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Pulmonary Vascular Sarcomas: Clinicopathologic Analysis of 14 Cases

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Background: Pulmonary vessel sarcomas are rare, and their pathogenesis is still unclear. **Methods:** We focus on the pathologic findings of fourteen pulmonary artery and/or vein sarcomas along with clinical prognosis. **Results:** Nine patients were male and five were female, and they ranged in age from 26 to 72 years (mean, 47 years). There were ten cases of pulmonary artery sarcoma, three cases of pulmonary artery and vein sarcoma, and one case of pure pulmonary vein sarcoma. Ten out of the fourteen cases were associated with pulmonary thromboembolism. Microscopically, all the tumors showed an undifferentiated sarcomatous portion. There were leiomyosarcoma portions in 8 cases, malignant fibrous histiocytomatous portions in 7 cases, angiosarcomatous differentiation in 3 cases, and osteosarcomatous portion in 1 case. All but two patients died during the follow up period (range, 1 to 78 months). The mean survival time of the patients who died was 14 months and the longest survival time was 78 months after surgical resection. **Conclusions:** The current study is one of the largest single institutional reviews of pulmonary artery and/or vein sarcoma. Regardless of the histological components and macroscopic growth patterns, these rare tumors have a grave prognosis.

Key Words: Sarcoma; Pulmonary artery; Pulmonary veins

Primary neoplasms of the great vessels (aorta, pulmonary vessels, and vena cava) are rare. The pathogenesis of these tumors is still unclear, and it has been suggested that these tumors arise from the mesenchymal cells of the muscle anlage in the bulbus cordis in cases of pulmonary artery sarcoma.[1] Most primary sarcomas of the elastic vessels such as the aorta and the pulmonary artery or vein are undifferentiated sarcomas, while leiomyosarcomas predominate in the muscular arteries and great veins such as the inferior vena cava.[2,3] Pulmonary vessel sarcomas are rare and show no characteristic symptoms, even though the thromboembolic events are the most severe events, and a subsequently delayed diagnosis may contribute to a fulminant prognosis. Recent advances in imaging modalities and accumulated clinical experience have made it possible to preoperatively diagnose these tumors. Most pulmonary vessel sarcomas have been reported as single case reports and there have only been a few radiologic review articles.[3,4] Here, we review pulmonary artery and vein sarcomas with their clinicopathologic features.

MATERIALS AND METHODS

The data were retrospectively collected on all patients who were pathologically confirmed as having pulmonary vessel sarcomas between 1994 and 2010. The included cases were unequivocal cases in which there was evidence of a primary tumor arising within the lumen of the pulmonary vessels radiologically and/or grossly at the time of surgical specimen examination. Clinical history and follow-up data were obtained from the Korea National Statistics Office and the referring physicians. This study was approved by the Institutional Review Board of Samsung Medical Center (IRB No. 2010-09-110). The resected specimens were fixed in 10% buffered formalin and embedded in paraffin. Each case was routinely stained with hematoxylin and eosin.

In order to confirm the specific differentiation of the tumors, immunohistochemistry was done using avidin-biotin-peroxidase complex methods. The employed primary antibodies were against the following antigens: CD31 (1:20, JC/70A, Dako, Glostrup, Denmark), factor VIII-related antigen (1:150, Z002, Biogenesis, Poole, UK), CD34 (prediluted, QBEnd 10, Dako),

CD68 (1:50, PG-M1, Dako), alpha-1-antitrypsin (1:40,000, A1AT, Dako), vimentin (1:100, V9, Zymed, San Francisco, CA, USA), smooth muscle actin (1:100, 1A4, Zymed), desmin (1:100, D33, Dako) and Ki-67 (1:250, polyclonal, Dako).

RESULTS

Clinical findings

A search identified ten cases of pulmonary artery sarcoma, three cases of pulmonary artery and vein sarcoma, and one case of pure pulmonary vein sarcoma. Among them, 8 previously reported cases were also included.[3,5,6] Nine patients were male and five were female. The ages of the patients ranged from 26 to 72 years (mean age, 47 years). The patients displayed non-specific presenting symptoms, such as cough (8/14, 57.1%), dyspnea (8/14, 57.1%), hemoptysis (4/14, 28.6%), chest pain (4/14, 28.6%), weight loss (3/14, 21.4%), superior vena cava (SVC) syndrome (1/14, 7.1%) and pleural effusion (1/14, 7.1%). One patient presented with pathologic vertebral fracture and paraspinal abscess-like masses, and the latter were initially misinterpreted as tuberculous spondylitis (1/14, 7.1%). Ten out of fourteen patients (10/14, 71.4%) were accompanied by pulmonary thromboembolism with no deep vein thromboses. The follow up duration ranged from 1 to 78 months (mean, 22 months). All except for two cases died. Their mean survival was 14 months. Case 13 is alive with recurrent masses for over 6 years. In particular, case 14 had been aware of an incidentally found lung mass 4 years before the surgical procedure, and he is stable with-

