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Clinical features, treatment and prognosis of primary pulmonary rhabdomyosarcoma: A systemic review

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Abstract

Background Primary pulmonary rhabdomyosarcoma (RMS), a rare soft tissue sarcoma, is characterized by a high recurrence rate and a poor prognosis. This systematic review aims to summarize the clinical characteristics, pathological features, treatment, and clinical outcomes of primary pulmonary RMS, and to analyze prognostic-related risk factors to provide robust evidence for future treatment strategies.

Methods Five databases (MEDLINE, Scopus, the Cochrane Central Register of Controlled Trials, EMBASE, and Web of Science) were searched using the relevant terms including "pulmonary", "lung", "alveolar", "ERMS", "ARMS", "RMS" and "rhabdomyosarcoma". Cases with a definitive pathological diagnosis of RMS, complete treatment information, a minimum follow-up period of at least three months, and detailed follow-up records were included. The primary study endpoints were cancer-specific survival (CSS) and progression-free survival (PFS).

Results 22 articles spanning from 1955 to 2023 met the inclusion criteria for the systematic review. The median patient age was 10.5 (2.0, 52.5) years. 22 cases (78.6%) presented with respiratory symptoms due to pulmonary masses at the initial diagnosis. 10 cases were diagnosed with the embryonal type, and the majority ($n=20$, 71.4%) underwent surgical treatment. The average PFS time was 60.9 ± 14.8 months. Patients at TNM stage IV were more prone to progression, and CSS was associated with factors including age ≥ 18 years, primary tumor size ≥ 10 cm, and non-surgical treatment. Surgery was identified as an independent factor that could shorten progression time (HR = 4.58 (1.32–15.90), $P=0.017$) and improve tumor-related survival (HR = 8.11 (1.45–45.50), $P=0.017$).

Conclusions Patients with higher TNM stages who did not undergo surgery tended to have more aggressive tumors, whereas primary tumors ≥ 10 cm and ages ≥ 18 years were associated with increased tumor-related mortality. Surgery, the primary treatment modality, independently improved CSS rates. Exploring optimal comprehensive treatment strategies that combine surgery, radiotherapy, and chemotherapy represents the principal direction for future research.

Keywords Primary pulmonary rhabdomyosarcoma, Prognostic factor, Cancer management, Lung

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Introduction

Rhabdomyosarcoma (RMS), a soft tissue sarcoma originating from the malignant transformation of striated muscle, can also occur in anatomical areas lacking normal skeletal muscle [1, 2]. As the most common soft tissue tumor in childhood, RMS accounts for approximately 6.5% of pediatric tumors and represents over 50% of pediatric soft tissue sarcomas, while it comprises less than 5% of adult sarcomas [3–5]. The primary sites of RMS include the head and neck region (35%), followed by the urogenital system, the limbs, and the trunk [6, 7].

RMS is sensitive to radiotherapy and chemotherapy; however, single-treatment modalities offer limited effectiveness. A multidisciplinary treatment approach combining surgery, radiotherapy, and chemotherapy is essential. Over the past decade, major pediatric oncology research groups, including the Intergroup Rhabdomyosarcoma Study Group (IRSG) in the United States and the European Pediatric Soft Tissue Sarcoma Study Group (EpSSG), have conducted long-term clinical studies [8–10]. Based on factors including age, tumor size, pathology, and clinical staging, these groups have categorized RMS into three risk levels: low, intermediate, and high, aiming to implement stratified management and comprehensive treatment. The precise selection of treatment modalities and implementation of multimodal treatment plans have significantly improved therapeutic outcomes and survival rates for RMS at various sites, with pediatric RMS survival now exceeding 70% [11, 12].

However, primary RMS in the pulmonary region, an anatomical site typically devoid of striated muscle, has been seldom reported in cases [13]. The profound rarity of primary pulmonary RMS has precluded the development of standardized treatment strategies, contributing to the unpredictability of its prognosis. In this study, we performed a systematic review of the literature, to summarize the clinical characteristics, pathological features, treatment modalities, and clinical outcomes of primary pulmonary RMS. Furthermore, we analyzed prognostic risk factors to provide robust, evidence-based medical insights for future treatment strategies of primary pulmonary RMS, ultimately aiming to improve prognosis and enhance survival rates.

