

A 3-days old infant with neonatal soft tissue sarcoma: a case report



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ABSTRACT

Background: The incidence of sarcomas occurs in less than 1% of all malignant tumors but sarcoma is the cause of 2% of total cancer deaths. Based on the National Registry the incidence of neonatal sarcomas is 2.6 per million live births.

Case Description: A baby girl aged 3 days, complaining of a mass on the patient's left thigh since birth. The mass was large from birth and there was only one on the left thigh. In the prenatal history, the mother and baby with routine ANC history to the midwife, and never having a control with an obstetrician, the mother was not sick during pregnancy and took medication during pregnancy outside of a doctor's prescription. In the perinatal history, the baby was born to mother G1P1A0, gestational age at term, was born by cesarean section, with birth weight 2800 grams, was born crying, active movement, reddish color, vomiting (-) genitalia (+) anus (+)

Conclusion : Soft tissue sarcomas (STS) require proper diagnosis and management. The results of rhomboid flap in the tumor resection area are better than skin grafts of similar shape and location and provide cosmetic advantages with very few complications.

Keywords: Soft tissue sarcoma, soft tissue sarcoma in neonatal , rhomboid flap.

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INTRODUCTION

Soft tissue sarcomas (STS) are a very heterogeneous group of extraskeletal nonepithelial malignancies, classified on the basis of histology according to the tissue predilection.¹⁻³ In the National Registry the incidence of neonatal sarcomas is 2.6 per million live births. Sarcomas account for 9.4% of all neonatal cancers in Asia.⁴

Soft tissue tumors in neonates are relatively common, these tumors consist of heterogeneous tumor types. STS in neonates raises concern for parents and diagnostic difficulties for health professionals. However, a small number of these lesions are found to be malignant and can present challenges in terms of management and poor prognosis. It is therefore important to develop an approach for the diagnosis and management of all soft tissue tumors so that they can be distinguished from benign STS which are

usually more common.⁵

Tumors in this age group have unique biological characteristics, the effects of long-term radiation therapy and difficulty in regulating the correct dose of chemotherapy agents, posing challenges to the management of STS in this age group.⁶

CASE REPORT

A baby girl aged 3 days, complaining of a lump on the patient's left thigh since birth. The lump is large from birth and there is only one fruit on the left thigh. In the perinatal history, the baby was born to mother G1P1A0, gestational age at term, was born by cesarean section, with a birth weight of 2800 grams. History of routine ANC, the mother was not sick during pregnancy and took medication during pregnancy outside of a doctor's prescription was denied. The patient's father and mother had a history of

previous malignancy. The patient was then taken to the Dr. Kariadi Semarang General Hospital.

On physical examination, the general condition was good, vital signs were within normal limits. Generalist status examination found no abnormalities. On examination of localist status on the left thigh, there was a lump with a firm border, blackish red, uneven surface, hard consistency, with a subcutis base, bumpy surface, uneven edges and firm borders.

On the macroscopic examination of anatomical pathology, it was found that the pieces of tissue had been split, weighing 132.6 grams, measuring 7 x 6x 5 cm, on the white, brown, solid, chewy pieces. Microscopically, a tumor in the left femur region shows spindle cells in the fibrovascular stroma with multiple mitosis according to the malignant soft tissue sarcoma.

The patient underwent an excision operation on the tumor followed by a rhomboid flap, and in making this case report, informed consent was made to the family.

DISCUSSION

Soft tissue tumors in neonates are relatively common, these tumors consist of heterogeneous tumor types. Epidemiology in this age group: rhabdomyosarcoma represented a third of all STSs (32.8%),



Figure 1. Clinical photo of a 3 day old baby with soft tissue sarcoma.

followed by infantile fibrosarcoma (24.5%) and malignant rhabdoid tumors (14.2%). In the SEER 17 database (1973-2007 period), only 47 cases of STS diagnosed at <1 month of age were registered: 18 cases of rhabdomyosarcoma, 12 cases of fibrosarcoma, 4 cases of malignant rhabdoid tumor, 3 cases of hemangiopericytoma, 10 other (or unidentifiable) cases.⁷

Collin et al,⁸ stated in their research that malignant soft tissue tumors, after neuroblastoma, are the second most common cause of cancer in neonates. Infantile fibrosarcoma (IF) is a rare tumor that most commonly affects the extremities of children 4 years of age or younger. Repeated t (12; 15) (p13; q25) fusion combining the ETV6 gene with the NTRK3 neurotrophin-3 receptor gene was identified on IF.