Table 1. The clinicoradiologic findings of the fourteen cases of pulmonary artery and vein sarcomas in the present study

Case No.	Age (yr), sex	Size of tumor (cm)	Presenting symptom	Thrombo-embolism	Symptom duration	Radiology	Procedure	Outcome (duration of follow up)
1	31, F	4	Cough, hemoptysis	+	4 yr	Hilar mass, RA mass	Wedge resection, chemotherapy, RT	Dead with RA mass (36 mo)
2	37, M	6	Cough, hemoptysis	+	1 mo	Mass-like opacity in LLL Invasion to LA, anterior wall of aorta	Lobectomy, chemotherapy, RT	Dead (14 mo) with metastatic masses in portal vein, splenic vein, SMV, adrenal gland
3	63, F	3	Asthma-like dyspnea with paroxysmal cough, hemoptysis, weight loss	-	7 yr	Hilar mass with eccentric calcification	Pneumonectomy, chemotherapy, RT	Dead with abdominal metastatic lesion (15 mo)
4	56, F	8	Orthopnea, chest pain, cough, weight loss	+	6 mo	Hilar mass with invasion to azygos vein and SVC	Pneumonectomy	Dead with brain metastasis (16 mo)
5	26, M	5	Orthopnea, dyspnea, chest pain	+	2 mo	Anterior mediastinal mass, invasion to LA, pericardium	Mass excision and pericardial biopsy, chemotherapy	Dead (2 mo)
6	39, M	5	Cough, dyspnea, weight loss	+	4 yr	Pulmonary infarct with multiple pulmonary nodules	Pneumonectomy	Dead (18 mo)
7	50, F	8	Cough, dyspnea	+	1 mo	Hilar mass	Pneumonectomy, chemotherapy, RT	Dead with multilple metastatic masses (4 mo)
8	46, M	8	Cough, blood-tinged sputum, chest pain	-	2 mo	Soft tissue mass surrounding bronchus intermedius and right lower bronchus	Bilobectomy, RT chemotherapy	Dead with pleural metastasis (40 mo)
9	61, F	8	Dyspnea	+	1 mo	Hilar mass with multiple masses in the entire lung	Pneumonectomy	Dead with metastatic IVC and pericardial masses (8 mo)
10	58, M	6	Cough, dyspnea	+	3 mo	Hilar mass, main pulmonary trunk, LA	Chemotherapy	Dead with acute cor pulmonale (1 mo)
11	72, M	6	Right flank and chest pain, dyspnea	+	1 mo	Pleural effusion, RUL mass	Lobectomy, RT, chemotherapy	Dead with brain metastasis (7 mo)
12	50, M	5.5	SVC syndrome, pathologic fracture due to metastatic spine mass	-		Paratracheal mass invading SVC	VAT's biopsy, RT, chemotherapy	Dead (7 mo)
13	28, M	10	Hemoptysis	+	3 mo	Unilateral pneumonia-like consolidation	Pneumonectomy, chemotherapy	Alive with local recurrence (42 mo) and tiny pleural seeding (78 mo)
14	50, M	8.5	Dyspnea, known lung mass	-	4 yr	Intraluminal filling defect in the Rt MPA	Pneumonectomy	Alive with no recurrent mass (65 mo)

F, female; M, male; RA, right atrium; RT, radiation therapy; LLL, left lower lobe; LA, left atrium; SMV, superior mesenteric vein; SVC, superior vena cava; IVC, inferior vena cava; RUL, right upper lobe; VAT, video-assisted thoracotomy; Rt MPA, right main pulmonary artery.