Materials and methods

To identify relevant literature, we utilized MEDLINE (OVID), Scopus, the Cochrane Central Register of Controlled Trials, EMBASE, and Web of Science as databases, searching for the following terms, including "pulmonary", "lung", "alveolar", "ERMS", "ARMS", "RMS" and "rhabdomyosarcoma". The literature search was conducted using the following search strategy: ("pulmonary" OR "lung" OR "alveolar") AND ("rhabdomyosarcoma" OR "ERMS" OR "ARMS" OR "RMS"). We focused our search on human studies, without restricting the types of clinical research, and excluded duplicate articles. Our systematic retrieval and screening process was carried out in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. All article selection processes were double-checked and verified by two authors (Y. Tao and W. Cheng).

Cases included in the systematic review met the following criteria: a definitive pathological diagnosis of RMS obtained through tissue biopsy or autopsy; detailed records of follow-up duration, survival, and recurrence times; complete treatment information for the patient; at the initial diagnosis, sufficient medical history, clinical presentation, and imaging evidence supported that the pulmonary lesion was primary rather than metastatic; and a minimum follow-up period of no less than three months, except when the patient died due to tumor-related reasons. The final inclusion of literature relied on a comprehensive review to ensure that they met the entry criteria.

Patient information was systematically extracted from all cases, covering gender, age, underlying diseases, clinical manifestations, auxiliary examinations, histopathological classification, tumor size, initial sites of metastasis, TNM staging, initial treatment strategies (such as surgery, chemotherapy, and radiotherapy), and follow-up duration. The TNM staging was based on the IRSG staging system for RMS [14, 15].

Survival information for patients was sourced from the literature, focusing on primary study endpoints including cancer-specific survival (CSS) and progression-free survival (PFS). CSS was characterized as the proportion of patients who did not succumb to tumor-related causes, while PFS was described as survival without progression. Kaplan-Meier survival analysis was performed on CSS and PFS using the collected clinical variables, and log-rank analysis was employed to elucidate the impact of different variables on patient outcomes. Variables significant in the univariate analysis ($P < 0.1$) were incorporated into the multivariate analysis to identify independent factors affecting CSS and PFS, with a P-value of ≤ 0.05 considered statistically significant. Survival and Cox regression analyses in this study were conducted using IBM SPSS 29.0, whereas Kaplan-Meier curves were plotted using GraphPad software.

Results

Figure 1 depicts the process of our systematic literature search and screening. From MEDLINE, Scopus, Cochrane Central Register of Controlled Trials, EMBASE, and Web of Science, we retrieved 2429, 890, 157, 740 and 240 articles, respectively. After removing 493 duplicate articles and excluding 3803 articles that failed to meet the research criteria based on titles and

