Problems and Countermeasures for Surgical Resection of Primary Pulmonary Artery Sarcoma

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Abstract

Background

This study clarified the problems and countermeasures of surgical treatment by examining surgical cases of pulmonary artery sarcoma.

Methods

The records of 10 patients with pulmonary artery sarcoma who underwent surgery at our hospital between January 2007 and October 2020 were retrospectively examined.

Results

The positive rate of vascular stumps was 7/10. Chest computed tomography scan showed positive margins of ≤ 20 mm between the tumor and surgical dissection in all cases (6/6). Additionally, the distance between the tumor location on computed tomography and the dissection line during surgery needed to be at least 20 mm (2/3). However, even at a distance of 25 mm, one case with a positive margin was observed. Postoperative recurrence was 6/9 cases, and the median recurrence period was as short as 9 months (range, 3-19 months). Postoperative treatment was required in 4/9 cases (chemotherapy/radiotherapy/chemoradiotherapy = 1/2/1). The median survival was 11 months (range, 0.5-24 months).

Conclusions

Extended surgery should be performed as much as possible, with a distance of at least 20 mm between the location of the tumor on computed tomography scan and the incision line during surgery. Intensive care for intrathoracic recurrence follow-up is required for 1 year after surgery.

Introduction

Pulmonary artery sarcoma (PAS) is a very rare and poorly understood tumor that arises from the intimal layer of the pulmonary artery (PA). In 1923, Mandelstamm published the first description of this sarcoma [1]. Since then, nearly 300 cases have been reported to date [2–4]. The age of onset of PAS ranges from 13-86 years [5, 6], with the majority of cases occurring in the middle-aged generation.

The results of chemotherapy or radiotherapy management alone for PAS are suboptimal.

Kruger et al. reported that the median survival time without surgical resection is 45 days, whereas it is 10 months with surgery [7], and surgery is still considered necessary as a treatment policy. Interventions reported for PAS include palliative stenting, total pneumonectomy, and endarterectomy, with or without pneumonectomy, and with or without PA reconstruction. Blackmon et al. reported that the 5-year survival rate was 49.2% for completely resected cases and 0% for incompletely resected cases [8]. In other words, since complete resection contributes to prognosis, reconstruction using artificial blood vessels is performed. However, since PAS is a tumor that grows while replacing the existing vascular endothelium, preoperative diagnosis does not indicate the extent to which the tumor has grown. Therefore, there are few positive resection reports that achieve complete resection. This study reviewed the management of 10 PAS patients, assessed short- and long-term outcomes, and assessed the likelihood of complete resection based on

preoperative computed tomography (CT), positron emission tomography (PET)-CT, and postoperative pathological results.

Methods

Study population

From January 2007 to December 2020, 10 patients underwent surgery for PAS at our institution. The operations were performed with curative intent in all 10 patients. The medical records were retrospectively reviewed to evaluate the clinical characteristics, operative findings, postoperative courses, and long-term results.

Surgical techniques

The operations were performed via thoracotomy and median sternotomy in three and seven patients, respectively. The diagnosis of PAS was histologically proven by frozen section biopsy during the operation. After confirmation of PAS, the tumor was radically resected, and effort was made to completely remove the tumor. Seven patients underwent surgical resection with hypothermic cardiopulmonary bypass. Nine patients underwent pulmonary resection, including pneumonectomy in eight patients and bilobectomy in one patient. As a surgical policy of our hospital, the operation of the heart and lung was divided into two stages when PA replacement was necessary.

Radiologic Evaluations of Thin-Section Computed Tomography Scan

Case No. 1 was operated on in 2007, so a thin-section CT scan was not performed. For the remaining nine patients, the findings of preoperative contrast thin-section CT scans were reviewed by the author (H.I.) and the tumor size was determined preoperatively based these findings. The size of the solid component was recorded as the maximum diameter in a single axial plane in the mediastinal window condition without a sharpening filter. In addition, all tumors were subsequently evaluated to estimate the tumor size using thin-section CT scans with a 2-mm collimator at our institute. We measured the distance from the site where the tumor was in contact with the PA to the surgical dissection line in all cross sections of the horizontal, sagittal, and coronal sections. The measurements for cases 2-10 are shown in Figures 1A-O.

