Pediatrics and Neonatology (2014) xx, 1–4

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.pediatr-neonatol.com

CASE REPORT

Congenital Ewing's Sarcoma/Peripheral Primitive Neuroectodermal Tumor: A Case Report and Review of the Literature

Shu-Guang Jin*, **Xiao-Ping Jiang**, **Lin Zhong**

Department of Pediatric Surgery, West China Hospital of Sichuan University, Chengdu 610041, China

Received May 26, 2013; received in revised form Oct 9, 2013; accepted Nov 2, 2013

Key Words
diagnosis;
Ewing's sarcoma;
newborn;
peripheral primitive neuroectodermal tumor;
prognosis;
treatment

Ewing's sarcoma (EWS) and peripheral primitive neuroectodermal tumor (pPNET) are small round cell malignancies that develop in soft tissue and bone. They very rarely affect newborns. A diagnosis of EWS/pPNET depends mainly on immunohistochemistry and molecular/genetic assays. Since these tumors are highly aggressive, patient prognosis is typically very poor, and treatment remains a challenge. Here, we report a 13-day-old newborn diagnosed with congenital EWS/pPNET and describe its treatment.

1. Introduction

Ewing's sarcoma (EWS) and peripheral primitive neuroectodermal tumor (pPNET) are two types of malignant tumor that are histologically characterized by the presence of small, round cells in soft tissues and bone.[1] This rare malignancy occurs primarily in children and adolescents, but it is extremely rare in newborns. Here, we describe a 13-day-old newborn diagnosed with EWS/pPNET in her upper left arm.

2. Case report

A 13-day-old newborn female was admitted due to a swollen mass on her upper-left arm after birth. She was a full-term cesarean neonate without a family history of genetic diseases. A physical examination revealed a round, soft, dark mass with clear boundaries, measuring about 5 cm × 5 cm × 4 cm, in her upper left arm. There was no increase in local skin temperature or any detectable fluctuation. An ultrasound showed a well-defined isoechoic mass close to the brachial artery with point-like blood flow within the mass, and rich blood flow surrounding the mass. Computed tomography (CT) detected an isodense soft tissue mass measuring 4.6 cm × 3.7 cm in the coronal maximum of her upper left arm, which was attached to the biceps and triceps (Figure 1). Enhanced scanning further showed a heterogeneously enhanced mass pressing onto the

* Corresponding author. Department of Pediatric Surgery, West China Hospital of Sichuan University, Chengdu 610041, China.
E-mail address: shgjin2003@aliyun.com (S.-G. Jin).

1875-9572/$36
http://dx.doi.org/10.1016/j.pedneo.2013.11.002

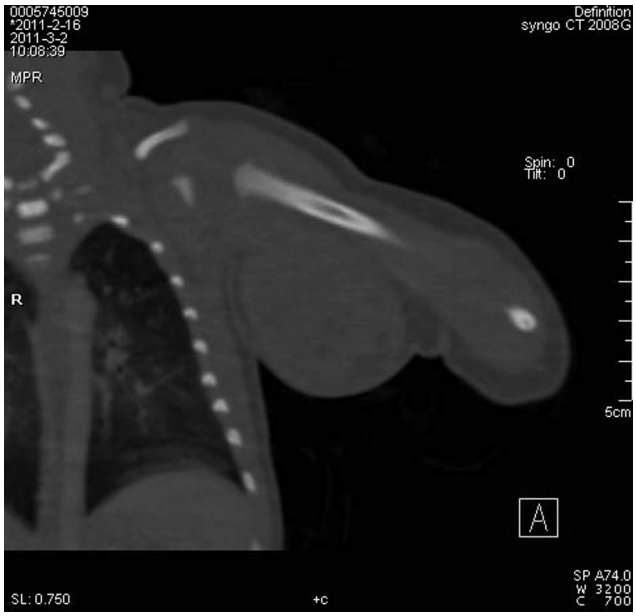

Figure 1 Computed tomography detects a mass present in the upper left arm.

left brachial artery, with branches of the brachial artery extending within the mass. The bone in the left humerus did not appear to be affected.