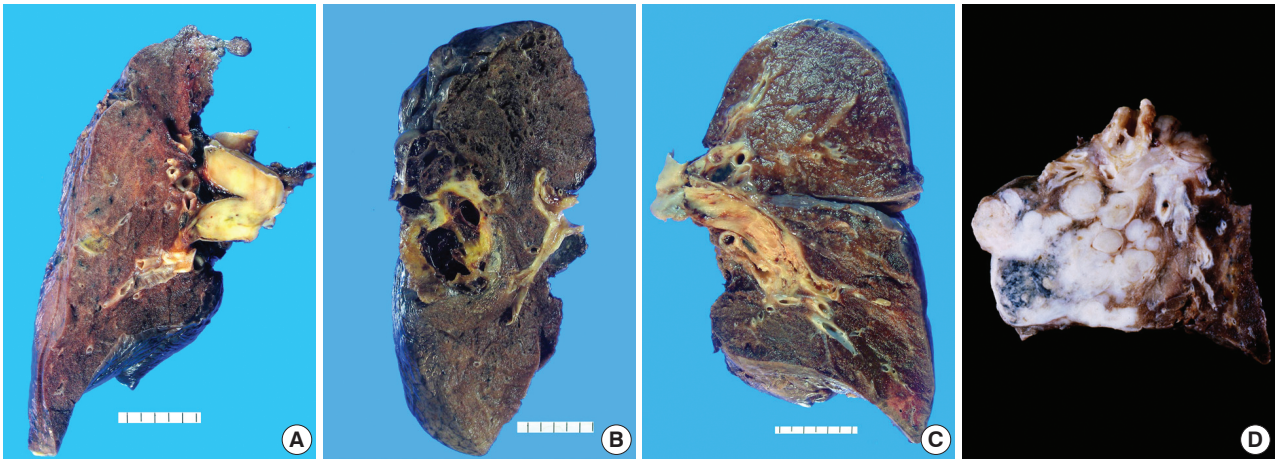

Fig. 1. Gross photographs of pulmonary artery and/or vein sarcomas. (A) In case 9, a yellow tan mass completely obstructs the lumen of right pulmonary artery with no invasion of lung parenchyma. (B) Case 11 shows a well demarcated lobulated ovoid mass with hemorrhagic changes and cavitation. Note visceral pleural invasion without bronchial involvement. (C) In case 13, a gray white mass is located in the pulmonary artery, and it grows into three lobes along the vascular tree. (D) Case 2 shows that the tumor extends along the pulmonary vasculature, like serpentine growth, and obstructs the lumen.