The lung was photographed with a window level of 500-700 H and window depth of 1000-2000 H, which was labeled as the “lung window,” and a window level of 30-60 H and window depth of 350-600 H, which was labeled as the “mediastinal window.”

Pathological examination

The hematoxylin and eosin-stained slides of all patients were examined by a pathologist experienced in the field of soft tissue tumor pathology. Electron microscopy and immunohistochemistry for cytokeratins, S-100 protein, desmin, and smooth muscle actin were also performed.

The PAS sub-classification consisted of 12 classes [9–11]: undifferentiated, rhabdomyosarcoma, osteroid sarcoma, angiosarcoma, fibrosarcoma, malignant mesenchymoma, myxosarcoma, chondrosarcoma, osteosarcoma, malignant fibrous histiocytoma, liposarcoma, and unclassified leiomyosarcoma.

Follow-up and Statistical Analysis

The follow-up comprised a combination of outpatient visits and telephone calls. Overall survival was defined as the interval between the date of the operation and the date of death from any cause or the date of the last follow-up (March

1, 2021). Survival analysis was performed using the Kaplan-Meier method and log-rank test. All data were analyzed using IBM SPSS Statistics (version 23.0; IBM Corp., Armonk, NY, USA). This retrospective study was approved by the ethics committee of our institution (2020115).

Results

Patient characteristics

The preoperative patient characteristics are summarized in Table 1. Three (30%) patients were male, and the age at presentation ranged from 26-77 years, with a median age of 61 years. All patients were symptomatic, and the most common symptom was respiratory distress (4/10, 40%). The time from symptom onset to surgery was 1-11 months, with a median of 2.5 months. The range of tumor diameter was 40-92 mm, and the median was 70 mm. PET-CT was performed in four patients with three positive cases (3/4, 75%).

Intraoperative and Pathologic Results

The surgical operative details are summarized in Table 2. There were three cases in which only lung resection (right middle and lower lobectomy and left pneumonectomy) was completed. However, the final pathological results showed positive findings in all three cases. There were six cases in which PA vascular replacement was required in addition to pulmonary resection. One was a case in which aortic valve replacement was added. The final pathological results showed that three out of six cases had a negative margin. Seven patients had extracorporeal circulation, and the day after, there were three cases in which lung resection was performed in two stages. The median surgical time and blood loss were 363 min (range, 144-595 min) and 515 ml (range, 85-8100 ml), respectively.

The histological types of tumors are summarized in Table 3. Five different pathologic tumor types were observed in this series: tumors with undifferentiated pleomorphic sarcoma (n= 6, 60%), leiomyosarcoma (n=1, 10%), angiosarcoma (n=1, 10%), osteogenic sarcoma (n=1, 10%), and chondroblastic osteosarcoma (n=1, 10%). The positive rate of vascular stumps was 7/10. It was not possible to measure cases in which the surgical procedure was pulmonary endarterectomy (PE) (Case No. 7). Case No. 4 could not be measured because it infiltrated the PAs in the lungs on both sides. All patients with a stump distance of < 20 mm had a positive stump (4/4). The distance between the tumor and the dissection line during surgery needed to be at least 20 mm (Cases No. 9 and 10). However, although the distance between the tumor and the dissection line during surgery was 25 mm, one case with a positive stump was found (Case No. 6).

Early Outcomes

The median lengths of intensive care unit (ICU) and postoperative hospital stays in this series were 4 days (range, 2-13 days) and 15 days (range, 6-134 days), respectively. The most common complication was hoarseness in four patients (median hospitalization was 18.5 days). In these four patients, the median hospitalization of three cases without complications was longer than 7 days. There was one in-hospital death (10%) in the ICU from right heart failure on postoperative day (POD) 12.

Long-term follow-up

The postoperative outcomes are summarized in Table 4. Postoperative recurrence was observed in 6/9 cases, and the median recurrence period was as short as 9 months (range, 3-19 months). All six patients had intrathoracic recurrence in varying locations: in the lungs (Cases No. 3 and 7), in the PA (Cases No. 2, 4, and 6), and in the superior vena cava (Case No 5). We examined all cases using chest contrast CT to identify postoperative recurrence. One patient (Case No. 6) underwent PET-CT after surgery. It was performed 10 months after the first operation, and the SUVmax was 15.2 due to

stump recurrence (tumor size: 34 mm). Postoperative treatment was required in 4/9 cases (chemotherapy/radiotherapy/chemoradiotherapy = 1/2/1). The chemotherapy regimen was cisplatin and docetaxel for both. The radiation course was that the stump of the PA was irradiated with 2 Gy 25 times. After surgery, two cases used pazopanib. One patient (Case No. 3) died of PAS 2 months after using pazopanib. Another patient (Case No. 7), who had been using pazopanib for 6 months, still had a stable disease. The median survival was 11 months (range, 0.5-24 months). All causes of death were sarcoma, except for one case of perioperative death.

