CASE REPORT

Synovial Sarcoma in a Neonate

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Received: July 28, 2015;
Initial review: September 29, 2015;
Accepted: December 28, 2015.

Background: Malignant tumors in neonates are rare. **Case characteristics:** A tumor was detected in the left biceps of a 3-day old neonate. Tumor biopsy and molecular study confirmed the diagnosis of synovial sarcoma. The child received multi-modality treatment with surgery and chemotherapy. **Outcome:** The child is disease-free on follow-up period of 12 months. **Message:** Synovial sarcoma can rarely occur in a neonate.

Keyword: *Chemotherapy, Malignancy, Neonate, Tumor.*

Malignancies in the neonatal age group are rare, and are usually congenital in origin [1]. Neuroblastoma and teratomas are the most common cancers seen in neonates. We present a neonate with synovial sarcoma who was successfully managed with multimodal treatment including chemotherapy and surgery.

CASE REPORT

A 21-day-old girl was brought to our hospital by her parents with complaints of swelling in her left arm. The child was born to non-consanguineous parents. She was a full-term normal vaginal delivery, and weighed 3 kg at birth. The mother had an uncomplicated pregnancy and her antenatal ultrasound studies were normal. The child received BCG vaccine in her left arm on the second day of life. The parents noticed a swelling in her left arm below the BCG vaccination site, 2 days after the vaccine was administered. The child was seen by a general surgeon, who suspected the swelling to be BCG vaccine-related abscess in the left biceps muscle, and attempted surgical drainage of the swelling. However, no pus was drained and the swelling could only be removed piecemeal. The histopathological examination (HPE) of the tissue was consistent with sarcoma. The swelling increased in size after the surgical exploration and the parents reported to our hospital for further management. On evaluation, a hard non-tender mass (5×3 cm) was palpable in the left biceps muscle (***Fig.* 1a**). The mass was mobile in the direction perpendicular to the axis of the muscle fibres. There were no dysmorphic features, and no other significant findings were noticed on examination. Magnetic Resonance Imaging (MRI) of the left arm showed a well-defined mass (3.4×3.1×2.4 cm) in left biceps muscle. The mass was iso-intense on T1 and hyperintense on T2 (***Fig.* 1b**). The computed tomographic (CT) imaging of chest and ultrasound imaging of abdomen and pelvis did not show any evidence of metastatic disease. Histopathological

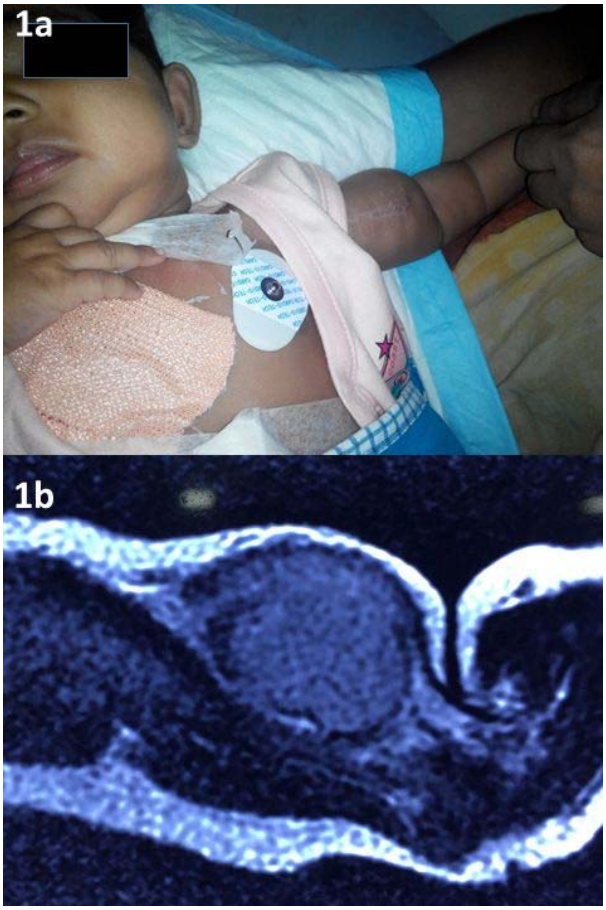

***FIG.* 1** *(a) soft tissue swelling in the left biceps muscles; (b): MRI showing mass in left biceps muscle.*

examination of patient's formalin-fixed operated specimen at our hospital confirmed the diagnosis of synovial sarcoma (***Web Fig.* 1**). Immunohistochemistry (IHC) showed that the tumor cells were positive for EMA, CD68, SMA, CD99, Vimentin, CD34 and CD56, and were negative for Keratin, Myogenin and Desmin. Polymerase Chain Reaction (PCR) of tumor specimen was positive for *SYT-SSX4* translocation, which confirmed the diagnosis of synovial sarcoma. The child received six cycles of chemotherapy with Ifosfamide 900 mg/m^2/day for three days and adriamycin 15 mg/m^2/day for two days given once in every three weeks. Three cycles of chemotherapy were given before surgical resection of the tumor, and three cycles were given after the resection. There was a decrease in size of the mass by 50% after the first 3 cycles of chemotherapy. Complete wide local excision of the tumor bearing biceps brachii muscle and adjoining soft tissue was done by a team of surgical oncologist and plastic surgeon. The histopathological examination of the grey white area of the specimen revealed only fibrosis, chronic inflammatory cells and foreign body giant cells with no residual tumor suggesting a complete pathological response to chemotherapy. Microscopic examination all the margins, adventitia and perineurium were free of tumor. The patient was not given radiotherapy to the local site in view of her age, anticipated significant long-term toxicity, and complete pathological response to chemotherapy. The patient is currently on follow-up for last 12 months and is disease-free. Her growth and development are normal.

DISCUSSION

Neonatal malignant tumors are extremely rare and histologically heterogeneous. They are challenging and difficult to treat when compared to older children. Chemotherapy and radiotherapy are associated with increased short-term and long-term toxicities in neonates because of the immaturity of liver, lungs, brain and kidneys. Soft tissue neoplasms in neonates are commonly benign and vascular in origin [2]. Rhabdomyosarcoma is the most common malignant soft tissue tumor in neonates. Synovial sarcoma in a neonate is exceedingly rare. In an earlier report [3], the diagnosis of synovial sarcoma in the neonate having mass in left arm was made on histopathological examination alone, and was not confirmed by molecular studies. The gold standard for diagnosing synovial sarcoma currently is the demonstration of t(x,18) translocation by PCR [4]. The t(x,18) translocation involves the translocation of SYT gene on chromosome 18 and either *SSX-1, SSX-2 or SSX-4* gene on X chromosome. The *SYT-SSX4* translocation seen in our patient is rarer than the commonly seen *SYT-SSX1* or *SYT-SSX-2* translocation [5]. This case is being presented for its rarity and successful management and also to alert the pediatrician that not all swellings in the neonatal period are benign. Early diagnosis and effective treatment is vital for improving outcomes in neonates with malignant tumors.

Contributors: VR, AJ: clinical management and drafting of report; SS: pathological examination and drafting of the report; AR: surgical management and drafting of the report.
Funding: None; *Competing interests*: None stated.

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