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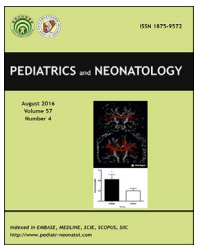

Letter to the Editor

Multidisciplinary management of a malignant rhabdoid tumor of the neck and mediastinum in an infant

Dear Editor:

Malignant rhabdoid tumor (MRT) is a highly aggressive tumor associated with an abysmal prognosis.[1] Here, we describe the case of a 9-month-old boy who presented with MRT in the anterior mediastinum.

This boy had flu-like symptoms, and he was brought to our emergency department because of progressive dyspnea, anorexia, vomiting, and decreased urine output. Chest X-ray imaging showed increased infiltration that led to his admission and treatment for suspicious pneumonia. However, computed tomography (CT) showed a multiseptated cystic lesion in the anterior mediastinum, which extended to the left lower neck, pushing the heart and airway to the right side (Fig. S1A). Because of severe respiratory distress and impaired consciousness, he was transferred to the intensive care unit for intubation. Surgical intervention was performed the next day, and a 15 × 10 × 8-cm anterior mediastinal tumor with feeding arteries and venous return systems was identified (Fig. S1B). However, the tumor could not be entirely resected because of the innominate vein and phrenic nerve involvement.

The histological examination demonstrated infiltrative and solid sheet-like proliferation of malignant epithelioid tumor cells with rhabdoid features (Fig. S1C). Immunohistochemically, the tumor cells were positive for cytokeratin (AE1/AE3), ERG, and BRG1 (Fig. S1D—G) and negative for INI-1, CD34, desmin, myogenin, S100, SALL-4, and glypican-3. The tumor exhibited necrosis and increased mitotic activity with thymus and lymph node encroachment. Whole-body CT showed residual enhancing soft tissue of ~12 mm in the left upper mediastinum, lateral to the great mediastinal vessels, without the brain, abdominal, or renal metastasis.

Given the aggressive characteristics of the remnant malignant tissue, six cycles of vincristine, dactinomycin, ifosfamide, and doxorubicin regimen were started, followed by maintenance target therapy of bevacizumab. As per the literature, bevacizumab reinitiation should be delayed to at least 28 days postoperatively to avoid wound healing complications.[2] Thus, we initiated bevacizumab with 15 mg/kg administered every 3 weeks after 28 days of the debulking surgery. The latest CT images showed regression of the mediastinal mass ~1 year postoperatively. To date, the boy had received 15 cycles of bevacizumab-targeted therapy. He tolerated the regimen well, with manageable side effects and was in a stable condition. However, other side effects of bevacizumab, such as hypertension and proteinuria, were still monitored.[2]

The tumor mutation burden (TMB) is often used to predict whether a patient shows a clinical response to immune checkpoint inhibitor therapy. The mutations of germline or somatic *SMARCB1* (INI1), or rarely *SMARCA4* (BRG1), are the oncogenetic driving events of MRT. Nevertheless, MRT usually harbors a very low TMB, which might indicate a poor response to immunotherapy; however, it might not exclude the possibility of a level of PD-L1 expression that implies the clinical benefit of immunotherapy.[3] However, immature thymus glands in infants and children might undermine the efficiency of PD-1/PD-L1 inhibitors because of insufficient well-trained T cells. Although current multidisciplinary therapies, such as surgery, chemotherapy, and targeted therapy, have shown anti-tumor efficacy in this patient, long-term follow-up is still warranted to understand the effects of persistent disease.

Author contributions

J.T. study conception and design; L.T, H.S, and C.Y. data collection; W.Y, and C.T. gave conceptual advice. All authors reviewed the results and approved the final version of the manuscript.

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Ethical approval

This study has been approved by our institutional review board.

Informed consent

The oral informed consent has been recorded and transcribed. This report does not contain any personal information that could lead to the identification of the patient.

Declaration of competing interest

The authors declare no conflict of interest.

Acknowledgments

The authors thank the nursing staff who participated in this study.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.pedneo.2022.09.007.

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