Surgical Case Report

INTERNATIONAL JOURNAL OF SURGERY OPEN

OPEN

Pleural effusion revealing pulmonary pleomorphic rhabdomyosarcoma in a 2-year-old child: a rare case report

Maria Iesa, MD[a], Mouhammed Sleiay, MD[b,*], Ahed Alhariri, MD[a], Mohammad Abd Alrahman Saif, MD[c], Batoul Ahmed Juneid, MD[a], Bilal Sleiay, MD[b], Abdulrahman Ahmad Othman, MD[d], Adnan Al Abbas, MD[d], Joud Zakkour Damlakhi, MD[d], Arej Alabdo, MD[d], Ari Maray Alnajary, MD[d]

Abstract

Pleural effusion in young children is most commonly associated with infectious or inflammatory conditions. However, rare malignant causes should also be considered, particularly when symptoms persist despite standard interventions. A 2-year-old boy presented with cough, fever, and dyspnea unresponsive to initial pleural drainage. Imaging revealed a large, complex left pleural effusion with mediastinal shift. Surgical exploration uncovered a cystic lesion in the upper lobe of the left lung. Histopathological analysis confirmed pulmonary pleomorphic rhabdomyosarcoma – a rare and aggressive pediatric malignancy. The lesion was resected, and the patient's condition improved postoperatively. A regional lymph node was tumor-free. The child was referred for further oncologic care. Although extremely rare, pulmonary pleomorphic rhabdomyosarcoma should be included in the differential diagnosis of persistent pleural effusion in children. Early surgical intervention and histological confirmation are essential for timely diagnosis and initiation of appropriate therapy.

Keywords: case report, dyspnea, pleural effusion, pulmonary pleomorphic rhabdomyosarcoma

Introduction

Rhabdomyosarcoma (RMS) is a type of soft tissue sarcoma characterized by the malignant transformation of striated muscle tissue. It can also manifest in anatomical regions that typically lack normal striated muscles[1,2].

As the most prevalent soft tissue tumor in childhood, RMS accounts for approximately 6.5% of pediatric tumors and represents more than 50% of pediatric soft tissue sarcomas[3–5].

The primary sites of RMS include the head and neck region (35%), followed by the urogenital tract, limbs, and trunk[6,7].

RMS exhibits sensitivity to both radiotherapy and chemotherapy; however, a multidisciplinary treatment approach incorporating surgery, radiotherapy, and chemotherapy is essential for optimal outcomes[8–10].

Based on various factors, including patient age, tumor size, histopathological findings, and clinical staging, RMS has been categorized into three risk levels: low, intermediate, and high. This classification aims to facilitate stratified management and comprehensive treatment strategies[11–13]. The meticulous selection of treatment modalities and the implementation of multimodal therapeutic plans have significantly improved clinical outcomes and survival rates for RMS across different sites, with the current survival rate for pediatric RMS exceeding 70%[11,12].

However, primary RMS in the pulmonary region, an anatomical site generally devoid of striated muscle, has been extremely rare in reported cases[13].

In this case report, we presented a unique case of pleomorphic pulmonary RMS in a 2-year-old child manifesting as refractory massive left pleural effusion with mediastinal shift.

Case presentation

A 2-year-old child presented with a 10-day history of cough, fever, and progressive dyspnea. Initial evaluation at an outside hospital included a chest radiograph demonstrating a left-sided pleural effusion. A left chest tube drainage was performed by an outside physician; however, there was no clinical or radiological improvement. A repeat chest X-ray revealed persistence and progression of the pleural effusion.

Given the lack of response to drainage, further imaging was pursued to investigate alternative etiologies. Chest wall ultrasonography demonstrated a profuse, bloody pleural effusion with an organoid internal architecture and multiloculated components within a multilobular hematoma. These atypical features were considered inconsistent with uncomplicated parapneumonic effusion and raised concern for a hemorrhagic or neoplastic process. Subsequently, a contrast-enhanced chest computed tomography (CT) scan revealed a massive left pleural effusion causing near-complete collapse of the left lung and significant rightward mediastinal shift (Fig. 1), further supporting the suspicion of an underlying space-occupying lesion.

