

Congenital Rhabdomyosarcoma of Shoulder

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Abstract

A 16-day-old female was referred with congenital swelling on her right shoulder. On examination, there was a hard, round, ecchymotic, nontender, slightly movable, warm and shiny 10x15 cm mass on the right axillary pits which was extended to the right side of neck and chest wall. The mass separated the shoulder from the chest wall causing paralysis of right hand. Chest X-ray, ultrasound and MRI with contrast demonstrated a soft tissue mass suspected to be a hemangioma. The mass rapidly increased in size despite aggressive steroid therapy with rupture and bleeding. On the 45th post natal day the baby was taken to operating room to control the bleeding and if possible total excision of the mass. The mass was separated easily from the surrounding tissue and was excised along with right upper extremity. At the end of surgery the baby had cardiac arrest, and apparently died of Disseminated Intravascular Coagulation (DIC). The final pathology report was Rhabdomyosarcoma (RMS).

Keywords: Rhabdomyosarcoma; Congenital; Newborn; Shoulder

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Introduction

Rhabdomyosarcoma (RMS) is the most common soft tissue malignancy in childhood. RMS can occur at any age; the peak incidence is between 2 and 6 years of age, and the second peak occurs at ages 14-18 years old [1] but extremely rare it may occur in the neonatal period [2, 3]. There are only a few reports for RMS in neonates [3, 4]. RMS may be present at birth: approximately 5-10% of all cases occur in patients aged <1 year, and 1-2% of all cases are congenital [5]. It may arise anywhere in the body, but it has a predilection for the head and neck area, genitourinary tract, and the extremities [3]. Chest wall is a rare site for RMS [3, 6]. Cooperative multidisciplinary treatment protocols have substantially improved the outcome of children with RMS over the past 25 years. Currently, patients with localized disease can be cured in >70% of cases [1, 7], nonetheless, the small percentage of patients aged <1 year are particularly difficult to manage.

The well-known physiologic immaturity of various organs is responsible for the vulnerability of infants to acute and late effects of therapy, and functional immaturity of liver leads to a different metabolism of drugs in infants as compared with older patients. Infants with RMS require tailored treatments, the use

of radiotherapy is restricted due to very high risk of side effects, and the morbidity of full-dose chemotherapy is unacceptable. The important points concerning treatment modality in infants have yet to be clarified completely. Various reports indicate that the outcomes for infants are the same as older children and are not affected by dose reduction [7,8] but more recent data suggest a less favorable outcome for patients aged <1 year [9].

Case Presentation

A 16-day-old female was referred with congenital swelling on her right shoulder. Patient was the 3rd born child to a healthy mother at 38th weeks of gestation by normal vaginal delivery followed by dystochia. On examination, there was a hard, round, ecchymotic, nontender, slightly movable, warm and shiny 10x15 cm mass on her right axillary pits that was extended to the right side of the neck and chest wall (Figure 1). The mass separated the shoulder from the chest wall and there was a paralysis of right hand. Chest X-Ray showed a round soft tissue shadow. Ultrasound showed a huge, heterogenic hypoechogenic mass 20 cm in size and enhanced vascular pattern and suspected to be a hemangioma. MRI with contrast and CT-angiography demonstrated a soft tissue mass suspected to be a

hemangioma and suggested the possibility of RMS or neurofibroma (Figures 2, 3). All laboratory profiles were normal.

The patient was treated with corticosteroid. The mass rapidly increased in size along with rupture and bleeding despite aggressive steroid therapy. On the 45th post natal day neonate was taken to operating room to control the bleeding and if possible total excision of the mass. The mass separated easily from the surrounding tissue and was excised along with right upper extremity amputation. Pathologic evaluation showed RMS in this patient which was confirmed by Immunohistochemistry (IHC) results (Myo-D: positive, CD99: negative). The subtype of tumor cannot be definitely identified according to histological features. Molecular biology (PAX11 and PAX7) is needed for definite diagnosis (Figure 4).

At the end of surgery the baby developed cardiac arrest and died due to DIC.