Based on an epidemiological study by Palumbo et al,⁹ the incidence of cancer in the United States occurring within the first 29 days of life is 36.5 per million live births. Half are diagnosed during the first day of life and two thirds in it the first week. The incidence of cancer in the first month of life is approximately 3.5 times more common than other types of disease.

Neonatal soft tissue sarcomas are divided into three histologically distinct subgroups; rhabdomyosarcoma represents more than one third of cases of soft tissue tumors with other cases being non-rhabdomyosarcoma and congenital infantile fibrosarcoma. The United Kingdom National Registry of Childhood

Tumours for babies born 1988–2007 and diagnosed within the first 4 weeks of life reflects this distribution. In the National Registry the incidence of neonatal sarcoma is 2.6 per million live births. Sarcomas account for 9.4% of all neonatal cancers in Asia.⁴

Soft tissue tumors can be diagnosed prenatally as an incidental finding on ultrasound examination. Very large tumors may prevent labor and a caesarean section is required, and frequent ultrasound monitoring in the last trimester is required. Soft tissue sarcomas are a heterogeneous group of tumors with more than 70 sub-histological types of sarcomas, so it is necessary to do a histological examination and biopsy. Recent molecular studies into detectable cytogenetic disorders that can be detected almost routinely show a variety of abnormalities including chromosomal translocation, inversion, amplification and gene rearrangement.⁷

Imaging studies should be discussed with a pediatric radiologist. Ultrasound imaging or MRI may be sufficient, avoid ionizing radiation. Plain radiographs may be indicated but tomography scanning should be avoided (due to high radiation doses) except in certain circumstances such as the need for imaging of lung pathology. Imaging can assist in the diagnosis or resectability assessment.¹⁰

The general principles of surgical management include complete wide excision of the primary tumor and excluding the surrounding margins while maintaining function. However, the operative strategy needs to be confirmed with a histologic diagnosis because some types of tumors may require only a more conservative surgical approach or may require chemotherapy before any definitive surgical procedures. is the right type of resection.¹²