Discussion

This is the first study to measure the distance from the tumor to the dissection margin using preoperative contrast-enhanced CT. Based on our investigation, it was considered that the distance from the tumor to the dissection margin should be at least 20 mm. However, Case No. 6 had a distance of 25 mm with a positive dissected stump [12]. Since PAS propagates and proliferates in the intima of the PA, it is difficult to determine how far it has progressed by contrast-enhanced CT and PET-CT. In terms of pathological findings, the PAS extension site is 0.01-0.02 mm for the normal PA intima, whereas the PA intima for PAS is 0.1-0.2 mm, which is not as thick (Figures 2A and B). Siordia et al. reported that primary PAS is better treated with pneumonectomy than with PE, which is better suited for palliative treatment [13]. With regard to the pathological findings, it is difficult to make a macroscopic judgment during surgery, and we agree with the opinions of Siordia et al. Extended PAS emergency surgery is often performed to save lives to remove a tumor that is originally symptomatic and has grown. However, since there are reports of patients living for more than 5 years due to extended surgery [14], one indicator would be that the distance between the tumor and the stump during surgery is 20 mm.

We also reported that cardiovascular and lung surgeries are performed on different days for PAS surgery. The advantage is that in the case of one-stage surgery, a surgical heart-lung machine tends to cause bleeding, so a two-stage helps ensure a clear surgical field of view. In addition, lung surgery is easier for general thoracic surgeons than surgery with a median sternotomy, by performing a posterior lateral incision. The disadvantage is that it is complicated because the surgery is performed in two stages. For Case No. 10, we attempted to perform the operation in two stages, but on the night of 0 POD of cardiovascular surgery, the bleeding from the trachea did not stop and the operation was accelerated. We believe that two-stage surgery is more effective, for example, when there is a high probability of adhesions in the chest cavity.

Kim et al. reported that early detection contributes significantly to prognosis [15]. For every doubling of time from symptom onset to diagnosis, the odds of death increased by 46% [16]. However, this tumor is often misdiagnosed as acute or chronic pulmonary embolism because it is characterized by luminal obstruction and intraluminal growth. Gan et al. reported that the wall eclipsing sign on PA-CT angiography is pathognomonic for PAS [17]. Endovascular catheter biopsy and endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) may be used to diagnose PAS [18–20]. However, EBUS-TBNA increases the risk of bleeding and causes massive hemoptysis. PET-CT has been previously reported to be helpful in diagnostic workup [21, 22]. Ito et al. reported that the median maximum standardized uptake value of FDG is 7.63 in PAS and 2.36 in pulmonary embolism [23]. In our case, PET-CT was performed in four cases, with three positive cases. However, Case No. 8 was suspected of having pulmonary embolism because of a negative PET-CT, and was initially treated with heparin, but chest CT after 2 weeks showed no change in the PA intravascular nodule. Therefore, we suspected PAS and performed the surgery. PAS should be suspected if there is a large intravascular filling defect and no clinical improvement on anticoagulant therapy.

Hoarseness was observed in 40% (4/10) of the patients as a minor complication after surgery. The incidence of postoperative recurrent laryngeal nerve palsy is 0.077% in general surgery [24], but the incidence after cardiovascular surgery is as high as 1.9%-6.9% [25, 26]. Itagaki et al. reported that macrovascular surgery was associated with a high