[a]Pediatric surgery Department, Children university Hospital, Damascus University, Damascus, Syria, [b]Faculty of Medicine, Hama University, Hama, Syria, [c]Thoracic surgery Department, Damascus university, Damascus, Syria and [d]Medizinische Fakultät der Universität des Saarlandes, Saarland, Germany

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Faculty of Medicine, Hama University, Alqosour, Hama, Syria. Tel.: +963947295398. E-mail: abdmouh1234mouhmouh@gmail.com (M. Sleiay).

International Journal of Surgery Open (2026) 64:108–112

Received 20 August 2025; Accepted 2 February 2026

Published online 13 March 2026

http://dx.doi.org/10.1097/IO9.0000000000000353

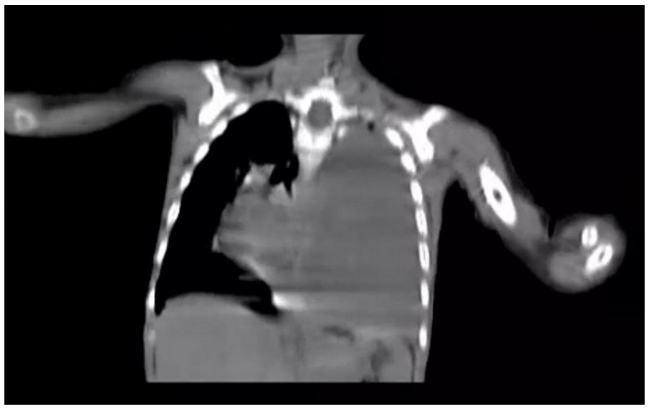

Figure 1. A chest CT scan showing a large left pleural effusion compressing and deflating the left lung, pushing mediastinal structures to the right.

Laboratory investigations showed a marked elevation of the inflammatory index, with C-reactive protein levels increased approximately 15-fold above normal values. Hemoglobin concentration and total leukocyte counts were within normal limits, arguing against severe ongoing infection or hemorrhagic shock. Although infectious causes, including bacterial pneumonia and tuberculosis, were initially considered, the absence of leukocytosis, failure of clinical improvement after adequate drainage, and the atypical radiological characteristics made an isolated infectious etiology less likely. Microbiological cultures and molecular testing did not yield pathogenic organisms.

In view of progressive respiratory compromise and imaging findings highly suggestive of a structural intrathoracic pathology, the decision was made to proceed with surgical exploration without prior pleural fluid cytology. A left thoracotomy was performed through the fifth intercostal space (Fig. 2). Intraoperatively, a cystic mass arising from the upper lobe of the left lung was identified (Fig. 3). Complete excision of the lesion, along with resection of the damaged portions of the left upper lobe, was achieved (Fig. 4). Additionally, an enlarged lymph node located in the left pulmonary hilum was excised and submitted for histopathological examination.

Histopathological analysis confirmed the diagnosis of pleomorphic RMS (Fig. 5). The resected lymph node showed no evidence of tumor infiltration. Postoperatively, the patient's clinical condition improved markedly, allowing removal of the chest tube without complications.

Following confirmation of the diagnosis, the patient was referred to pediatric hematology-oncology for further staging and management. The planned therapeutic approach included systemic multiagent chemotherapy, most commonly based on a vincristine, actinomycin D, and cyclophosphamide (VAC) regimen, in accordance with established pediatric RMS treatment protocols. Radiotherapy was not immediately planned, given the complete surgical excision and absence of nodal involvement, but remained under consideration pending multidisciplinary tumor board evaluation. Early postoperative follow-up demonstrated sustained clinical improvement, and the patient was enrolled in ongoing oncologic surveillance.