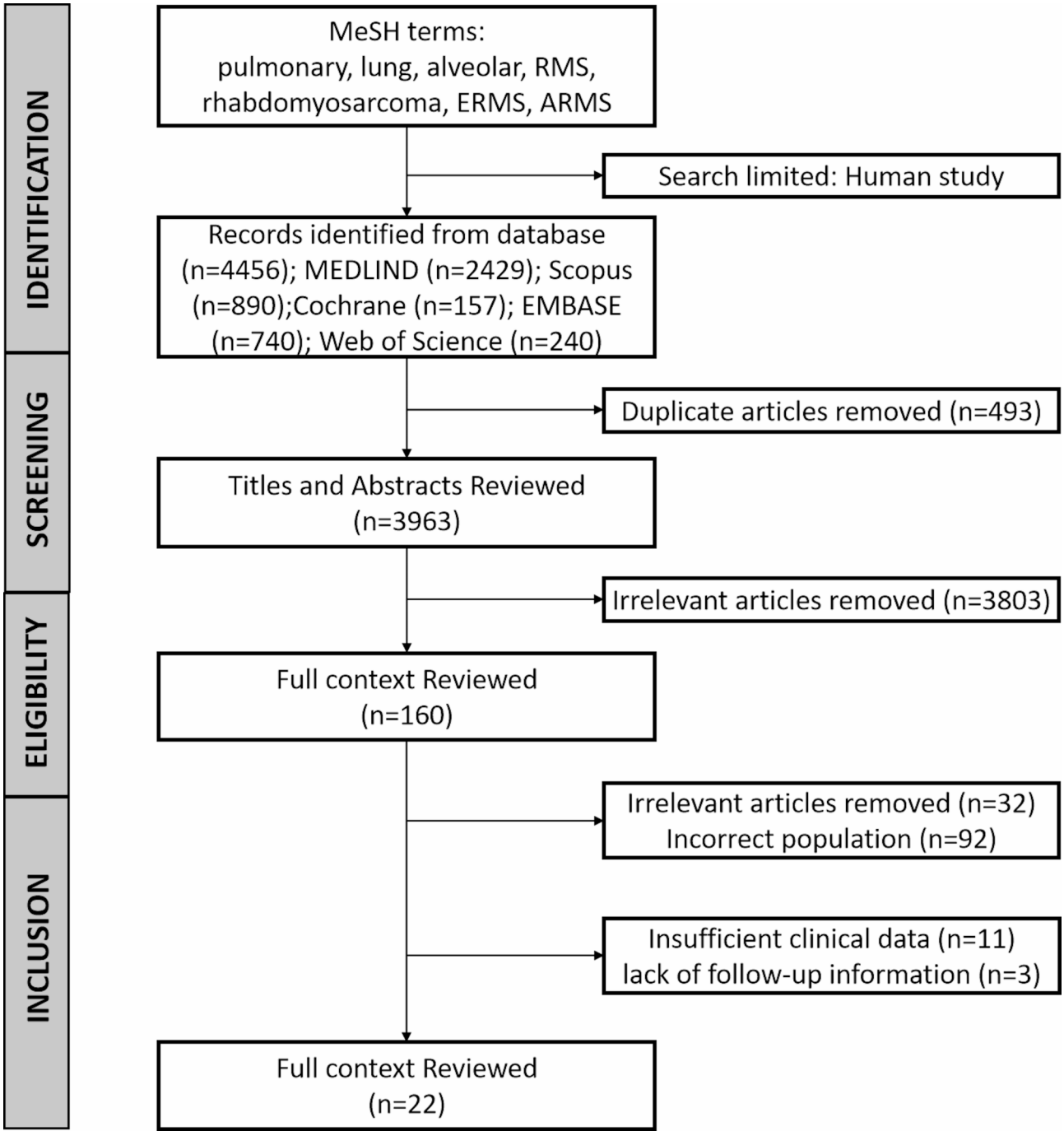

Fig. 1 Systematic review of the literature according to PRISMA. PRISMA: Preferred Reporting Items for Systematic Reviews and Meta-Analyses

abstracts, we further removed 124 articles after full-text review for not matching the theme or targeting inappropriate research groups. Additionally, we excluded 11 articles with incomplete clinical data and 3 with incomplete follow-up data. Ultimately, 22 articles met the criteria for inclusion in the systematic review.

The analysis included 22 case reports on primary pulmonary RMS, documenting 28 patient cases. Table 1 summarizes all reported cases, listing authors, publication years, patient ages, genders, histopathological classification, lesion sizes, sites of metastasis, TNM staging, initial treatments, and prognosis outcomes concerning recurrence and survival. Supplementary Table 1 compiles information on patients' smoking histories, clinical manifestations, physical examinations, laboratory tests, imaging examinations, and diagnostic methods.