Discussion

Only 2% of childhood tumors occur in the first month of life, of which 20% are mesenchymal in origin [10]. The incidence of soft tissue sarcoma is believed to be approximately 8 in 1,000,000 and more than half of these are rhabdomyosarcoma [11]. There are only a few reports about neonatal RMS in the literature [4]. Lobe TE, et al. reported only 4% of neonatal RMS in a large series of involved RMS patients [10]. RMS is traditionally subdivided into embryonal, alveolar and pleomorphic. Embryonal RMS is the most common type (60-70%), and also is the most predominant in neonates, infants and young children [3]. RMS usually occurs in the head and neck



Figure 1. Patient with right axillary mass that was extended to the right side of the neck and chest wall before operation

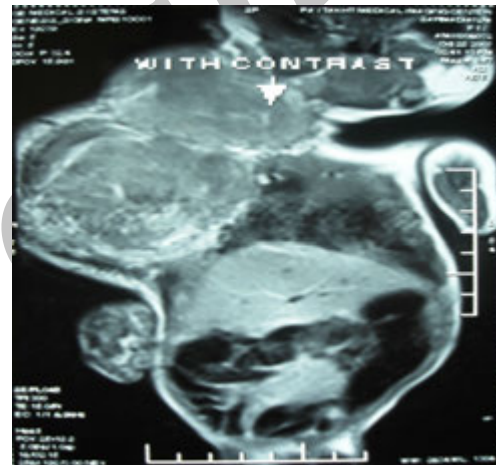


Figure 2. MRI with contrast that confirmed huge soft tissue mass of right side chest and neck



Figure 3. CT-angiography that showed hypervascular soft tissue mass of right neck, axilla and chest wall

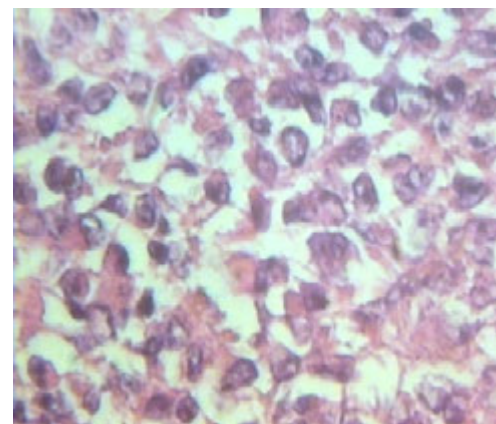


Figure 4. Histopathology which confirmed RMS in our patient

region, and approximately 20% in extremities [3, 12].

It has been noted that in congenital RMS, the disease may be metastatic at birth (organs and placenta too) [3]. MRI is the primary imaging modality in RMS with its superior ability to depict soft-tissue changes [3]. CT-scan of chest is mandatory in order to assess pulmonary metastases [13]. In our case CT-scan of chest and upper abdomen was done, which suggested the possibility of RMS or neurofibroma. The clinical management of malignancies in patients aged < 1 year is particularly difficult and requires special therapy modifications and careful monitoring to prevent severe complications [7]. Treatment of neonatal RMS requires a multidisciplinary approach, where surgery and chemotherapy both have their own specific roles. Complete resection of tumor is recommended; embryonal RMS generally responds to chemotherapy very well, and a long-term follow-up is necessary to evaluate the outcome of treatment. A good response to chemotherapy allows surgery less aggressively if needed, and the use of radiotherapy is restricted due to very high risk of side effects, and it should be avoided in the newborns [3]. Therefore, pediatric oncologists, radiologists, pediatric surgeons and pathologists play very important roles [3]. The overall survival rates for patients under 1 year of age and patients over 1 year of age did not differ significantly [10].

Conclusion

RMS is the most common soft tissue sarcoma in childhood. Only 2% of them present at birth. Huge congenital RMS is very rare. The treatment for neonatal RMS requires a multidisciplinary approach where surgery and chemotherapy both have their own specific roles. Surgical option is radical or partial resection, and chemotherapy is recommended in all cases, but radiotherapy is necessary when the tumor cannot be completely removed. When the tumor size is huge we should firstly consider biopsy and chemotherapy followed by a surgery.

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Conflict of Interest

The authors have no conflict of interest in this article.

Authors' Contribution

Ahmad Khaleghnejad Tabari and Alireza Mirshemirani contributed to surgical performance and prepared the manuscript. Atoosa Gharib carried out pathologic evaluation. Shahin Nariman and Nasibeh Khaleghnejad Tabari carried out literature review. Mohsen Rouzrokh contributed to study design. Shaghayegh Hassas Yeganeh monitored the case and analyzed the data.

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