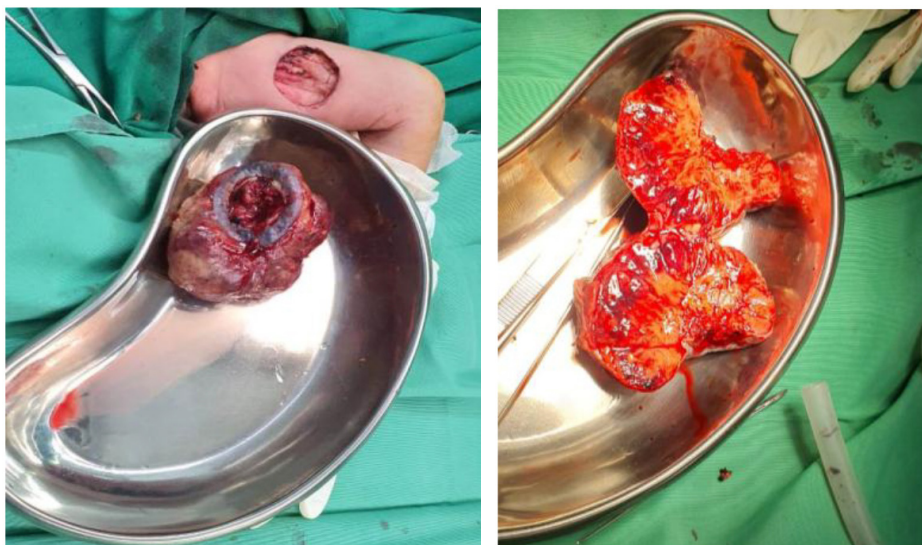


Figure 2. Tumor excision



Figure 3. Rhomboid flap

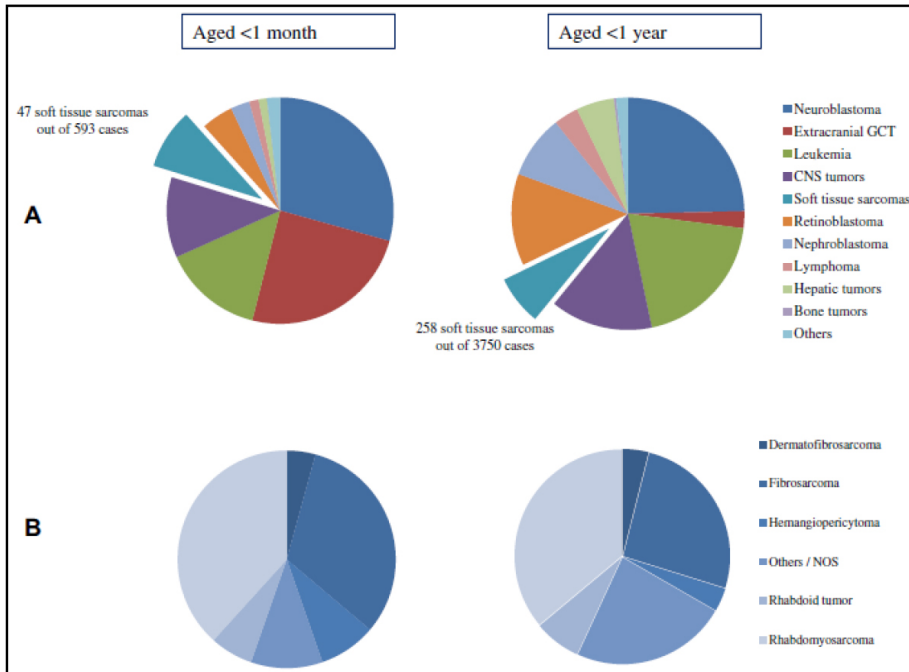


Figure 4. Number of cases registered in the public access database surveillance, Epidemiology, and End Results (SEER) 17 (1973-2007 period): (A) all tumor types, <1 month and <1 year old, respectively ; (B) all soft tissue sarcoma subtypes. GCT, germ cell tumor; CNS, central nervous system.⁷

Postoperative adjuvant radiation therapy can be applied to the area of the graft without fear of wound complications, especially if the graft is allowed to heal adequately and if chemotherapy is not given at the same time as radiation. Neo-adjuvant chemotherapy. The reasons for administering neo-adjuvant chemotherapy for advanced local STS include: (i) tumor reduction to compensate for less extensive local surgery; (ii) early treatment of metastases; and (iii) in vivo evaluation of tumor chemosensitivity.

In soft tissue sarcomas, the degree of malignancy affects the likelihood and speed of recurrence. A risk assessment based on grade, size, and location of the tumor will determine the follow-up policy. Patients with high risk will generally have a recurrence within 2 to 3 years whereas patients with low risk will have a relapse with a longer time, although it is unlikely. Recurrence is often metastasis to the lungs. Early detection of local recurrence and metastases in the lung is prognostic.¹²

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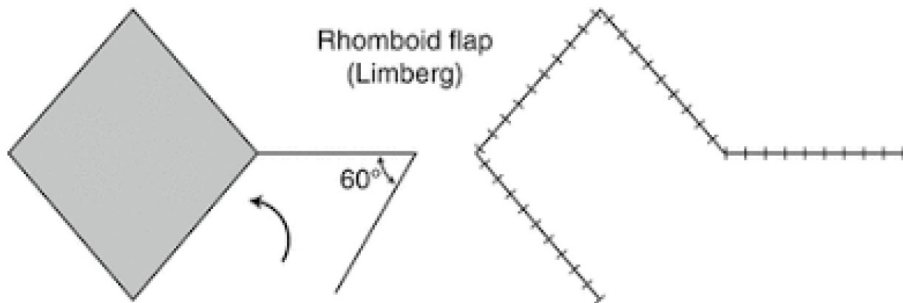


Figure 5. Basic rhomboid flap construction.

Extensive surgical closure may be accompanied by a transposition flap. The transposition flap is one of the complex random flap pattern designs. The technique that is often used is the Rhomboid flap. Rhomboid flaps can be performed for small to large defects in various anatomical locations. Rhomboid or Limberg flap is a flap design that is often used to cover defects in skin cancer.¹³

Rhomboid flap is one of the transpositional flap designs that is made in the shape of a rhombus with an angle of 120° from the proximal to the skin defect. Rhomboid flap results were reported to be better than skin grafts of similar shape and

location. This is because the sub-papillary and sub-dermal vascular plexuses are retained on a flap that is not found on grafts. The simplicity and efficacy of this flap makes it a cosmetically superior flap with very few complications.^{14,15}

The main advantage of the Rhomboid flap is that it can direct skin tension from only one axis to the defect. Rhomboid flaps can only be performed well on areas with substantial skin laxity to allow easy closure of the secondary defect.¹³

Appropriate post flap therapy is required to reduce the risk of wound complications which are a significant problem in this age population.

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