risk (odds ratio 5.6) [27]. After branching from the vagus nerve, the recurrent laryngeal nerve travels posteriorly at the subclavian artery on the right and the ductus arteriosus on the left and ascends the tracheoesophageal groove. It then splits into anterior and posterior branches before reaching the upper edge of the cricoid cartilage. The causes of paralysis are thought to be indirect disorders, in addition to direct disorders of the recurrent laryngeal nerve due to surgical operations on large blood vessels. The first is due to compression of the recurrent laryngeal nerve. Traction of the subclavian artery is triggered by intraoperative repositioning, curvature of the endotracheal tube due to cervical extension, intraoperative operations such as sternum traction, displacement of the cuff position due to repositioning, gastric tube and transesophageal echocardiogram (TEE), and excessive sternum traction. The second is hypoperfusion of the recurrent laryngeal nerve feeding blood vessels due to peripheral circulatory insufficiency during use of the heart-lung machine and transient recurrent laryngeal nerve palsy due to hypothermia. In general, the risk factors for postoperative recurrent laryngeal nerve palsy include hypertension, diabetes, female sex, macrovascular surgery, heart-lung machine, TEE, long intubation, and surgery time. The median hospitalization time of patients with hoarseness complications was 18.5 days, which was longer than that in cases without complications. Although it is a major operation, it requires careful operation.

In this case, one perioperative death was due to right-sided heart failure. Kruger et al. reported an early postoperative mortality rate of 22% [7]. In other reports, perioperative mortality was 0–15%, with right heart failure and respiratory failure being the most common causes [15, 28–30]. In addition, Mussot et al. reported that two out of 31 cases required reoperation and two deaths were due to adult respiratory distress syndrome [31]. Based on this result, it should be remembered that surgery for PAS is major surgery.

Limitations

Our study had a few limitations. First, the sample size was small owing to the rarity of the original disease and was a retrospective observational analysis spanning nearly 15 years. Further research with a larger population and a longer follow-up period is necessary.

Conclusions

In conclusion, because postoperative chemotherapy and radiation therapy are likely to be ineffective, we may need to perform extended surgery as much as possible and have a distance of at least 20 mm between the location of the tumor on the CT and the surgical incision line during surgery for PAS. In addition, intensive care for intrathoracic recurrence follow-up is required for 1 year after surgery.

Declarations

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Author’s contributions

H.I., K.T., M.F., A.H., T.M., and K.S. conceived and planned the experiments. H.I., K.T., and A.H. planned and performed the simulations. H.I., T.M., and K.S. contributed to sample preparation. H.I., K.T., and K.S. interpreted the results.

H. I. took the lead in writing the manuscript. All authors provided critical feedback and helped shape the research, analysis, and manuscript.

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Conflict of Interest Statement

The authors declare no conflicts of interest.

Availability of data and materials

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

Ethics approval and consent to participate

This retrospective study was approved by the ethics committee of Juntendo University Hospital (2020115).

Consent for publication

Individual consent was waived.

Competing interests

The authors declare that they have no competing interests

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Tables

Table 1. Preoperative information on pulmonary artery sarcoma

No	Age	Sex	Symptoms	Time from symptom	Medical history	Tumor diameter	PET-CT
				to surgery (Months)		(mm)	(SUV max)
1	58	Female	Dyspnea	2	Rheumatoid arthritis	74	Not enforced
					Cervical cancer		
2	43	Male	Chest pain	11	Hypertension	40	9.4
3	49	Female	Fever	4	Cervical cancer	88	6.2
			Cough				
4	66	Female	Cough	4	None	70	Not enforced
5	75	Female	Hemoptysis	1	None	70	Not enforced
6	69	Male	Back pain	2	Hypertension	67	8.8
					Type 2 diabetes		
7	26	Female	Dyspnea	3	None	92	Not enforced
			Back pain				
8	77	Female	Dyspnea	2	Hypertension	40	Negative
					Type 2 diabetes		
9	64	Female	Dyspnea	3	Organized pneumonia	59	Not enforced
10	33	Male	Chest pain	1	None	75	Not enforced

Table 2. Intraoperative information on pulmonary artery sarcoma

No	First surgery content	Tumor localization	Extracorporeal circulation	Two-stage surgery	Surgery time (Minute)	Blood loss (Ml)
1	LPN, PAVR, AVR	LPA (C and P) PT, AV	Presence	No	595	440
2	LPN	LPA (C and P)	Absence	No	167	120
3	RPN, PAVR	RPA (C and P), PT	Presence	Yes	312±125	1110±235
4	RPN, PAVR	RPA (C and P), PT	Presence	Yes	428±96	1520±40
5	RPN, PAVR	RPA (C and P) PT, LPA(C)	Presence	Yes	296±120	2080±130
6	RMLL	RPA (P)	Absence	No	240	85
7	PAE	RPA (C), LPA (C)	Presence	No	240	100
8	LPN	LPA (C and P)	Absence	No	144	200
9	LPN, PAVR	RPA (C), LPA (C and P) PT	Presence	No	309	590
10	LPN, PAVR	RPA (C), LPA (C and P) PT	Presence	No	559	8100