Discussion

RMS is recognized as the most prevalent soft tissue malignancy in pediatric populations, accounting for approximately 6.5% of all pediatric tumors and over 50% of pediatric soft tissue sarcomas, while constituting less than 5% of sarcomas in adults[1–4].

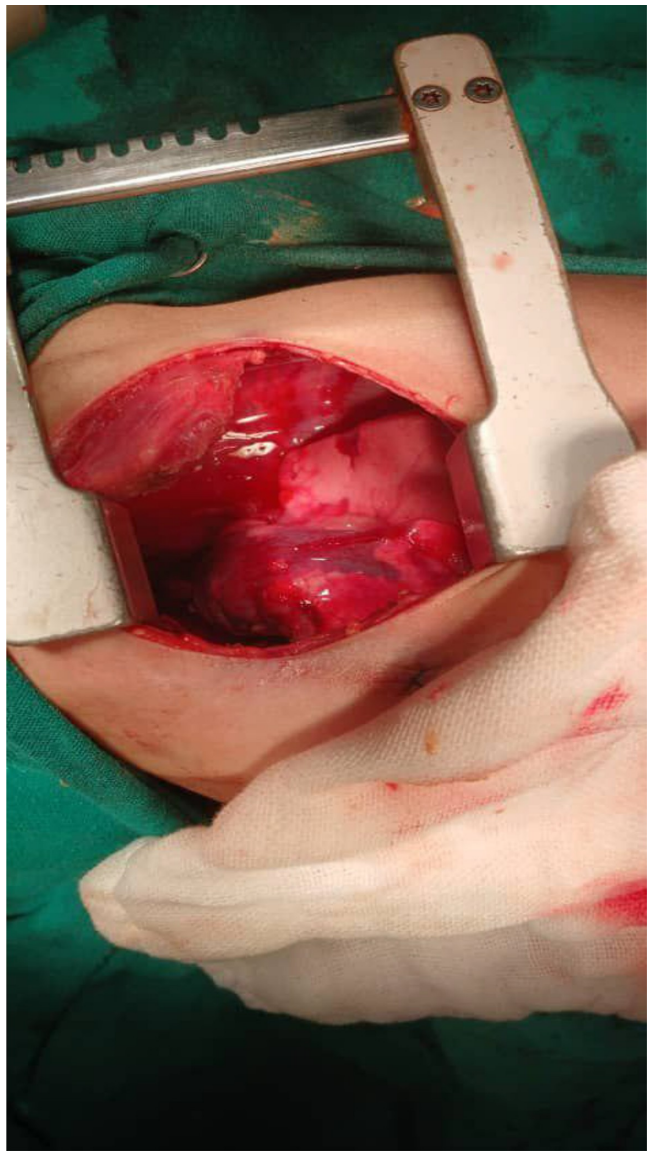

Figure 2. A surgical procedure performed via an incision located in the left thoracic region, specifically at the level of the fifth rib.

Pleomorphic RMS is an exceptionally rare entity in the pediatric population and is classically encountered in adults, in contrast to childhood RMS, which most commonly occurs between 2 and 6 years of age and is predominantly of the embryonal or alveolar subtype[3–5]. Therefore, the occurrence of pleomorphic RMS in a 2-year-old child, as observed in the present case, is highly unusual and falls outside the expected age-histology distribution. Furthermore, the absence of regional lymph node involvement or distant metastasis at diagnosis represents a favorable prognostic indicator, as localized disease that is amenable to complete surgical resection is associated with improved outcomes[2–4]. The cystic appearance of the pulmonary lesion is also atypical for pulmonary RMS, which generally presents as a solid mass[4–6], and this unusual morphology likely contributed to the initial diagnostic challenge and misinterpretation as a complicated pleural effusion.

