

## An unusual localization of embryonal rhabdomyosarcoma in a neonate

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Rhabdomyosarcoma (RMS) is a common, highly malignant, uniformly fatal childhood malignancy, which presents extremely rarely in the neonatal period; there are only a few reports about this tumor in this age group. While the primary tumor may arise virtually anywhere in the body, the extremity, orbit and genitourinary region are the most frequent sites; the retromammary region is extremely rare. Herein, we report a neonate with embryonal RMS arising from the anterior chest wall musculature at birth.

*Key words:* rhabdomyosarcoma, newborn.

Rhabdomyosarcoma (RMS), a malignant tumor of immature mesenchymal cell origin, is the most common soft tissue sarcoma in children<sup>1,2</sup>, but this type of tumor is extremely rare in the neonatal period, constituting only 1-2% of childhood RMS<sup>2,3</sup>. Of more than three thousand patients registered in the Intergroup Rhabdomyosarcoma Study (IRS) I-IV, only 14 were in the neonatal period at the time of diagnosis<sup>4</sup>. In the first three IRS trials, approximately 35% to 40% of all tumors arose from the head or neck region, slightly fewer than 25% from the genitourinary tract, approximately 20% from an extremity, 10% from truncal primary tumors and the remaining 10% from other miscellaneous sites<sup>5</sup>. Because primary congenital RMS in the retromammary region has not been reported in the literature, we report such a neonate to emphasize the malignant perspective of the anterior chest wall masses in newborns.

### Case Report

A one-day-old female baby was admitted with a swelling on her right breast. She was born to a G1, 31-year-old healthy mother at 40 weeks gestation by cesarean section with a birth weight of 3500 g. There was no history of nipple discharge or redness. Physical examination revealed a firm, round, movable

and nontender mass measuring 7x4 cm in the right breast region. The mass extending to the axillary region was fixed to the chest wall at the median border (Fig. 1). Contralateral breast was normal. No palpable axillary or supraclavicular lymph nodes were found. The general physical examination was unremarkable. Ultrasound revealed a soft tissue mass with heterogeneous, vascularizing areas that did not originate from the breast tissue; there was no calcification. On day 4, magnetic resonance imaging (MRI) examination was performed and showed a mass 6.5x3.5 cm in diameter, localized in the retromammary region, with a possible diagnosis of congenital fibrosarcoma or RMS (Fig. 2). At 12 days of age, total excision of the mass measuring 7x4x3 cm was performed (Fig. 3). Pathological diagnosis was embryonal RMS with high grade. The parents of our patient did not accept treatment in spite of our detailed explanation, and she was discharged from the hospital. No information about the newborn could be obtained by telephone.

### Discussion

Two unusual features were present in our case. First, the patient was a neonate. RMS is the most frequent soft-tissue sarcoma seen in the pediatric age group, accounting for approximately 4% of all childhood



Fig. 1. Photograph of the patient on admission.



Fig. 2. Oblique sagittal postcontrast T1-weighted image shows enhancing mass lesion in the retromammary region.

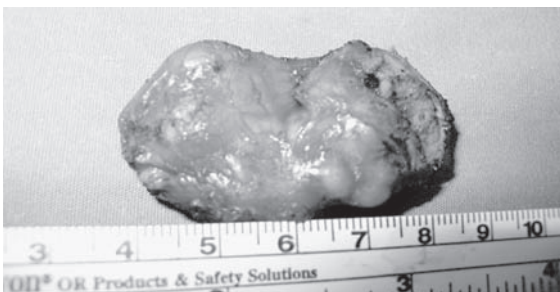


Fig. 3. Postoperative photograph of the mass.