The literature reviewed in this study spanned nearly 70 years, from 1955 to 2023. Across all 28 cases, the

Table 1 Cases of primary pulmonary rhabdomyosarcoma arranged by publication date

Author; Year of publication	Sex	Age (years)	Histology	Tumour size (cm)	Metastatic site	TNM Stage	Initial treatment	Recurrence (time, Months)	Survival Outcome (time, Months)
Jin et al. [20]; 2023	M	75	N/A	5–10	None	3	CT	No	DDD (2 M)
Nishioka et al. [13]; 2018	F	66	Alveolar	5–10	Lymphatic node	3	PC + CT	No	Alive (9 M)
Kandola et al. [25]; 2018	M	20	Alveolar	>10	None	3	RT + CT	No	DDD (2 M)
Ji et al. [26]; 2013	M	51	Alveolar	5–10	None	3	CT	No	DDD (8 M)
Guo et al. [21]; 2013	F	3	Embryonal	5–10	Brain	4	None	No	DDD (1 M)
Yoko et al. [27]; 2009	M	64	N/A	>10	None	3	None	No	DDD (1 M)
Gupta et al. [22]; 2007	M	38	Pleomorphic	>10	Pleura and lymphatic node	4	SC + CT	Yes (1.5 M)	DDD (2.5 M)
Doladzas et al. [28]; 2005	F	2	Pleomorphic	1–5	None	3	PC + RT + CT	No	Alive (120 M)
Comin et al. [29]; 2001	M	62	N/A	5–10	Lymphatic node	3	PC + RT	No	Alive (9 M)
Agostino et al. [30]; 1997	F	2	Embryonal	>10	None	3	PC + CT	No	Alive (72 M)
Schiavetti et al. [23]; 1996	F	2.5	Embryonal	>10	None	3	PC + RT + CT	Yes (14 M)	DDD (14 M)
Schiavetti et al. [23]; 1996	F	1.5	Embryonal	>10	None	3	PC + CT	Yes (2 M)	DDD (9 M)
Noda et al. [31]; 1995	M	2	Alveolar	5–10	None	3	PC + CT + RT	Yes (6 M)	Alive (38 M)
Doval et al. [32]; 1994	M	10	Embryonal	1–5	None	3	CRT	No	DDUD (36 M)
Murphy et al. [33]; 1992	F	2	Embryonal	5–10	Pleura	4	PC + CT	No	Alive (12 M)
Murphy et al. [33]; 1992	F	3	Embryonal	5–10	None	3	PC + CT	No	Alive (3 M)
Murphy et al. [33]; 1992	M	3.5	Embryonal	5–10	Pleura and lymphatic node	4	PC + CT	No	Alive (6 M)
Shariff et al. [34]; 1988	F	1	N/A	5–10	None	3	PC	No	Alive (3 M)
Allan et al. [19]; 1987	F	2.5	N/A	>10	Pleura	4	PC + CT	Yes (5 M)	Alive (11 M)
Allan et al. [19]; 1987	F	2	N/A	1–5	None	2	PC + CT	No	Alive (48 M)
Avagnina et al. [35]; 1984	F	43	N/A	5–10	Small intestine	4	PC	Yes (11 M)	Alive (23 M)
Hartman et al. [24]; 1983	M	11	N/A	1–5	None	2	PC + RT + CT	No	Alive (24 M)
Hartman et al. [24]; 1983	F	13	Embryonal	1–5	None	3	PC + RT + CT	Yes (40 M)	Alive (60 M)
Lee et al. [36]; 1981	M	69	N/A	>10	Pleura, adrenal gland and brain	4	None	No	DDD (1 M)
Thomas et al. [37]; 1981	M	2	Embryonal	>10	None	3	PC	No	Alive (60 M)
Conquest et al. [38]; 1965	M	53	Pleomorphic	5–10	None	3	PC	No	DDUD (152 M)
Conquest et al.[38]; 1965	F	66	Pleomorphic	1–5	None	2	PC	No	Alive (40 M)
Gordon et al. [39]; 1955	F	20	N/A	>10	Intrapulmonary metastasis and liver	4	None	No	DDD (12 M)

CRT: chemoradiotherapy; CT: chemotherapy; DDD: die due to disease; DDUD: die due to unrelated disease; F: female; M: male; N/A: not available; PC: partial cystectomy; RT: radiotherapy; SC: simple cystectomy

median patient age was 10.5 (2.0, 52.5) years, with 15 cases (53.6%) involving patients under 18 years old. The male-to-female ratio was nearly equal (13:15). 17.8% of the patients reported a history of smoking, while five pediatric patients had congenital cystic adenomatoid malformation (CCAM), accounting for 1/3 of all pediatric cases. Nearly all patients exhibited clinical symptoms at the initial diagnosis, with 22 cases (78.6%) presenting respiratory symptoms attributed to pulmonary masses, including cough, chest pain, shortness of breath, difficulty breathing, and even spontaneous pneumothorax. Six patients (21.4%) experienced systemic inflammatory symptoms, including fever, fatigue, and weight loss; two cases (7.1%) reported abdominal discomfort; and one (3.6%) initially presented with unexplained swelling of the ankles, hands, and knee pain, as detailed in Supplementary Table 1.