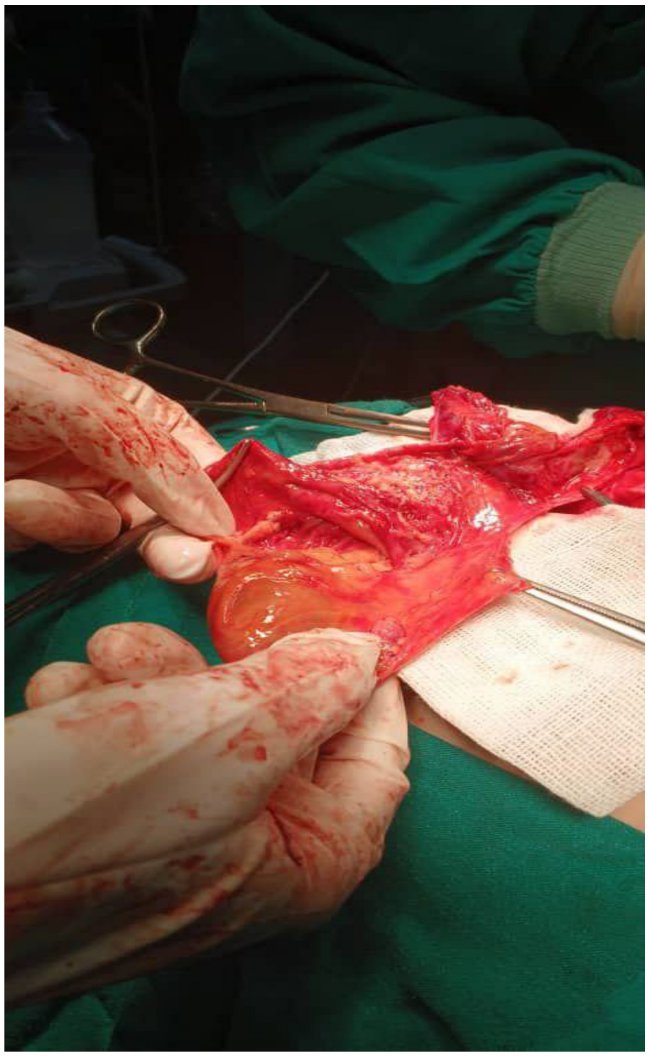

Figure 3. Atypical cystic lesion, significantly involving the upper lobe of the left lung.

Among the various presentations of RMS, primary pulmonary involvement is notably rare, with studies indicating that less than 3% of patients harbor tumors in the mediastinum, pleura, or lungs. When RMS occurs in the pulmonary system, it predominantly manifests as embryonal or alveolar variants[7]. Overall, the late presentation of symptoms, particularly in cases affecting the respiratory system, underscores the complexity of diagnosis and management in this disease[8].

The demographic analysis of the patient cohort indicated a median age of approximately 10 years, characterized by a nearly balanced male-to-female ratio. A minority of participants disclosed a history of smoking; however, a substantial proportion of pediatric cases were associated with congenital cystic adenomatoid malformation, thereby underscoring its clinical relevance within this population[1].

Upon initial diagnostic assessment, nearly all patients manifested clinical symptoms. A significant proportion presented with respiratory complications related to pulmonary masses, exhibiting symptoms such as cough, pleuritic chest pain, dyspnea, and generalized respiratory distress[8]. Moreover, instances of spontaneous pneumothorax were documented in several cases.

The diagnosis is often delayed due to the non-specific nature of early symptoms and the rarity of the tumor, which can lead to initial misattribution to more common pediatric conditions such as pneumonia or tuberculosis[1–4].

From a diagnostic standpoint, the presence of a pleural effusion necessitates thorough evaluation. Initial imaging, such as chest X-ray and CT scan, is crucial for assessing the extent of pulmonary and pleural involvement. Pleural fluid analysis, although often non-diagnostic, should include cytological examination to detect malignant cells. However, in many cases, definitive diagnosis relies on tissue biopsy, obtained via image-guided techniques or thoracoscopy. Histopathological evaluation with immunohistochemical staining is essential to confirm the diagnosis and differentiate pleomorphic RMS from other small round blue cell tumors of childhood[8–10]. Markers such as desmin, myogenin, and MyoD1 play a pivotal role in establishing myogenic differentiation[1–3].