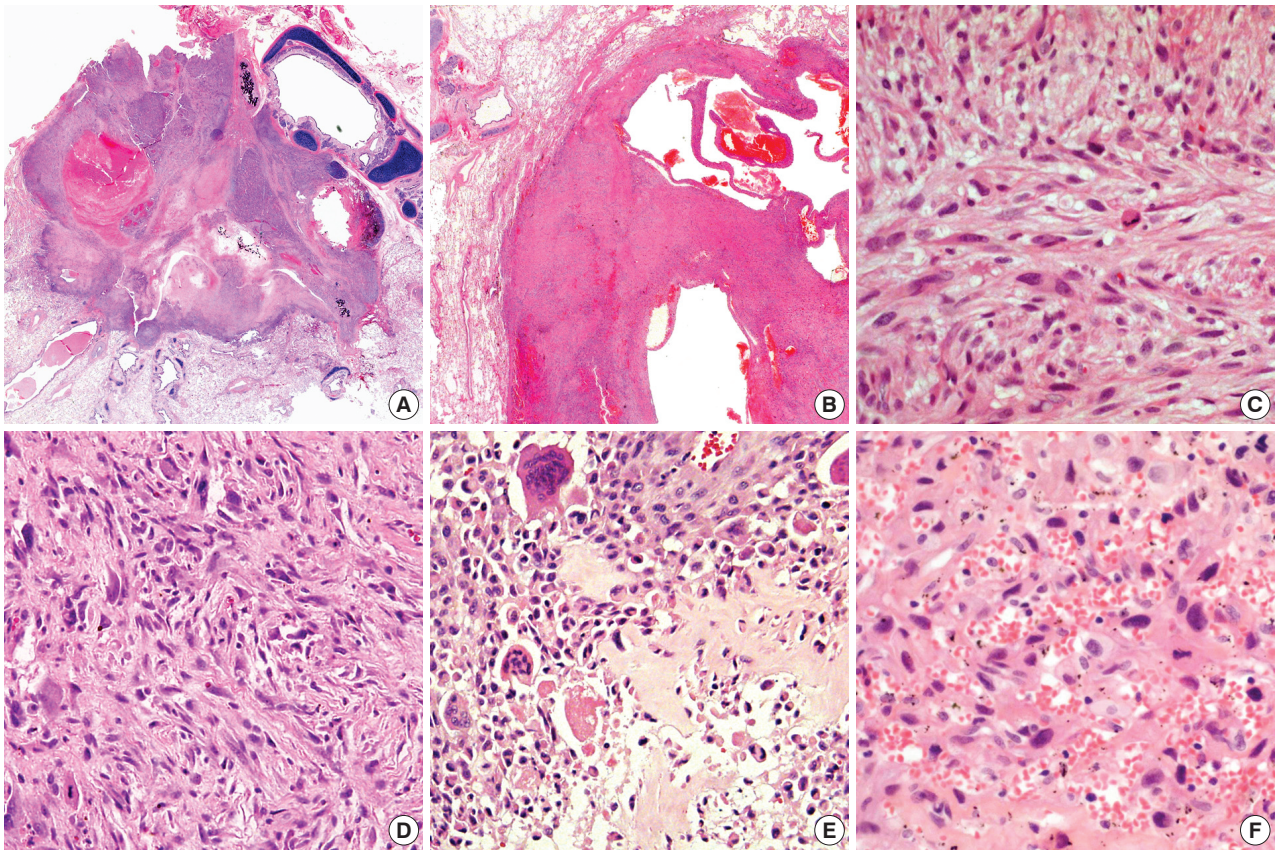

Fig. 2. (A) In case 2, low power view shows an irregular shaped tumor infiltrating to the bronchial wall. (B) In case 11, the tumors show intratumoral hemorrhage and cystic changes. (C) In a leiomyosarcomatous portion of case 13, spindle cells having the cigar-shaped nuclei are arranged in interlacing fascicles. (D) In a storiform-pleomorphic malignant fibrous histiocytomatous differentiation of case 8, short fascicles of spindle cells are radiating from a central portion which is intermixed neoplastic pleomorphic cells. (E) The round and spindle shaped tumor cells with lace-like eosinophilic osteoid as well as abundant giant cells are seen in a portion of osteosarcomatous differentiation of case 2. (F) In case 5, irregular shaped vascular channels are lined by atypical tumor cells, indicating angiosarcomatous differentiation.

out any evidence of recurrence, with no additional chemotherapy for 5.4 years of postoperative follow up. The clinical summary is shown in Table 1.

Gross findings

Intraoperatively, the tumors of cases 1, 2, 5, and 10 had invaded the left or right atrium (4/14, 28.6%). All the masses obstructed the lumina of the pulmonary artery or vein (Fig. 1A). In case 8, a necrotic mass encircling the pulmonary artery was grossly infiltrating the vascular wall. In case 11, the tumor showed extensive hemorrhagic cavity formation (Fig. 1B). In cases 3, 6, 8, and 9, poorly defined irregular masses involved the adjacent lobar bronchi. Blood clots intermingled with masses filled the upper and lower lobar bronchus in cases 3 and 6. Direct mucosal invasion and erosion were seen in case 8. In case 5, the mass directly invaded the pericardium. In cases 2 and 13, irregular parenchymal masses arose from the pulmonary artery or vein, and they grew through the vascular pathway (Fig. 1C, D). In case 12, the pulmonary artery sarcoma directly invaded the SVC and the azygus vein.

Microscopic findings

Microscopic bronchial infiltration was demonstrated in cases 2 and 4 (Fig. 2A). In cases 2 and 11, the tumors showed parenchymal hemorrhage and thrombi in the intrapulmonary or main pulmonary artery and veins (Fig. 2B). Direct invasion to lymph nodes occurred in cases 7, 13, and 14. Microscopically, all the cases of the totally resected 12 masses showed the tumors had portions of undifferentiated or poorly differentiated sarcoma with fibroblastic differentiation. Among differentiated sarcomatous portions identified, leiomyosarcomatous portions in 8 cases, malignant fibrous histiocytomatous portions in 7 cases, angiosarcomatous differentiations in 3 cases, and giant cell-rich osteosarcomatous portion in 1 case (Fig. 2C-F) were identified. All the cases except for case 14 showing low grade myxofibroblastic sarcomatous portion were categorized to grade 3 sarcomas according to a 3-tiered grading system by Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC).[7,8] Immunohistochemically, the spindle cells were reactive for vimentin in all fourteen cases. The leiomyosarcomatous portions were stained with smooth muscle actin and desmin in cases 2, 4, 5, 6, 7, 9, 11, and 13. The malignant fibrous histiocytomatous differentiations in the cases with in cases 2, 3, 7, 8, 9, 11, and 14 were positive for CD68 and A1AT. Angiosarcomatous differentiations in cases 3, 5 and 9 were positive for CD31, CD34 and factor VIII-related antigen. The proliferation indices ranged from 5 to 80% in Ki-67 immunostaining. Gross and microscopic characteristics of the tumors are summarized in Table 2.

