



Problems and countermeasures for surgical resection of primary pulmonary artery intimal sarcoma

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Background: Pulmonary artery intimal sarcoma (PAIS) is a rare but aggressive malignancy. This study clarified the problems and countermeasures of surgical treatment by examining surgical cases of PAIS.

Methods: Between January 2007 and October 2020, 10 patients with PAIS who underwent surgery at our hospital were retrospectively examined.

Results: The surgical procedure that aimed at complete resection was pulmonary resection only (three cases), along with pulmonary artery vascular replacement (six cases) and pulmonary endarterectomy (PE) (one case). The positive rate of vascular stumps was 7/10. In all cases, chest computed tomography scan showed positive margins of ≤ 20 mm between the tumor and surgical dissection (6/6). In addition, the distance between the location of the tumor 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 8/9 cases, and the median recurrence period was as short as 10 months (range, 3–19 months). Postoperative treatment was required in 7/9 cases (operation/chemotherapy/radiotherapy/chemoradiotherapy/heavy ion radiotherapy = 1/2/2/1/1). The median survival was 15 months (range, 0.5–36 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. The median postoperative recurrence period was as short as 10 months. Therefore, intensive care for intrathoracic recurrence follow-up is required for 1 year after surgery.

Keywords: Pulmonary artery intimal sarcoma (PAIS); surgical margin; treatment; survival; cardiopulmonary bypass

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Introduction

Pulmonary artery intimal sarcoma (PAIS) 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 (2-4). The age of onset of PAIS 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 PAIS are suboptimal. Krüger *et al.* reported that the median survival time without surgical resection is 1.5 months, whereas it is 10 months with surgery (7), and surgery is still considered necessary as a treatment policy. Interventions reported for PAIS 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 PAIS 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 PAIS patients at our institution, 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. We present the following article in accordance with the STROBE reporting checklist (available at <https://tcr.amegroups.com/article/view/10.21037/tcr-22-1945/rc>).

Methods

Study population

We retrospectively reviewed the clinical records of all patients who could be identified through a computer-assisted search of all medical records with a pathological diagnosis of PAIS, primarily diagnosed at the Juntendo University Hospital. From January 2007 to December 2020, 10 patients underwent surgery for PAIS 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, operable findings, postoperative courses, and long-term results. They all underwent contrast-enhanced CT before surgery. Case No. 1 was operated on in 2007; therefore, a thin-section CT scan was not performed. The remaining nine patients underwent a thin-section CT. The follow-up comprised a combination of outpatient visits and telephone calls. We defined early outcomes as pre-discharge and long-term outcomes as post-discharge. 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, 2022).

We excluded the following patients from this study: (I) patients with lung parenchymal sarcomas; (II) patients with metastatic sarcomas of the pulmonary vessels and (III) patients with inoperable primary PAISs in a fully palliative situation. This retrospective study was approved by the ethics committee of Juntendo University Hospital (No. 2020115), and was conducted in accordance with the Declaration of Helsinki (as revised in 2013). Informed consent for this retrospective research was waived.

Surgical techniques

The operations were performed via thoracotomy and median sternotomy in three and seven patients, respectively. The diagnosis of PAIS was histologically proven by frozen section biopsy during the operation. After confirmation of PAIS, the tumor was radically resected, and effort was made to completely remove the tumor. Seven patients underwent

Highlight box

Key findings

- The distance between the location of the pulmonary artery intimal sarcoma (PAIS) on computed tomography and the dissection line during surgery needed to be at least 20 mm.
- The median postoperative recurrence period was as short as 10 months.

What is known and what is new?

- It is known that complete resection of PAIS by surgery has a good prognosis, but there are no papers aiming to predict complete resection using preoperative chest CT.
- This is the first study to measure the distance from the tumor to the dissection margin using preoperative contrast-enhanced CT.

What is the implication, and what should change now?

- Intensive care for intrathoracic recurrence follow-up is required for 1 year after surgery even in patients with complete resection pathologically.

