

Case Report

Non-rhabdomyosarcoma soft tissue sarcoma in a neonate, a rare and aggressive disease: case report

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ABSTRACT

Neonatal soft tissue sarcomas are a rare group of tumors. The behavior and aggressiveness of neonatal STS is variable. Surgical excision has been noted to be most important factor affecting outcome. However, when non-mutilating surgery is not possible, or resection is incomplete, chemotherapy and radiotherapy have been tried with variable success. We encountered a case of a neonate having large soft tissue mass at anterior abdominal wall. Biopsy and immunohistochemistry confirmed it as undifferentiated soft tissue sarcoma. Surgical excision was incomplete with margin positivity. His disease showed recurrence in short duration of time and despite adjuvant chemotherapy, progression was noted. The child abandoned the treatment and died within 3 months of life.

Keywords: Abdominal wall tumor, Neonatal soft tissue sarcoma, Neonatal cancer, Non-rhabdomyosarcoma soft tissue sarcoma

INTRODUCTION

Malignancy in first month of life is a rare ailment. Neonatal soft tissue sarcoma is the third common cancer of this age group. While neuroblastoma and teratoma are more frequently reported, soft tissue sarcoma (STS) is a rarer entity.¹ More than 75% of soft tissue tumors diagnosed in infancy are pathologically benign. They may include infantile hemangioendothelioma, lymphangioma or infantile myofibromatosis. 10% of tumors are borderline lesions and only 15% are malignant. The most common diagnosis among malignant lesions during this age group is rhabdomyosarcoma, peripheral primitive neuroectodermal tumor and undifferentiated sarcomas.² Due to sparse incidences and paucity of pooled data, etiopathogenesis, behavior and treatment guideline for neonatal STS is lacking. The prognosis for such tumors is seen to depend on histology. The management remains a

challenge due to the immaturity of physiological systems and thus constraints of therapy. The risk of long-term sequelae always is a concern in the therapy of this age group. Undifferentiated sarcomas are aggressive tumors with poor outcomes. We report a case of neonatal undifferentiated sarcoma.

CASE REPORT

S, full term, male, appropriate for gestational age (3200g), born to an unbooked mother. Antenatal records were not available. Perinatal events were unremarkable. At birth, a soft tissue mass was noticed in the right iliac fossa. No other phenotypic abnormality was noted. CECT Abdomen was done which was s/o a large solid heterogeneously enhancing mass [7.3 x 6.6 x 6.5cm] seen arising from the soft tissue planes of the lower anterior abdominal wall on right side extending inferiorly up to the pubic region and superiorly up to the periumbilical

region (Figure 1) Chest scan showed no abnormal shadow. Surgical excision of the mass was done on day 4 of life. Excision was with positive margins. Histopathology and immunohistochemistry confirmed the diagnosis of High-grade undifferentiated sarcoma (Vimentin positive, S-100 few dispersed cells positive and Ki-67 - 50-60%).

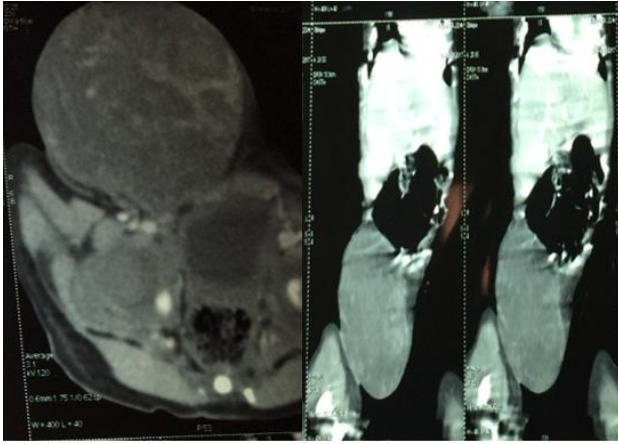


Figure 1: CECT Abdomen showing a large soft tissue mass in the pelvis.

Post-operative CT imaging showed no gross disease. However, in view of margin positivity and high-grade tumor, adjuvant therapy with Vincristine and Actinomycin D was suggested after complete recovery and suture healing. He was admitted on day 18 of life with progressively increasing swelling at right side of lower abdominal wall (Figure 2) Ultrasound evaluation showed soft tissue mass. A recurrence of disease was confirmed with fine needle aspiration cytology. He received chemotherapy, Inj. Vincristine 0.5 mg/kg, Actinomycin-D 0.045 mg/kg and Inj. Cyclophosphamide 30 mg/kg. He tolerated the chemotherapy well and was discharged in a stable condition. A decrease in the size of the swelling was observed post chemotherapy. However, on day 15 post chemotherapy, swelling size started to increase. Family opted for alternative treatment after this. The child continued to have progressive disease and succumbed to the illness at the age of 3 months.



Figure 2 : Recurrence of mass on the opposite side of scar.

DISCUSSION

Neonatal malignancies are heterogeneous in histology and variable in natural history. While almost all histology can be seen in this age group, drawing a parallel from older counterpart is over simplification. STS account for 7.4% of all childhood cancers, the incidence being 11 cases/million population.³ They are essentially divided into Rhabdomyosarcoma (RMS) and Non-Rhabdomyosarcoma Soft Tissue sarcomas (NRSTS). The NRSTS group is less common than RMS group. It is rarely found among neonates but when present is usually an aggressive disease. Neonatal STS is different from STS affecting older child or adult. Etiopathogenesis is less clear for this subset and probably pre-conceptual and transplacental oncogenesis has an effect on it. Exact genetic fault and mutations are yet to be defined.⁴ While for neonatal cancers, congenital abnormalities are common associations but in a series of neonatal malignancy from France, only 4.5% cases were found to have association. That study noted STS in 8% of all neonatal cancer cases.³ There is no separate large data for such entity to support or refute any specific abnormality with neonatal STS.

Neonatal STS has been classified into three broad categories- benign group, intermediate group and malignant or aggressive group. While malignant category is rare, undifferentiated sarcoma from abdominal wall has not been reported.⁴ Rhabdomyosarcoma from same anatomical location has been reported. Fibrosarcoma of neonatal age group is the most common malignant STS reported.⁵ Index case represented a rare and aggressive STS.

Treatment modality of Neonatal STS has not been standardized due to paucity of data. However, cohort from France noted that surgical excision remains the most important prognostic factor.³ From fibrosarcoma experience, an attempt to downsize the non-operable tumor should be done.^{5,6} Study noted effective cytoreduction with vincristine and actinomycin-D chemotherapy. Index case underwent upfront excision, although resection margins were positive, no gross residual disease was left. The re-appearance of tumor at opposite side abdominal wall showed aggressiveness of tumor. Adjuvant therapy with effective agents were administered but had only a temporary effect on tumor size. Literature search showed no clear guideline of choosing second line therapy for such aggressive and rare tumor. A multidisciplinary team is warranted for optimal management including neonatology, pediatric oncology and pediatric surgery.⁷

Antenatal detection of such malignancy is possible with antenatal ultrasonography.³ But as neonatal tumors have variable natural history, its impact on pregnancy or treatment decision is not predictable. Histological typing is necessary to predict outcome. Grace period or self-subsiding phenomena of neonatal tumors seen in many

histology, has not been reported with undifferentiated STS.^{5,8,9} Neonatal sarcomas are a small proportion of all neonatal tumours. Most are benign but aggressive, nonetheless.¹⁰

The outcome of the aggressive neonatal soft tissue sarcomas not amenable to complete excision is dismal. Such cases should be pooled for analysis and treatment.

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