Table 2. Gross and microscopic characteristics of the tumors of the present study

Case No.	Involved large vessels of sarcoma	Grading by FNCLCC	Gross findings	Histologic types of sarcoma (tumor, %)		
				Undifferentiated sarcoma	Leiomyosarcoma	Other sarcomatous components
1	Artery sarcoma	3	Ill-defined firm mass in the lumen of the PA	100	-	-
2	Vein sarcoma	3	Serpentine growth along the pulmonary vessels	25	60	MFH (10), osteosarcoma (5)
3	Artery and vein sarcoma	3	Bronchial protruding mass	≤3	-	MFH (90), angiosarcoma (7)
4	Artery sarcoma	3	Serpentine growth along the bronchial artery	10	90	-
5	Artery and vein sarcoma	3	[a]	65	10	Angiosarcoma (25)
6	Artery sarcoma	3	Multiple tiny nodules	80	20	-
7	Artery sarcoma	3	Ill-defined mass in the PA	50	10	MFH (40)
8	Artery sarcoma	3	Bronchial protruding mass without involvement	40	-	MFH (60)
9	Artery sarcoma	3	Ill-defined firm mass in the lumen of the PA	10	60	Angiosarcoma (15), MFH (15)
10	Artery and vein sarcoma	3	[a]	Biopsy only (100)	-	-
11	Artery sarcoma	3	A firm RUL mass with tight adhesion to pleura	30	50	MFH (20)
12	Artery sarcoma	3	An elongated bulging mass between SVC and RA junction	Biopsy only (100)	-	-
13	Artery sarcoma	3	A mass at the PA extending to upper and lower trunk	40	60	-
14	Artery sarcoma	2	An intravascular mass in the lumen of Rt MPA	50	-	Myxoid MFH (50)

[a]The specimen is submitted in fragmented aggregates of the mass.
FNCLCC, Fédération Nationale des Centres de Lutte Contre le Cancer; PA, pulmonary artery; MFH, malignant fibrous histiocytoma; RUL, right upper lobe; SVC, superior vena cava; RA, right atrium; Rt MPA, right main pulmonary artery.

DISCUSSION

Primary pulmonary artery and/or vein sarcomas were first described by Mandelstamm[9] in 1923 and they are very rare tumors by themselves. Diagnosing these tumors is even more difficult due to the nonspecific symptoms.[4,10] Radiologically, the most common presentation is a hilar mass or a hilar infiltrative lesion, followed by decreased vascular markings, atelectasis and/or volume loss, multiple metastatic pulmonary nodules and an intravascular soft-tissue mass with decreased or absent perfusion on a ventilation/perfusion study.[11] In particular, the lesion can appear as an intraluminal filling defect that expands the vessel.[3] One of the specific computed tomography (CT) findings of extraluminal extension of the tumor was direct spread into the adjacent lung along the bronchial wall and lymph nodes.[1]

In the present series, grossly, there was the presence of a convex intraluminal bulging mass that mostly spread from the pulmonary trunk, which had continuously spread into the peripheral pulmonary artery branches, filling and dilating the vessels, and six cases showed macro- and microscopic bronchial infiltration of the tumor cells. The growth patterns of the tumors were characteristic in the present series. The lesion arose from the intimal layers of the right, left or main pulmonary artery or vein and the lesion extended as a polypoid mass into the small pulmonary arteries or branches of the vein. Less commonly, the mass grows in a retrograde fashion to involve the pulmonary valve and right ventricle for the artery sarcoma and the mass involved the left atrium for the pulmonary vein sarcoma, as was seen in case 2 of the present series.

From a histologic point of view, most pulmonary vessel sarcomas show undifferentiated spindle cell sarcomas of grade 3, leiomyosarcomas or malignant fibrous histiocytoma.[12] Leiomyosarcomas are uncommon tumors and have a predilection for deep soft tissues, with a rare group arising in the medium-sized or large veins far less frequently than in arteries. Immunohistochemical staining of these pulmonary vessel sarcomas is helpful in distinguishing the tumors. Although focal angiosarcomatous differentiation was seen in 3 cases, most pulmonary vessel sarcomas were composed of undifferentiated, multipotential, nonendothelial intimal stromal cells that were focally stained with endothelial markers such as CD31, CD34, and factor VIII-related antigen. Among variable histologies shown in pulmonary vascular sarcomas, angiosarcomas and low grade myofibrosarcomas are the least common.[13] Even with relatively favorable prognosis of low grade myofibrosarcomas, the grade but not the histologic subtype of the tumors has a significant bearing on the prognosis, based on results of the present series as well as previous studies.[14]

