Congenital Ewing's sarcoma of the humerus—a case report

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On June 14, 1995 a full-term female baby was born at our hospital. Her birth weight was 2900 g. Immediately after birth, she was found to have a very large mass on her right arm. There was no history of a difficult delivery or birth injury. The family history was unremarkable.

Physical examination revealed a healthy baby, except for a huge 10 cm erythematous mass on her right arm. Radiographs revealed a very large tumor mass in the humerus, with extensive sun-burst periosteal reaction and soft tissue extension (Figure 1). A radionuclide bone scan showed increased activity in the right humerus, but not in other parts of the skeleton. MRI showed a very large tumor arising from the right humerus with soft tissue extension. Investigation for neuroblastoma, including urine vanillylmandelic acid was within normal limits. An open biopsy was performed. On histological examination, it showed dense sheets of round to ovoid malignant cells which were separated by fibrovascular stroma (Figure 2). Perivascular aggregation of the cells was also evident. The periodic acid-Schiff (PAS) stain for glycogen was positive in most of the neoplastic cells. The immunohistochemical studies for neural markers, such as neurospecific enolase, chromogranine A, synaptophysin and S-100 protein were all negative. The pathologic diagnosis was Ewing's sarcoma. The family refused any suggestions of radiotherapy or chemotherapy. Half a year later, the arm mass had become extremely large, measuring 26 × 15 × 20 cm with superficial venous engorgement and the overlying skin had almost been pierced by the tumor (Figure 3). At that time, chest roentgenogram and radionuclide bone scan were still normal. On December 23, 1995, she underwent a disarticulation of the right shoulder. The postoperative course was uneventful. Her family still refused chemotherapy. The child developed lung and liver metastases in December 1996 and died of the disease 6 months later.

Discussion

The diagnosis of Ewing's sarcoma seems very likely given the radiologic picture and the histopathology including the positive PAS stain. Since the immunohistochemistry was negative, PNET is

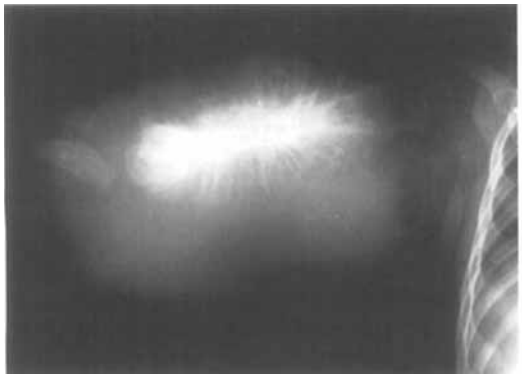

Figure 1. Osteoblastic lesion in the right humeral shaft with extensive sun-burst periosteal reaction and a very large soft tissue mass.

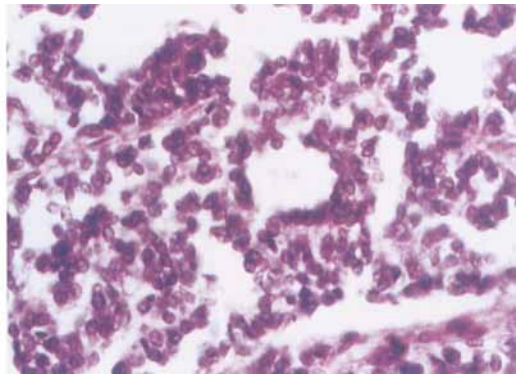

Figure 2. Uniform round to ovoid cells with intervening fibrous tissue. Note the perivascular arrangement of the tumor cells (HE, ×130).

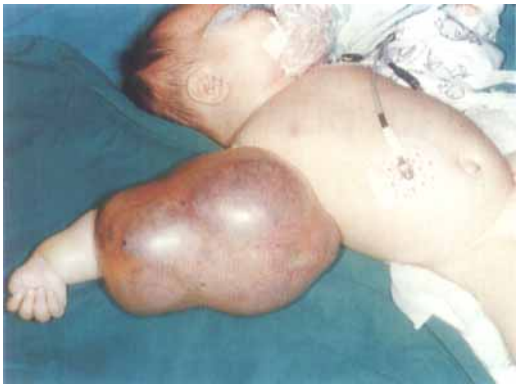

Figure 3. At the age of 6 months.

excluded. Cytogenetics was not carried out at that time. To our knowledge, this is the first reported case of congenital Ewing's sarcoma of bone in the English literature (Mirra 1989, Unni 1996). There was no family history of malignancy. The mother had no medications or exposure to radiation during her pregnancy.

The huge size of the tumor, with the threat of rupture of the skin 6 months later, made the amputation unavoidable. Kim et al. (1986) reported a successful limb salvage in a 5.5-month-old infant with Ewing's sarcoma, using radiation therapy, followed by chemotherapy with vincristine, cyclophosphamide, dactinomycin and doxorubicin. 7 years later, their patient was disease-free with a leg length discrepancy of 3 cm. The parents' refusal prevented us from giving our patient chemotherapy and radiotherapy.

Kim T H, Zaatari G, Atkinson G O, McLaren J R, Ragab A H. Ewing's sarcoma of a lower extremity in an infant. A therapeutic dilemma. Cancer 1986; 58 (1): 187-9.

Mirra J M. Bone tumors. Clinical, radiologic and pathologic correlations. Lea and Febiger, Philadelphia, London 1989: 1088.

Unni K K. Dahlin's bone tumors. General aspects and data on 11087 cases. 5th Ed. Lippincott, Raven, Philadelphia, New York 1996: 249.