RPN: right pneumonnectomy; LPN: left pneumonectomy;

PAVR: pulmonary artery vascular replacement; AVR: aortic valve replacement;

PAE: pulmonary artery endarterectomy; RMLL: right middle and lower lobectomy;

PT: pulmonary trunk; RPA: right pulmonary artery;

LPA: left pulmonary artery; AV: aortic valve; C: center; P: peripheral

Table3. Relationship between CT evaluation and pathology

No	Tumor diameter (mm)	Distance between tumor and the dissection line on CT (mm)	Pathology	Pathological stump results
1	74	20	Leiomyosarcoma	Negative
2	40	11	Undiffrentiated	Positive
3	88	10	Undiffrentiated	Positive
4	70	8	Undiffrentiated	Positive
5	70	11	Angiosarcoma	Positive
6	67	25	Undiffrentiated	Positive
7	92	Unmeasurable	Osteogenic sarcoma	Positive
8	40	6	Undiffrentiated	Positive
9	59	25	Condroblastic osteosarcoma	Negative
10	75	23	Undiffrentiated	Negative

Table 4. Postoperative information on pulmonary artery sarcoma

No	ICU	Hospital	Complications	Recurrence	Reccurence	Postoperative	Survival	Cause of death
	days	stays		(Months)	site	course		
						(Months)	(Months)	
1	12	12	Right heart	No	None	Right heart	Dead	Perioperative
			Failure			failure	(0.5)	death
2	2	6	None	Yes (19)	MPA, RPA	No PT	Dead	Cancer death
							(21)	
3	4	23	Hoarseness	Yes (10)	Lung, Liver	RT (2Gy×25) (3)	Dead	Cancer death
					Kidney, Bone	CT (10)	(13)	
						Pazopanib (11)		
4	13	33	Difficulty sputum	Yes (3)	MPA, RPA	CRT (3)	Dead	Cancer death
			PAF				(8)	
5	4	16	Hoarseness	Yes (8)	SVC, RA	No PT	Dead	Cancer death
							(8)	
6	2	7	None	Yes (5)	PA stump	Operation (11)	Dead	Cancer death
						Operation (19)	(24)	
7	5	12	None	Yes (10)	RUL	CT (3)	Alive	
						Operation (9)	(24)	
						Pazopanib (18)		
8	3	134	Takotsubo	No	None	RT (2Gy×25) (3)	Alive	
			Cardiomyopathy				(12)	
			Postoperative					
			myastenic crisis					
9	3	21	Hoarseness	No	None	No PT	Alive	
							(10)	
10	5	15	Hoarseness	No	None	No PT	Alive	
							(3)	

ICU: intensive care unit; PAF: paroxysmal atrial fibrillation; MPA: main pulmonary artery;

RPA: right pulmonary artery; SVC: superior vena cava; RA: right atrium; PA: pulmonary artery;

RUL: right upper lobe; PT: postoperative therapy; RT: radiotherapy; CT: chemotherapy;

CRT: chemoradiotherapy

Figures

Figure 1

How to measure tumor and dissection line

1A: Transverse plane of Case No 2, Arrow distance: 11 mm

1B: Transverse plane of Case No. 3

1C: Coronal plane of Case No. 3, Arrow distance: 10 mm

1D: Transverse plane of Case No. 4

1E: Sagittal plane of Case No. 4

1F: Transverse plane of Case No. 5

1G: Sagittal plane of Case No. 5, Arrow distance: 14 mm

1H: Transverse plane of Case No. 6, Arrow distance: 25 mm

1I: Coronal plane of Case No. 6

1J: Transverse plane of Case No. 7

1K: Transverse plane of Case No. 8, Arrow distance: 6 mm

1L: Transverse plane of Case No. 9

1M: Sagittal plane of Case No. 9, Arrow distance: 25 mm

1N: Transverse plane of Case No. 10

1O: Sagittal plane of Case No. 10, Arrow distance: 23 mm

Figure 2

A: Normal pulmonary artery (PA) structure B: PA structure of PA sarcoma