Pulmonary vessel sarcomas occur during the fifth decade of life with a nearly equal gender distribution although the present study showed a slight male predominance. However, the presenting signs and symptoms included weight loss, fever, anemia and clubbed digits with the exceptional occurrence of SVC syndrome if the SVC was involved or compressed.[15] The presenting symptoms such as cough, dyspnea, hemoptysis, chest pain, weight loss and pleural effusion were nonspecific and subtle. Ten patients out of the fourteen (10/14, 71.4%) had associated pulmonary thromboembolism because all the pulmonary vessel sarcomas showed filling of the lumen of the vessels to a variable extent. Pulmonary vessel sarcomas including intimal sarcomas were frequently misdiagnosed as pulmonary thromboembolism because of the tumors' rarity and insidious growth, leading to subsequent inappropriate therapy such as prolonged anticoagulation or thrombolysis. Therefore, pulmonary thromboembolism should be distinguished from these rare tumors for early diagnosis. Enhancement of an intraluminal filling defect with gadolinium on magnetic resonance imaging is a sensitive way of differentiating a tumor mass from thrombus.[16,17] The CT findings showing a low-attenuation filling defect occupying the entire lumen of the main or the proximal pulmonary artery with expansion of any segment of the pulmonary artery raise suspicion for pulmonary artery sarcoma.[3] In addition, the absence of predisposing factors for thromboembolism, the persistence of symptoms despite anticoagulation therapy, and the unilateral distribution of a massive perfusion defect indicate the possibility of pulmonary artery sarcoma.[1,18] In our series, the case presenting as a bronchial mass was unique in that the disease appeared to be an intractable asthma. The symptom of bronchial obstruction like in case 3 has been reported once previously.[2] Spindle cell carcinomas of the lung with vascular invasion and metastatic sarcomas to the lung should be distinguished in differential diagnosis.

The pathogenesis of pulmonary vessel sarcomas has not been understood and is mostly presumed as intimal fibroblastic or myofibroblastic in origin.[14] Recently, several molecular genetic studies of pulmonary vessel sarcomas have been attempted.[19-21] Overexpression of platelet derived growth factor α, dysregulation of the cell cycle due to overexpression of MDM2 and cdk4 have also been tried.[19,20] Adenomatous polyposis coli (*APC*), a tumor suppressor gene in the Wnt-signaling pathway, stabilizes β-catenin and controls cell growth. Loss of heterozygosity in microsatellite markers flanking chromosome 5q21.1 (*APC* gene)

was found. No mutations were detected in *APC* or β-catenin.[21]

Surgical treatment can either involve a pneumonectomy or excision of the tumor from the vessel with vascular reconstruction. As seen in case 14, pulmonary artery and/or vein sarcomas showed insidious slow growth and low histologic grade, and thus aggressive surgical intervention could have the potential to be curative even for high histologic grade tumors. Surgery with adjuvant chemotherapy or radiation might improve short-term survival. Yet, overall prognosis is poor with a five-year-survival rate of 0 to 6%. Compared with pulmonary parenchymal sarcoma, which behaves less aggressively and can be cured by resection with or without adjuvant therapy, the outcome of pulmonary artery and vein sarcoma is uniformly fatal. However, recently, long survivals of 5 years were reported despite surgical resection, like in cases 13 and 14 in the present series.[22,23] The patients who achieved long-term survival in our series underwent complete surgical resection with metastatectomy and this offered the chance of prolonged survival. As seen in the present review, the growth patterns of the tumors, such as retrograde ingrowing to the heart chambers and pulmonary parenchymal invasion, limits the extent of surgical resection.[24] Combined heart and lung transplantation is theoretically and technically feasible for highly selected patients with localized advanced pulmonary vessel sarcoma, the same as for primary cardiac sarcomas.[25] The high incidence of metastatic spread and the lack of donors usually prevent this type of dual transplantation.

In summary, the pathogenesis of pulmonary vessel sarcoma has not yet been clarified even after recent molecular cytogenetic trials. The grade rather than histologic subtypes might be more related to the disease course of pulmonary vascular sarcoma. Cumulative experiences and more reports about pathologic subtypes as well as molecular aspects are required to better define their biological behavior.

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