The diagnosis for all patients was ultimately confirmed through tissue biopsy or autopsy. Of the 18 patients with a clear pathological classification, 10 (55.6%) were diagnosed with the embryonal type, while 4 (22.2%) were identified as either alveolar or pleomorphic types. Tumor size was determined from pathological specimens or, when unavailable, from imaging examinations. At initial treatment, all patients' tumors measured over 1 cm, with 6 cases (21.4%) between 1 and 5 cm, 42.9% between 5 and 10 cm, and 35.7% exceeding 10 cm at the primary pulmonary site. 10 cases (35.7%) showed local lymph node or distant organ metastasis at initial diagnosis, confirmed through imaging or surgery, with pleural metastasis as the most common type. TNM staging at diagnosis revealed that only 3 patients (10.7%) were at stage II, while the majority, 17 (60.7%), were at stage III.

Table 2 Cox proportional hazard univariate analysis of factors with outcomes of recurrence and cancer-specific survival

Characteristics	PFS		CSS	
	HR (95% CI)	*P*-value	HR (95% CI)	*P*-value
Sex (male vs. female)	1.71 (0.62–4.76)	0.30	2.138 (0.60–7.62)	0.24
Age (<18 yrs vs. ≥18 yrs)	0.47 (0.17–1.31)	0.15	0.243 (0.062–0.95)	**0.042**
Histology				
Embryonal	1	0.45	1	0.77
Alveolar	0.86 (0.27–2.72)	0.80	0.78 (0.19–3.26)	0.73
Other	2.22 (0.53–9.28)	0.28	1.52 (0.29–7.88)	0.62
Tumor size (<10 cm vs. ≥10 cm)	0.37 (0.13–1.02)	0.055	0.20 (0.052–0.78)	**0.020**
TNM stage (II, III vs. IV)	0.30 (0.096–0.94)	**0.038**	0.45 (0.12–1.61)	0.22
Surgery (NO vs. YES)	5.76 (1.96–16.87)	**0.001**	11.37 (2.84–45.52)	**0.001**
Radiotherapy (NO vs. YES)	1.086 (0.37–3.20)	0.88	2.17 (0.46–10.36)	0.33
Chemotherapy (NO vs. YES)	0.98 (0.33–2.88)	0.97	1.37 (0.39–4.87)	0.63

*Bold values indicate statistical significance ($P < 0.05$)

CI: confidence interval; CSS: cancer-specific survival; HR: hazard ratio; PFS: progression-free survival; yrs: years

Table 3 Cox proportional hazard multivariate regression analysis to determine factors of age, tumor size, TNM stage and surgery with outcomes of recurrence and cancer-specific survival

Characteristics	PFS		CSS	
	HR (95% CI)	*P*-value	HR (95% CI)	*P*-value
Age (<18 yrs vs. ≥18 yrs)	0.95 (0.27–3.41)	0.94	0.53 (0.085–3.52)	0.52
Tumor size (<10 cm vs. ≥10 cm)	0.54 (0.17–1.66)	0.28	0.24 (0.052–1.12)	0.069
TNM stage (II, III vs. IV)	0.48 (0.14–1.72)	0.26	1.32 (0.27–6.42)	0.73
Surgery (NO vs. YES)	4.58 (1.32–15.90)	**0.017**	8.11 (1.45–45.50)	**0.017**

*Bold values indicate statistical significance ($P < 0.05$)

CI: confidence interval; CSS: cancer-specific survival; HR: hazard ratio; PFS: progression-free survival; yrs: years