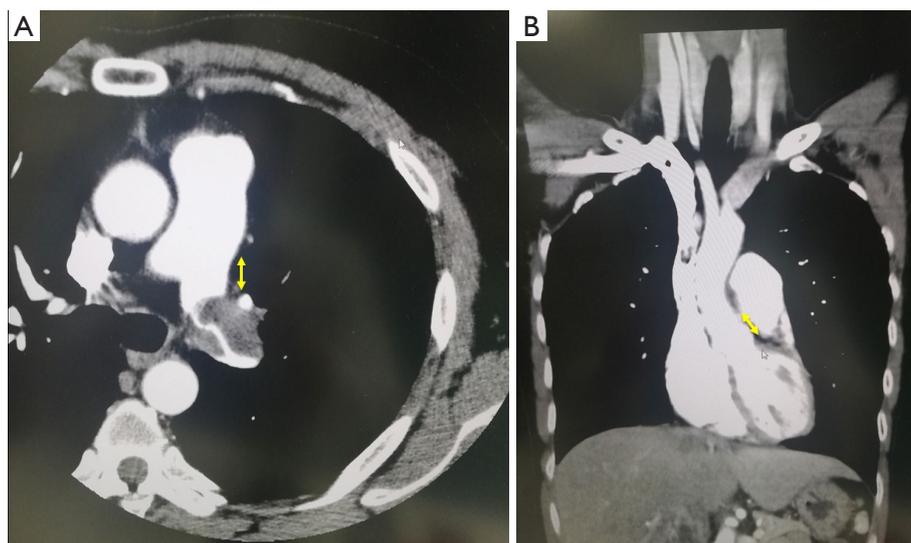


Figure 1 How to measure tumor and dissection line. (A) Transverse plane of Case No. 2, arrow distance: 11 mm. (B) Coronal plane of Case No. 3, arrow distance: 10 mm.

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

The findings of preoperative contrast thin-section CT scans were reviewed by the author (HI) and the tumor size was determined preoperatively based on 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 and 3 are shown in *Figure 1A,1B*.

The lung was photographed with a window level of 500–700 H and window depth of 1,000–2,000 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 thoracic tissue tumor pathology. Microscopy and immunohistochemistry for cytokeratins, S-100 protein, desmin, CD34, CD31, Factor VIII and smooth muscle actin were also performed.

We diagnosed PAIS according to the 2021 WHO classification (9). PAIS are malignant mesenchymal neoplasms arising in the large vessels of the pulmonary circulation, which are composed of spindle cells with varying degrees of atypia and variable cellularity.

Statistical analysis

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).

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

Table 1 Preoperative information on pulmonary artery intimal sarcoma

No.	Age (years)	Sex	Symptoms	Time from symptom to surgery (months)	Medical history	Tumor diameter (mm)	PET-CT (SUVmax)
1	58	Female	Dyspnea	2	Rheumatoid arthritis Cervical cancer	74	Not enforced
2	43	Male	Chest pain	11	Hypertension	40	9.4
3	49	Female	Fever, cough	4	Cervical cancer	88	6.2
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, type 2 diabetes	67	8.8
7	26	Female	Dyspnea, back pain	3	None	92	Not enforced
8	77	Female	Dyspnea	2	Hypertension, type 2 diabetes	40	Negative
9	64	Female	Dyspnea	3	Organized pneumonia	59	Not enforced
10	33	Male	Chest pain	1	None	75	Not enforced

PET-CT, positron emission tomography-computed tomography; SUVmax, maximum standardized uptake value.

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 3/6 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, 85–8,100 mL), respectively.

The CT findings and pathological margin results are summarized in *Table 3*. In this series, 4/10 cases were observed to have a very small amount of three different morphological components: a leiomyosarcomatous component (No. 1), an osteosarcomatous component (No. 7, 9) and a chondroblastomatous component (No. 8). 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 (6/6). 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 8/9 cases, and the

Table 2 Intraoperative information on pulmonary artery intimal 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*	1,110+235*
4	RPN, PAVR	RPA (C and P), PT	Presence	Yes	428+96*	1,520+40*
5	RPN, PAVR	RPA (C and P), PT, LPA (C)	Presence	Yes	296+120*	2,080+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	8,100

*, first surgery + second surgery. LPN, left pneumonectomy; PAVR, pulmonary artery vascular replacement; AVR, aortic valve replacement; LPA, left pulmonary artery; C, center; P, peripheral; PT, pulmonary trunk; AV, aortic valve; RPN, right pneumonectomy; PAE, pulmonary artery endarterectomy; RMLL, right middle and lower lobectomy; RPA, right pulmonary artery.

Table 3 Relationship between CT evaluation and pathological stump results

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

CT, computed tomography.

median recurrence period was 10 months (range, 3–19 months). All eight patients had intrathoracic recurrence in varying locations: in the lungs (Cases No. 3 and 7), in the PA (Cases No. 2, 4, 6, 9, and 10), 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 7/9 cases (operation/chemotherapy/radiotherapy/chemoradiotherapy/heavy ion radiotherapy =1/2/2/1/1). The chemotherapy regimens included cisplatin and doxorubicin in two cases (No. 3 and No. 7) and only paclitaxel in one case because of the patients’ concomitant heart failure (No. 10). The radiation course involved irradiation of the stump of the PA with 2 Gy 25 times. After surgery, two cases used pazopanib. One patient (Case No. 3) did not respond to pazopanib and died from progression of PAIS due to poor performance status 2 months after using pazopanib. Another patient (Case No. 7), who had been using pazopanib for 18 months, still had a stable disease.

The median survival was 15 months (range, 0.5–36 months). The mean recurrence times for margin-positive and margin-negative cases were 9.6 months (range, 3–19 months) and 13.5 months (range, 9–18 months), respectively. The mean survival times for margin-positive and margin-negative cases were 19.1 months (range, 8–36 months) and 19.5 months (range, 17–22 months), respectively. All causes of death were sarcoma, except for one case of perioperative death. No patient was lost to follow-up and no patients switched to another hospital.