Persistent pleural effusion in pediatric patients encompasses a wide differential diagnosis, with parapneumonic effusion or empyema representing the most common etiology and typically responding to antibiotics and adequate drainage. Tuberculous pleuritis, particularly in endemic regions, is characterized by chronic symptoms, lymphocytic exudate, and supportive microbiological or radiological findings. Congenital pulmonary malformations, such as congenital pulmonary airway malformation or bronchogenic cysts, may present with recurrent or nonresolving effusions secondary to infection or mass effect. Malignant causes, including lymphoma, often manifest with systemic symptoms, mediastinal lymphadenopathy, and recurrent effusions[3–6]. In contrast, the present case was distinguished by a bloody, multiloculated, organoid pleural effusion, lack of response to drainage, absence of typical infectious or tuberculous features, and intraoperative identification of a primary pulmonary mass, ultimately leading to the diagnosis of pleomorphic RMS.

In terms of treatment, multimodal therapy remains the cornerstone of management for RMS. This typically involves a combination of surgical resection, chemotherapy, and radiotherapy, tailored according to the tumor location, stage, and resectability. However, for primary pulmonary tumors, especially with pleural involvement, complete surgical excision may be challenging due to the anatomical constraints and the extent of disease at presentation[7–11].

Chemotherapy regimens, often based on VAC protocol, have demonstrated efficacy in RMS, though their success in pleomorphic variants remains less well-defined due to limited data. Radiotherapy is considered in non-resectable or residual disease. Despite aggressive treatment, the prognosis of pleomorphic RMS in children is generally poor, with survival outcomes significantly worse than embryonal or alveolar subtypes[8–13].

Conclusion

Pleural effusion in young children should prompt thorough investigation, as it may reveal rare malignancies such as pulmonary pleomorphic RMS. Early diagnosis and a multidisciplinary treatment approach are crucial to improve outcomes, despite the aggressive nature and poor prognosis associated with this rare pediatric tumor.