Regarding treatment and prognosis, three patients died without a confirmed diagnosis and received no treatment. The majority of patients ($n = 20$, 71.4%) underwent surgical intervention, with all but one undergoing a lobectomy; the exception involved excision of only the pulmonary tumor. Additionally, 18 patients (64.3%) received chemotherapy, and 8 patients (28.6%) underwent radiotherapy. Notably, one patient opted for concurrent chemoradiotherapy instead of surgery. The median follow-up duration for all patients was 11.5 (3.0, 39.5) months. Within this cohort, 12 patients ultimately died, including two from non-tumor-related causes. Additionally, post-treatment progression occurred in 7 patients, with a median progression time of 6.0 (2.0, 14.0) months. In terms of prognosis, the average PFS time was 60.9 ± 14.8 months, while the overall survival (OS) time averaged 85.6 ± 16.0 months.

Cox proportional hazard univariate and multivariate regression analysis of factors with outcomes of PFS and CSS are detailed in Tables 2 and 3, respectively. Regarding tumor progression post-treatment, patients at TNM stage IV were more prone to progression compared to those at stages II and III (HR = 0.30 (0.096–0.94), $P = 0.038$). Patients who underwent surgery showed a higher likelihood of progression compared to those who did not (HR = 5.76 (1.96–16.87), $P = 0.001$). Additionally, patients with primary tumor size ≥ 10 cm tended to exhibit greater progression, although these results were not statistically significant (HR = 0.37 (0.13–1.02), $P = 0.055$). In multivariable regression analysis, surgery was identified as an independent factor that could shorten progression time (HR = 4.58 (1.32–15.90), $P = 0.017$). Regarding CSS, factors significantly associated with increased tumor-related mortality included age ≥ 18 years (HR = 0.243 (0.062–0.95), $P = 0.042$), primary tumor size ≥ 10 cm (HR = 0.20 (0.052–0.78), $P = 0.020$), and non-surgical treatment (HR = 11.37 (2.84–45.52), $P = 0.001$), as detailed in Fig. 2. Multivariate analysis confirmed surgery as an independent factor improving tumor-related survival (HR = 8.11 (1.45–45.50), $P = 0.017$). Additionally, patients with tumor size ≥ 10 cm exhibited a reduced survival rate, although this was not statistically significant (HR = 0.24 (0.052–1.12), $P = 0.069$).

Discussion

RMS is the most prevalent soft tissue malignancy in pediatrics, accounting for approximately half of all pediatric soft tissue sarcomas [16]. The primary sites of occurrence include the head and neck region, genitourinary tract, and limbs, with pathological classifications primarily comprising embryonal, alveolar, and pleomorphic variants [5]. Primary pulmonary RMS is exceptionally rare, with the first case reported by McDonald et al. in 1939 [17]. Data from the Intergroup Rhabdomyosarcoma Study (IRS) indicate that fewer than 3% of enrolled patients had primary tumors in the mediastinum, pleura, or lungs [18]. In children, primary pulmonary RMS predominantly presents as embryonal or alveolar variants, whereas in adults, it is primarily seen as pleomorphic variants [19]. This review systematically summarizes global case reports of pulmonary RMS for the first time, analyzing its clinical and pathological characteristics, along with survival-related prognostic factors, to offer