maligancies. However, occurrence of this tumor in the neonatal period is extremely rare: approximately 1-2% of all cases are congenital<sup>2</sup>. Second, it was located in the retromammary region. RMS may arise from any part of the body where the skeletal muscle is present, including head, neck, genitourinary tract, extremities, and trunk, but primary retromammary region is extremely rare. There are only a few reports about neonatal RMS in the literature<sup>6,7</sup>. Buyukpamukcu et al.<sup>6</sup> reported three patients with RMS located in the orbit, vagina and left tibia, all of whom died. The differential diagnosis of RMS located in the retromammary region includes other oncologic and non-oncologic conditions such as primary breast RMS, congenital fibrosarcoma, infantile myofibromatosis, and abscess. MRI has been frequently used for differential diagnosis. RMS was also in the form of a palpable breast mass suggesting a primary breast tumor in our patient. However, ultrasonographic and MRI studies showed that it was located in retromammary region and originated from the intercostal muscle of her anterior chest wall. Total excision was performed for a definitive diagnosis and treatment. Histopathological examination revealed an embryonal RMS. Location of the primary lesions, age of the patient, metastatic status and histopathologic subtype are important prognostic factors for RMS. Data from the IRS-II study showed that subjects with nonmetastatic primary lesions at favorable sites, including non-parameningeal regions of the head and neck, and the genitourinary system, had long survival rates, while lesions at less favorable sites, such as the parameningeal areas, extremities and trunk, were associated with short survival rates<sup>8</sup>. Nearly all patients diagnosed in the neonatal period and reported in the literature died. The authors have mentioned that the clinical management of malignancies during the first year of life is particularly difficult and the use of radiotherapy is restricted by the very high risk of side effects. So, age less than one year has emerged as an independent adverse prognostic factor for RMS<sup>9,10</sup>. Unfortunately our patient's embryonal RMS had originated from an uncommon location and begun at a very early age, both of which suggest poor prognosis for long-term survival. Due to lack of parental consent, we could not manage or follow-up our patient.

In conclusion, embryonal RMS may occur even in utero. When a baby is born with a palpable hard mass on her/his breast region, embryonal RMS should be kept in mind in the differential diagnosis.

#### REFERENCES

1. Pappo AS, Lyden E, Breneman J, et al. Up-front window trial of topotecan in previously untreated children and adolescents with metastatic rhabdomyosarcoma: an intergroup rhabdomyosarcoma study. *J Clin Oncol* 2001; 19: 213-219.
2. Weiss SW, Goldblum JR. Rhabdomyosarcoma. In: Weiss SW, Goldblum JR (eds). *Enzinger and Weiss's Soft Tissue Tumors* (4<sup>th</sup> ed). St Louis: CV Mosby; 2001: 785-835.
3. Ragap AH, Heyn R, Tefft M, et al. Infants younger than 1 year of age with rhabdomyosarcoma. *Cancer* 1986; 58: 2606-2610.
4. Lobe TE, Wiener ES, Hays DM, et al. Neonatal rhabdomyosarcoma: the IRS experience. *J Pediatr Surg* 1994; 29: 1167-1170.
5. Wexler LH, Crist WM, Helman LJ. Rhabdomyosarcoma and the undifferentiated sarcomas. In: Pizzo PA, Poplack DG (eds). *Principles and Practice of Pediatric Oncology* (4<sup>th</sup> ed). Philadelphia: Lippincott Williams Wilkins; 2002: 940-971.
6. Buyukpamukcu M, Varan A, Tanyel C, et al. Solid tumors in the neonatal period. *Clin Pediatr* 2003; 42: 29-34.
7. Isaacs H Jr. Congenital and neonatal malignant tumors: a 28-year experience at Children's Hospital of Los Angeles. *Am J Pediatr Hematol Oncol* 1987; 9: 121-129.
8. Lawrence W Jr, Anderson JR, Gehan EA, Maurer H. Pretreatment TNM staging of childhood rhabdomyosarcoma: a report of the Intergroup Rhabdomyosarcoma Study Group. *Cancer* 1997; 80: 1165-1670.
9. Dillon PW, Whalen TV, Azizkhan RG, et al. Neonatal soft tissue sarcomas: the influence of pathology on treatment and survival. *Children's Cancer Group Surgical Committee. J Pediatr Surg* 1995; 30: 1038-1041.
10. Lobe TE, Wiener ES, Hays DM, et al. Neonatal rhabdomyosarcoma: the IRS experience. *J Pediatr Surg* 1994; 29: 1167-1170.