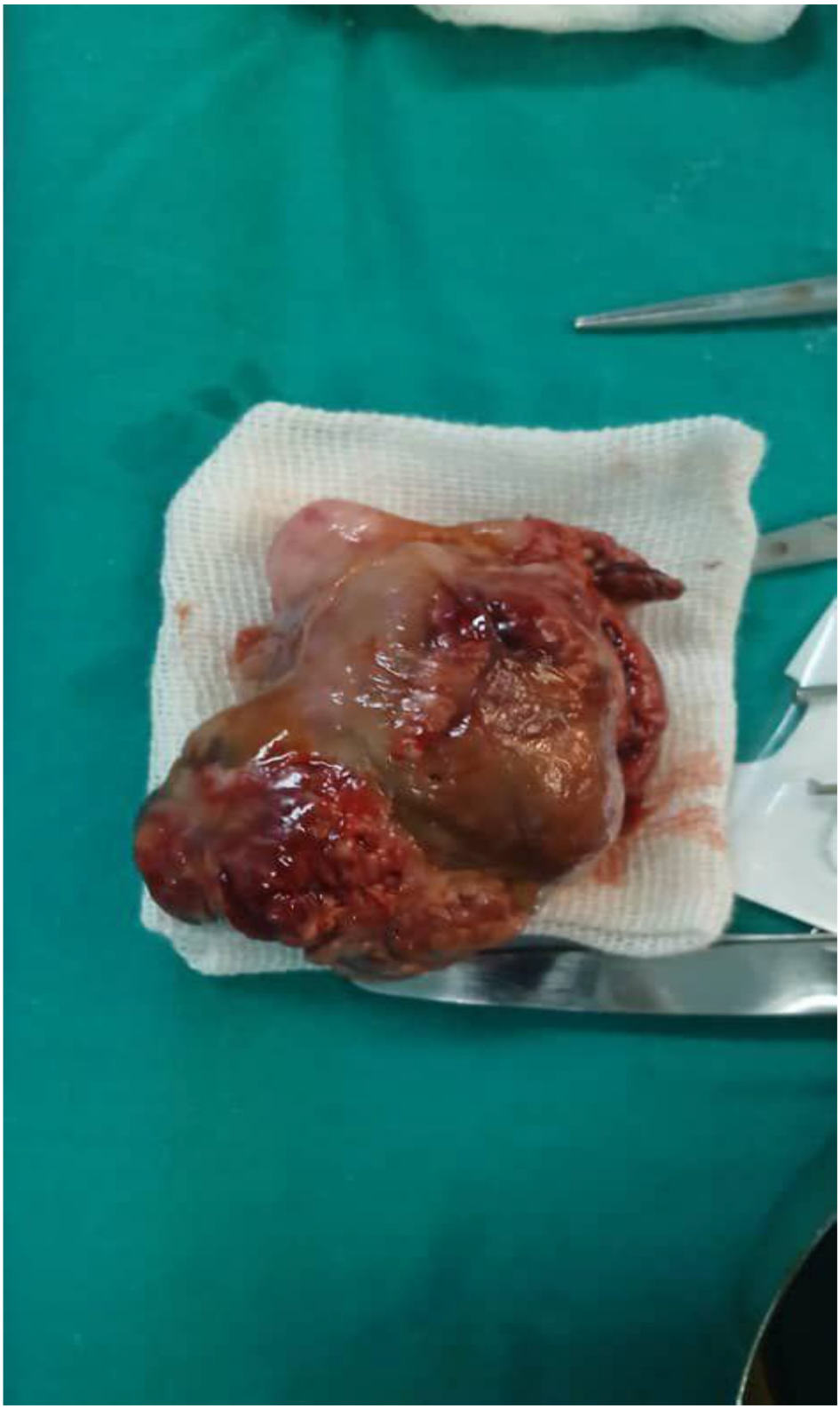

Figure 4. The excision process involving the removal of the identifiable lesion along with the compromised sections of the upper lobe of the left lung.

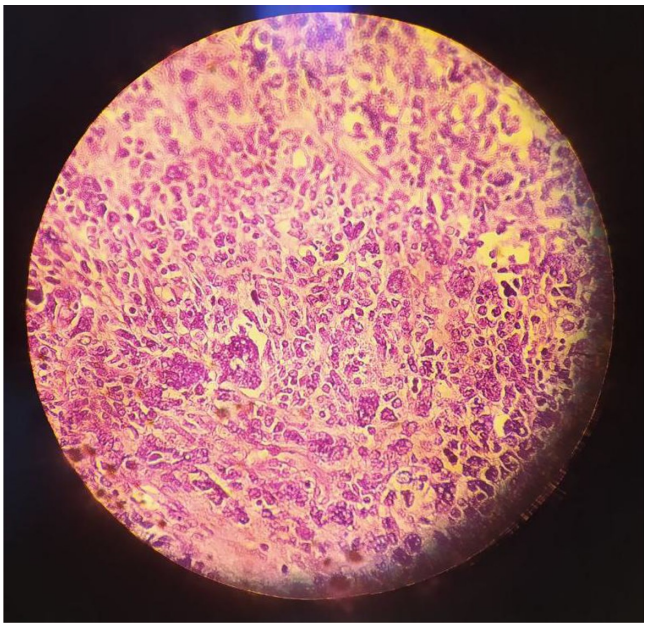

Figure 5. Pathological assessment using immunohistochemical marker (Myogenin) identified pleomorphic rhabdomyosarcoma, characterized by cellular atypia, significant pleomorphism, and increased mitotic activity.

Ethical approval

Ethical approval is not required for case reports in our institution (Hama University).

Consent

Written informed consent was obtained from the patient's legal guardian.

Sources of funding

None declared.

