Surg* 1987;22:637–9.
- Jayasinghe UW, Taylor R, Boyages J. Is age at diagnosis an independent prognostic factor for survival following breast cancer? *ANZ J Surg* 2005;75:762–7.

Learning points

- ▶ Breast carcinoma can occur in children.
- ▶ Diagnosis of breast carcinoma in young females is late because of the low index of suspicion.
- ▶ Axillary metastasis is rare in young females with breast carcinoma.
- ▶ Fine-needle aspiration cytology and MRI are used to confirm the diagnosis.
- ▶ Treatment is wide local excision for early stage disease with modified radical mastectomy for late stage disease.

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