CONJONCTIVE SOFT TISSUE EWING’S SARCOMA
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ABSTRACT
A 15 days old female baby presented with a 6 x 7 cm reddish lump with irregular margins on her right scapular region, since birth. Histopathology and immunohistochemistry of excised tissue revealed it to be soft tissue Ewing’s sarcoma. Postoperatively, she received only one dose of chemotherapy but could not survive and expired at the age of one month.

Key words: Ewing’s sarcoma. Soft tissue mass. Chemotherapy. Treatment. Congenital tumour.

INTRODUCTION
Ewing’s sarcoma, a close kin of peripheral primitive neuroectodermal tumours, is a highly aggressive and poorly differentiated, small round blue cell tumour, with unknown histiogenesis. Ewing’s sarcoma family tumours (ESFT) include; classical Ewing’s sarcoma of bone, soft tissue Ewing’s sarcoma, Askin tumour of chest wall and peripheral primitive neuroectodermal tumours of bone and soft tissues.1 The most common chromosomal translocation is between chromosomes 11, 22 t (11:22) q24:q12) and also between chromosomes 21:22.2,3 Congenital or neonatal Ewing’s sarcoma is extremely rare4, hence this case is reported.

CASE REPORT
A 15 days old female baby weighing 2.8 kg was brought with a reddish lump on her right scapular region since birth. The lump was approximately 6 x 7 cm in size with irregular margins and oozing at places as shown in Figure 1. The baby had no other obvious pathology. She was born at 39th week of gestation with uncomplicated delivery. Mother had no exposure to drugs or radiation during pregnancy and there was no significant illness in family. The antenatal period has been unremarkable. Investigations were performed including complete blood count, X-ray chest and ultrasound abdomen, all of which were within normal limits. The lump was excised and sent for biopsy.

Histopathology showed wide infiltration of the dermis by solid sheets of small uniform round blue cells with well-developed vascular network (Figure 2). These cells had indistinct outline giving syncial appearance, with cytoplasm containing abundant glycogen, and conspicuous nuclei. The nuclei were round showing indentation, and fragment mitosis. At places, cells were seen encroaching on the skin surface with resultant ulceration. The tissue was negative for CD31, LCA, S100, Tdt, Desmin and Cytokeratin AE1/AE3 stains, but positive for MIC2 favouring Ewing’s sarcoma. Molecular study showed chromosomal translocation between chromosomes 11, 22 t(11:22)q24:q12).

On the 10th postoperative day, baby developed metastatic swelling on right side of neck approximately 3x3 cm in size and rapidly progressive in size (Figure 3). The new-onset lump was also excised and the patient was referred to paediatric oncologist for management. Two-drug (Vincristine and Adriamycin) chemotherapy was started with the consent of parents. Baby could not survive and expired on the 3rd post-chemotherapy day due to respiratory problem and severe sepsis.

DISCUSSION
Congenital solid tumours are rare and account for 2% of all childhood malignancies.5 Congenital soft tissue Ewing’s sarcoma have been reported at various parts of body; including face, hand, anterior chest wall, and retroperitoneum,1 while in this case it was seen on the scapular region. ESFT usually appear in young adults and adolescents, but are rarely seen in new born. Coffin et al. reported that paediatric soft tissue tumours account for 4% and 14% of the patients were under the age of 5 years.6 Diagnosis of ESFT is based on histopathology, immunohistochemistry and molecular study. On histopathology, round blue cells are seen in variety of ESFT, as reported in this patient, but these cells are also seen in; alveolar rhabdomyosarcoma, desmoplasic round cell tumours, lymphoma, neuroblastoma, poorly differentiated synovial sarcoma and midline carcinoma with t(5:19) translocation.7

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Immunohistochemistry is a reliable diagnostic modality in differentiating ESFT from other round blue cell tumours. Though MIC2 is strongly indicative of ESFT as was observed in this case, it is not restricted to ESFT and so the use of a panel of antibodies is necessary. Moreover, poor sampling and improper staining may cause diagnostic difficulties, while molecular study is the final and most accurate diagnostic tool which shows the translocation of genes as was seen in this case.

Treatment options include complete excision of lesion, chemotherapy and radiotherapy but standard protocol of treatment has not been established yet, because of small volume of patients. Meaaza et al. used two-drug regimen; Vincristine and Adriamycin in non-metastatic cases, while Kim et al. believe that patients presenting with metastasis should receive 5 drugs including Vincristine, Adriamycin, Cyclophosphamide, Ifosfamide and Etoposide. European paediatric soft tissue group recommends avoiding the use of Ifosfamide and Anthracycline in infants less than one and 3 months old respectively, due to immaturity of organs and slow clearance leading to higher rate of complications. Excision of mass and intense chemotherapy along with counselling to parents is preferred mode of treatment as suggested by Kim et al.

Prognosis for congenital ESFT is poor, while in metastasis cases it is very poor. Tumour cell fusion gene has also an impact on prognosis. Optimal therapy in neonates is debatable as the cure rate is very low, while morbidity and mortality is very high. Introduction of new treatment approaches including the stem cell transplantation and targeting of EWS-FL11-related molecules may be helpful in improving the prognosis.

The diagnostic modalities and treatment regimen for ESFT in neonates still need further studies.

REFERENCES