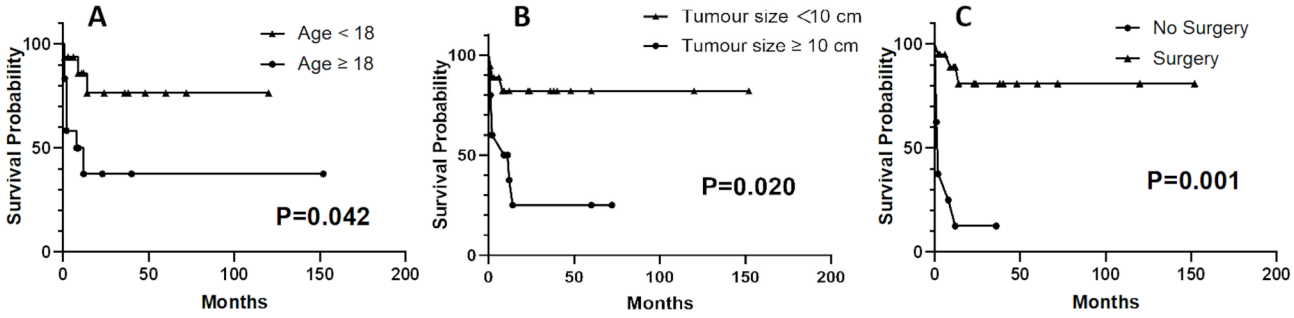

Fig. 2 Kaplan-Meier cancer-specific survival curves of patients according to age, tumor size, and surgery. (A) Kaplan-Meier survival curves according to age. (B) Kaplan-Meier survival curves according to tumor size. (C) Kaplan-Meier survival curves according to surgery

insights for clinical treatment strategies for this rare disease. The study results revealed that the median age of patients was 10.5 (2.0, 52.5) years, with a nearly equal male-to-female ratio. Nearly all patients exhibited respiratory system-related clinical symptoms at initial diagnosis, with embryonal variants being predominant.

In this cohort, the average PFS time was 60.9 ± 14.8 months, and the OS time was 85.6 ± 16.0 months, reflecting a generally pessimistic outlook. Furthermore, seven patients experienced progression after treatment, with a median progression time of 6.0 (2.0, 14.0) months. The high recurrence rate and rapid progression further underscored the lethality of primary pulmonary RMS. Difficulty in early diagnosis likely contributes significantly to the poor prognosis. The onset age of primary pulmonary RMS peaks in children and the elderly [20]. Particularly for pediatric patients and infants who cannot verbalize their symptoms, routine chest computed tomography (CT) screenings are seldom performed. Additionally, the initial symptoms of primary pulmonary RMS, which include non-specific respiratory manifestations like cough, chest pain, shortness of breath, and difficulty breathing, are often underestimated. Patients often seek hospital care only when the tumor exacerbates, causing hypoxia or other clinical symptoms like local infiltration damage, thus delaying diagnosis. The rarity of pulmonary RMS and challenges in determining its pulmonary origin also complicate pathological diagnosis. Some patients require referral to specialized cancer centers, which further delays early diagnosis [21]. Currently, no standard treatment protocol exists for primary pulmonary RMS. Surgery is the primary treatment modality; however, curative resection is not feasible for all patients. In such cases, radiotherapy is critical for achieving local control [12, 22]. Furthermore, pediatric patients often have reduced tolerance to radiotherapy and chemotherapy, the key treatment options, which may lead to poorer survival outcomes.

Regarding prognosis, univariate and multivariate analyses of PFS and CSS revealed that higher TNM staging, primary tumor sizes ≥ 10 cm, and ages ≥ 18 years were associated with increased progression or tumor-related mortality, likely due to a positive correlation with disease severity. Additionally, surgery, the primary treatment method, independently improved tumor-related survival and could facilitate precise pathological diagnosis of primary pulmonary RMS. However, several studies indicated that RMS patients, particularly adults, who underwent only surgery exhibited a lower survival rate [11, 12]. Therefore, exploring alternative treatment strategies, such as radiotherapy or chemotherapy, is essential to improve the survival and prognosis of pulmonary RMS patients.

RMS is sensitive to both radiotherapy and chemotherapy, making them essential components of comprehensive treatment strategies. However, in this study, univariate and multivariate analyses related to survival revealed that neither radiotherapy nor chemotherapy independently improved CSS rates or reduced tumor progression in treated patients. In radiotherapy contexts, when complete tumor removal is unfeasible, radiotherapy aids in enhanced local tumor control. The radiation field should encompass the grossly apparent tumor with a margin of normal tissue [22]. Nonetheless, for excessively large primary tumors, further increasing the radiation dose becomes impractical, thereby limiting treatment efficacy and emphasizing the critical role of early diagnosis. Furthermore, the rarity and low incidence of primary pulmonary RMS, coupled with the lack of large-scale clinical cohorts, mean that optimal radiation doses and fractionation schemes require further exploration. Regarding chemotherapy, the standard regimen is Vincristine + Adriamycin + Cyclophosphamide (VAC) chemotherapy [6]. However, the responsiveness of primary pulmonary RMS to this treatment warrants further investigation, as some cases have shown tumor progression, recurrence, or metastasis during chemotherapy [20, 23, 24]. Additionally, children exhibit poorer tolerance to chemotherapy than adults, which increases the risk of treatment-related adverse reactions.

