Primary Primitive Neuroectodermal Tumor of the Breast: A Rare Case Presentation

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Abstract A primitive neuroectodermal tumor (PNET) in the breast developed in a 36-year-old Indian woman who initially underwent lumpectomy and was diagnosed as the malignant phyllodes tumor of the right breast. Within 2 months it recurred, clinicoradiologically appearing like organized collection. Incision and drainage along with biopsy was done. The tissue diagnosis was reported as PNET. The histopathology report showed the tumor cells as malignant round cells, immunohistochemically positive for CD99, vimentin and neuron-specific enolase (NSE) (patchy) and negative for CD45, cytokeratin, S100, and desmin. Extended simple mastectomy was carried out. She came after another interval with recurrence. Chemotherapy as well as radiotherapy was given. After 18 months of surgery, the patient is having persistent stable disease without distant metastasis. PNET in adults is rare and has been reported in the chest wall (Askin tumor) and other visceral sites. To our knowledge, only a few cases have been reported of a primary PNET of the breast.

Keywords Primitive neuroectodermal tumor · Malignant phyllodes tumor · Breast

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Case Report

A 36-year-old married Indian woman was referred to Cancer Centre Welfare Home and Research Institute after lumpectomy as a malignant phyllodes tumor in the right breast.

On examination of the right breast, no mass was felt except a scar mark of previous lumpectomy. Mammogram and ultrasonography of both breasts did not reveal any positive finding. USG abdomen and chest X-ray reports were normal. The review of lumpectomy histopathology slides in our institute reported it as same (i.e., malignant phyllodes tumor). Wide excision was done with a good margin; after nearly 1 month of the first surgery, histopathological examination (HPE) reported no residual tumor.

After 2 months, the patient presented again with the lump measuring 8 cm × 5 cm and pain in the right breast. Clinical impression was serous collection with some solid component. USG breast showed multiple areas of heterogenous echotexture with multiple septae, impressing organized collection. Fine-needle aspiration cytology (FNAC) was reported as duct carcinoma but it did not corroborate clinically. Incision and drainage was done. Blood-mixed fluid with a few friable tissues came out. The histopathology report showed sheets of neoplastic round cells with high nucleocytoplasmic ratio and coarse chromatin. Brisk mitoses were noted. A few pseudorosettes and rosettes were seen. With immunohistochemistry (IHC), the report came as PNET (vimentin positive, CD99 strongly positive, NSE patchy positivity; CD45, cytokeratin, S100, and desmin negative).

Right extended simple mastectomy with en-bloc resection of pectoralis major muscle was done (Fig. 1a). On gross examination of specimen, a grayish white fleshy soft mass with partly necrotic area was found. Final diagnosis was given as PNET of the right breast based on IHC findings (Fig. 2). Level I lymph nodes were reactive. The pectoralis major muscle was involved, but all margins were free. The

Fig. 1 **a** Right extended simple mastectomy specimen, **b** Recurrent tumor, **c** CECT showing the recurrent tumor

initial lumpectomy slides were reviewed and confirmed as PNET of the breast.

The patient was discharged uneventfully with plan of chemotherapy.

The patient did not turn for follow-up. After 4 months she again presented with the recurrent tumor over the medial part of right pectoral region covering sternum (Fig. 1b and c) of 9 cm × 6 cm. She received six cycles of vincristine, doxorubicin, and cyclophosphamide followed by radiotherapy. There was partial response, so she again received three cycles of chemotherapy (ifosfamide, etoposide, and mesna). After 18 months of surgery, the patient is having persistent stable disease without distant metastasis. Chemotherapy is still continued.

Fig. 2 Small round cells with pseudo-rosette, H/E stain high power x40; [Inset—strong CD99 membrane positivity(brown colour)]

Discussion

PNET belongs to Ewing's sarcoma family of tumors (ESFT) [1]. It is a small, round, blue cell sarcomatous neoplasm of bone and soft tissue that arises from neural crest [2, 3]. It gets its name PNET because the majority of the cells in the tumor are derived from neuroectoderm, but have not devolved and differentiated in the way a normal neuron would do and so the cell appears primitive [3]. PNET primarily occurs in children and adolescents [3]. It is a rare tumor in the breast [2, 4, 5].

The defining feature of ESFT includes a characteristic (11:22) chromosomal translocation and near universal expression of CD99 antigen, though other translocations do occur [1, 2].

Primary PNETs demonstrate a predilection for the truncal and axial soft tissues including the chest wall (Askin tumor), the paravertebral region (50–60 % of cases), and the extremities (20–25 % of cases) [4]. The thoracopulmonary region (Askin tumor) is the single most common primary site [4]. Primary PNET of many organs such as the kidney, ureter, bladder, testis, seminal vesicles, ovary, pancreas, uterus, parotid gland, and lungs have been documented but only a very few case reports of primary PNET of the breast exist [4, 5].

In our case, radiation and prolonged chemotherapy was necessary because of initial misdiagnosis.

Since the treatment is radically different from a duct carcinoma and malignant phyllodes tumor, we insist that rare-appearing tumors should always be confirmed by immunohistochemistry. This will definitely help the

patient to achieve a long disease-free survival if the appropriate treatment with surgery and chemoradiation therapy is given.

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