The mass was completely resected and, upon gross inspection, had the appearance of a fish-like cut surface. A pathologic examination found histological evidence of malignant, small round tumor cells that were not accompanied by a margin of residual tumor cells (Figure 2). The tumor cells were immunohistochemically positive for CD99 (Figure 2) and Ki67 (40%; Figure 2), and negative for desmin, myogenin, synaptophysin, leukocyte common antigen, S-100 protein, P63 protein, smooth muscle antibody, and epithelial membrane antigen. To detect chromosome 22q12 translocation, fluorescence *in situ* hybridization (FISH) was employed using LSI EWSR1 (22q12) Dual Color, Break Apart Rearrangement Probe (Vysis; Abbott Molecular, Des Plaines, IL, USA). Fluorescence was detected using a microscope with a Y-Fl Epi-Fluorescence Attachment (Nikon, Tokyo, Japan). Tumor cell nuclei that exhibited a split of signal pair were scored as positive for translocation and rearrangement of chromosome 22q12 (Figure 2).

After surgery, enhanced chest CT and abdominal ultrasonography were performed. No evidence of abnormality or metastasis was detected. However, it was recommended that the upper left limb be amputated. The patient's parents refused, and the infant was transferred to the oncology department for further treatment 7 days after surgery. Due to the infant's young age and the immaturity of her organs, she received four courses of chemotherapy with cisplatin (20 mg/m^2.day, quaque die $\times$ 4 days, once every 3 weeks) and cyclophosphamide (0.2/ m^2·d, qd $\times$ 4 days, once every 3 weeks). However, 3 months later, distant metastasis involving the lung and liver were detected without local recurrence. Gradually, the infant became emaciated and infirm, and eventually she died of dyscrasia.

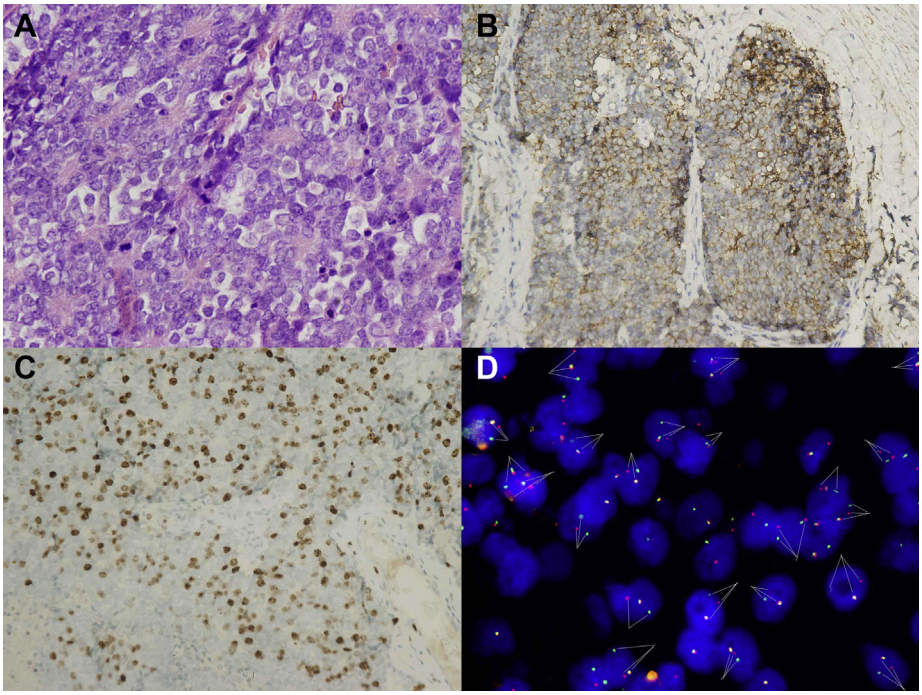

Figure 2 (A) A uniform population of small round cells with minimal cytoplasm and fine granular chromatin nuclei stained with hematoxylin and eosin (magnification, 300×). (B) Neoplastic cells with strong expression of CD99 (En Vision; magnification, 300×). (C): Expression of Ki67 (40%; En Vision; magnification, 300×). (D) Fluorescence *in situ* hybridization reveals that tumor cells are characterized by one fused, one red, and one green signal pattern (labeled with lines), and chromosome translocations of the *EWSR1* region have occurred in 22/50 nuclei.

Please cite this article in press as: Jin S-G, et al., Congenital Ewing's Sarcoma/Peripheral Primitive Neuroectodermal Tumor: A Case Report and Review of the Literature, Pediatrics and Neonatology (2014), http://dx.doi.org/10.1016/j.pedneo.2013.11.002