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Congenital Ewing's Sarcoma/Primitive Neuroectodermal Tumor of the Upper Extremity

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Dear Editor,

The patient was a female newborn, 3200 g in weight, 50 Cm in length, who presented at birth with a firm mass on the left upper arm, measuring 25 × 16 × 45 mm in size. Physical examination revealed a non-movable mass with firm consistency and mild tenderness on the left upper arm. Family and maternal histories were unnoteworthy, and the laboratory examination was normal. An excisional biopsy of the mass was performed and the histopathological findings of the excised lesion were ES/PNET. Immunohistochemistry was positive for CD99, NSE, PCK, desmin, and negative for CK. In enhanced chest CT, abdominal ultrasonography, and whole body bone scan no evidence of abnormality or metastasis was detected. Bone marrow biopsy and further treatment were refused by her parents.

Unfortunately, the sarcoma recurred one year later. Seven cycles of VIDE regimen (Vincristine 0.25 mg d1; Ifosfamide 500 mg d1-4; Doxorubicin 3.5 mg d1-4; Teniposide 50 mg d1, 25 mg d2-4) was administered. After radiation therapy (3040cGy/19 fractions, 6 MeV electrons), the patient underwent autologous stem cell transplantation and achieved a complete remission. Two years later, the patient came to our institution presenting with a subcutaneous mass near the area of surgical scar on the left upper arm. The ultraphonic revealed several hypoechoic nodules between the skin and subcutaneous fat layer on the left upper arm, the biggest one measured 15 × 5mm in size. X-ray (Panel A) showed no adjacent humerus destruction. Magnetic Resonance Imaging showed several nodules on the shallow surface of biceps muscle of the left arm with slight enhancement and marked swelling of the surrounding soft tissue (Panel B, C, arrow). Chest CT scan and whole body bone scan were normal. Subsequent excisional biopsies were performed on 18/11/2013, and the postoperative pathologic results were Ewing's sarcoma. The patient was treated by a course of three-dimensional conformal radiotherapy (3D-CRT) after surgery. Dose of external irradiation was 4680cGy/26 fractions (6 MeV X-ray) in 30 days. The patient is alive 20 months after the second relapse.

ES and PNET share common antigenic profiles, cytogenic aberrations and proto-oncogene expression which are derived from the same primordial bone marrow-derived mesenchymal stem cell (1). At the molecular level, ES/PNET is characterized by chromosomal translocations that fuse the EWSR1 gene to some members of the ETS family of transcription factors, being FLI1 the most frequently implicated [t (11; 22) (q24; q12)] (2). The incidence of these tumors in early childhood is rare, accounting for less than 10 cases per million each year (3), and the incidence in newborns is exceedingly rare. Wong et al reported 1975 patients with Ewing's sarcoma, only 39 (2.0%) were diagnosed at < 12 months of age (4). ES/PNET with a slight male predominance occur predominantly in primarily white and Hispanic patients; however, these tumors are extremely rare in individuals of African or Asian origin (2). ES/PNET account for approximately 10% of primary malignant bone tumors and mainly affect the long bones (47%), pelvis or ribs (5). While 30% are seen in an extraskeletal location of the soft tissues, 5% occurred in extremities among these extraskeletal Ewing's sarcomas (6, 7). Manifestation in extraskeletal location of the soft tissues of extremities rarely occurs and this would appear to be the first description of a congenital ES/PNET arising in the soft tissue of upper extremity.

Multiagent systemic therapy for localized ES/PNET

achieved a 5-year survival of approximatly 70 - 75% (8-10). The similarity in survival between older and younger patients with Ewing sarcoma was reported in literature (11). A study including 2202 ES/PNETs demonstrated that extraskeletal ES/PNETs have a superior prognosis compared with skeletal Ewing sarcoma. About 30 – 40% of patients suffer from recurrent tumors and have a very poor prognosis (12). The 5-year survival following recurrence of these patients is approximately 10 – 15%, while it was 7% for those whose disease recurred within 2 years (13). In our case of disease recurrence after surgery, the patient was treated with multiagent chemotherapy, stem cell transplantation and aggressive surgical treatment, and is still surviving.

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