Currently, there are no guideline-recommended standard treatment protocols specifically for primary pulmonary RMS, and the existing studies suggest a lack of uniform treatment strategies. We believe that surgical treatment remains recommended for early-stage lesions. Although this study, due to its limited sample size, could not compare the benefits of complete resection versus partial resection, we can infer from the experience with RMS in other anatomical sites that complete resection with a negative margin is optimal. Chemotherapy, radiotherapy, or a combination of both should be selected based on the individual patient's condition. However, this certainly warrants further study. Our understanding of the pathophysiological characteristics of primary pulmonary RMS and how it differs from RMS at other sites remains limited. The responsiveness of pulmonary RMS to radiotherapy and chemotherapy requires further validation through animal models and large-scale clinical cohort studies, a direction we will pursue in future research.

As the inaugural effort to systematically summarize global case reports on pulmonary RMS, this study spanned related literature from 1955 to 2023, covering nearly 70 years. Regrettably, the number of cases collected remained small, and the follow-up period was limited, suggesting that patients recorded as recurrence-free in the literature might experience delayed recurrences. Additionally, the potential heterogeneity between cases due to such a wide age range needs to be acknowledged. For example, differences in the level of radiotherapy across different time periods, leading to variations in patient survival, are unavoidable. These limitations impede more precise subgroup analyses and contribute to statistical biases. Future research on primary pulmonary RMS requires larger clinical cohorts and extended follow-up periods to more accurately evaluate risk factors affecting patient survival prognosis and to offer more substantive references for clinical treatment strategies.

Conclusions

Primary pulmonary RMS, a rare soft tissue sarcoma, is associated with high recurrence rates and poor survival outcomes. This study involved a systematic review of the literature, summarizing clinical characteristics, pathological features, treatment approaches, and clinical outcomes of primary pulmonary RMS, and analyzed prognostic-related risk factors to inform clinical treatment strategies for this condition. The findings revealed that patients not undergoing surgery with higher TNM stages tended to have more aggressive tumors, whereas primary tumors ≥ 10 cm and ages ≥ 18 years were associated with increased tumor-related mortality. Surgery, the primary treatment modality, independently improved CSS rates. Exploring optimal comprehensive treatment strategies that combine surgery, radiotherapy, and chemotherapy represents the principal direction for future research.

Abbreviations

CCAM	Congenital cystic adenomatoid malformation
CI	Confidence interval
CRT	Chemoradiotherapy
CSS	Cancer-specific survival
CT	Computed tomography
DDD	Die due to disease
DDUD	Die due to unrelated disease
EpSSG	European Pediatric Soft Tissue Sarcoma Study Group
F	Female
HR	Hazard ratio
IRS	Intergroup Rhabdomyosarcoma Study
IRSG	Intergroup Rhabdomyosarcoma Study Group
M	Male
N/A	Not available
PC	Partial cystectomy
PFS	Progression-free survival
PRISMA	Preferred Reporting Items for Systematic Reviews and Meta-Analyses
RMS	Rhabdomyosarcoma
RT	Radiotherapy
SC	Simple cystectomy
VAC	Vincristine + Adriamycin + Cyclophosphamide
yrs	Years

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12887-025-05521-y .

Supplementary Material 1

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Not applicable.

Author contributions

All authors contributed to the conceptualization of the study. Y.T. and W.C. were responsible for methodology, data curation, investigation, formal analysis, writing– original draft and writing– review and editing. H.Z., J.S. and H.G. were responsible for data curation and investigation. Z.L. was responsible for project administration, supervision, funding acquisition and writing– review and editing. All authors read and approved the final manuscript.

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Data availability

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Competing interests

The authors declare no competing interests.

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