Author contributions

A.A.: Writing a part of the manuscript. M.S.: Writing a part of the manuscript. M.I.: Writing a part of the manuscript. B.J.: Writing a part of the manuscript. B.S.: Writing a part of the manuscript. M.Sa.: Writing a part of the manuscript. A.O.: Writing a part of the manuscript. J.Z.: Writing a part of the manuscript. A.M.: Writing a part of the manuscript. A.A.A..: Writing a part of the manuscript. A.A.: Writing a part of the manuscript. All authors approved the final manuscript.

Conflicts of interest disclosure

None declared.

Provenance and peer review

This work was not commissioned and has been externally peer-reviewed.

Research registration unique dentifying number (UIN)

Not Applicable.

Guarantor

Mouhammed Sleiay.

Methods

The work has been reported per the SCARE criteria[14].

Acknowledgements

The authors express their sincere appreciation to the SMSR Laboratory team for their dedicated efforts in bringing the research team together. The authors also extend their gratitude to Dr. Imad Eddin Alshikh Saleh for his valuable efforts in securing the necessary resources to conduct this study.

References

[1] Nguyen A, Fassas S, Freidberg N, *et al*. Recurrence and treatment of adult primary nonmetastatic bladder rhabdomyosarcoma: a systematic review. Urol Oncol 2021;39:774–80.

[2] Rudzinski ER, Kelsey A, Vokuhl C, *et al*. Pathology of childhood rhabdomyosarcoma: a consensus opinion document from the children's oncology group, European paediatric soft tissue sarcoma study group, and the cooperative weichteilsarkom studiengruppe. Pediatr Blood Cancer 2021;68:e28798.

[3] Rudzinski ER, Anderson JR, Hawkins DS, *et al*. The world health organization classification of skeletal muscle tumors in pediatric rhabdomyosarcoma: a report from the children's oncology group. Arch Pathol Lab Med 2015;139:1281–87.

[4] Sultan I, Qaddoumi I, Yaser S, *et al*. Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and end results program, 1973 to 2005: an analysis of 2,600 patients. J Clin Oncol 2009;27:3391–97.

[5] Weiss SW, Enzinger FM. Malignant fibrous histiocytoma: an analysis of 200 cases. Cancer 1978;41:2250–66.

[6] Healy JN, Borg MF. Paediatric nasopharyngeal rhabdomyosarcoma: a case series and literature review. J Med Imaging Radiat Oncol 2010; 54:388–94.

[7] Guo Y, Xie D, Yan J, *et al*. Primary pulmonary rhabdomyosarcoma with brain metastases in a child: a case report with medico-legal implications. J Forensic Leg Med 2013;20:720–23.

[8] Tao Y, Cheng W, Zhe H, *et al*. Clinical features, treatment and prognosis of primary pulmonary rhabdomyosarcoma: a systematic review. BMC Pediatr 2025;25:185.

[9] Benkhaled S, Mané M, Jungels C, *et al*. Successful treatment of synchronous chemoresistant pulmonary metastasis from pleomorphic rhabdomyosarcoma with stereotaxic body radiation therapy: a case report and a review of the literature. Cancer Treat Res Commun 2021;26:100282.

[10] Punyko JA, Mertens AC, Baker KS, *et al*. Long-term survival probabilities for childhood rhabdomyosarcoma: a population-based evaluation. Cancer 2005;103:1475–83.

[11] Dasgupta R, Fuchs J, Rodeberg D. Rhabdomyosarcoma. Semin Pediatr Surg 2016;25:276–83.

[12] Nishioka Y, Tane S, Nishio W, *et al*. A rare resected case of pulmonary rhabdomyosarcoma. Gen Thorac Cardiovasc Surg 2019;67:1089–92.

[13] Tomida A, Chiyonobu T, Tokuda S, *et al*. Pleomorphic rhabdomyosarcoma in a young adult harboring a novel germline MSH2 variant. Hum Genome Var 2022;9:8.

[14] Kerwan A, Al-Jabir A, Mathew G, *et al*. Revised Surgical CAse REport (SCARE) guideline: an update for the age of Artificial Intelligence. Prem J Sci 2025;10:100079.