Discussion

This is the first study to measure the distance from the

Table 4 Postoperative information on pulmonary artery intimal sarcoma

No.	ICU days	Hospital stays	Complications	Recurrence [months]	Recurrence site	Postoperative course [months]	Survival [months]	Cause of death
1	12	12	Right heart failure	No	None	Right heart failure	Dead [0.5]	Perioperative death
2	2	6	None	Yes [19]	MPA, RPA	No PT	Dead [21]	Cancer death
3	4	23	Hoarseness	Yes [10]	Lung, Liver Kidney, Bone	RT (2 Gy ×25) [3], ChT [10], Pazopanib [11]	Dead [13]	Cancer death
4	13	33	Difficulty sputum, PAF	Yes [3]	MPA, RPA	CRT [3]	Dead [8]	Cancer death
5	4	16	Hoarseness	Yes [8]	SVC, RA	No PT	Dead [8]	Cancer death
6	2	7	None	Yes [5]	PA stump	Operation [11], operation [19]	Dead [24]	Cancer death
7	5	12	None	Yes [10]	RUL	ChT [3], operation [9], Pazopanib [18]	Alive [36]	
8	3	134	Takotsubo cardiomyopathy, postoperative myasthenic crisis	No	None	RT (2 Gy ×25) [3]	Alive [24]	
9	3	21	Hoarseness	Yes [18]	PA stump	HIR [18]	Alive [22]	
10	5	15	Hoarseness	Yes [9]	MPA, RPA	ChT [9], RT [3 Gy ×15] [10]	Alive [15]	

ICU, intensive care unit; MPA, main pulmonary artery; RPA, right pulmonary artery; PT, postoperative therapy; RT, radiotherapy; ChT, chemotherapy; PAF, paroxysmal atrial fibrillation; SVC, superior vena cava; CRT, chemoradiotherapy; RA, right atrium; PA, pulmonary artery; RUL, right upper lobe; HIR, heavy ion radiotherapy.

tumor to the dissection margin using preoperative contrast-enhanced CT. Based on our investigation, the positive rate of vascular stumps was 7/10, and all patients with a stump distance of ≤ 20 mm had a positive stump (6/6). The distance between the tumor and the dissection line during surgery needed to be at least 20 mm. However, Case No. 6 had a distance of 25 mm with a positive dissected stump. Since PAIS 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 PAIS extension site is 0.01–0.02 mm for the normal PA intima, whereas the PA intima for PAIS is 0.1–0.2 mm, which is not as thick (Figure 2A,2B). Siordia *et al.* reported that primary PAIS is better treated with pneumonectomy than with PE, which is better suited for palliative treatment (10). 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 PAIS 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 (11), 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 PAIS 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 (12). For every doubling of time from symptom onset to diagnosis, the odds of death increased by 46% (13). 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 (14). Endovascular catheter biopsy and endobronchial

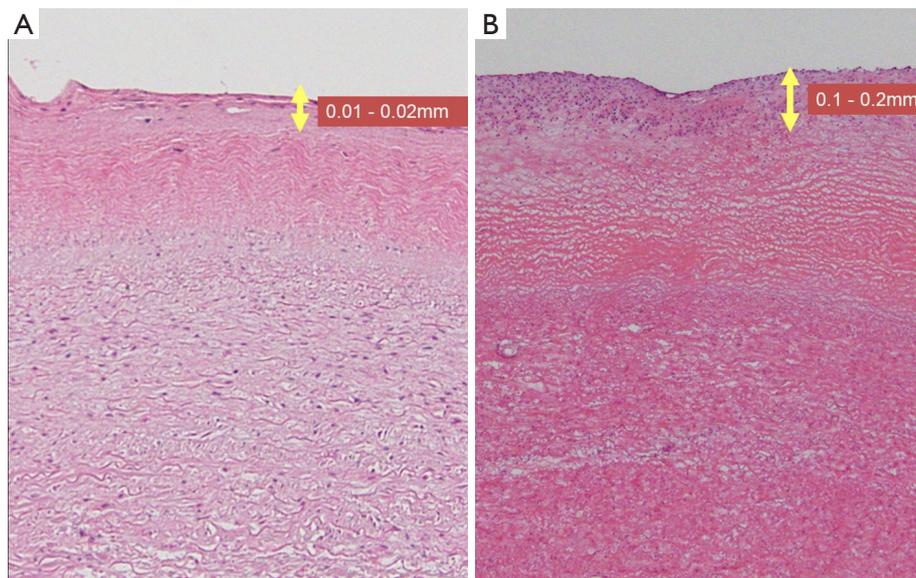


Figure 2 Differences in microscope images between normal PA and PAIS. (A) Normal PA structure (100× haematoxylin and eosin stain). (B) PA structure of PAIS (100× haematoxylin and eosin stain). PA, pulmonary artery; PAIS, pulmonary artery intimal sarcoma.

ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) may be used to diagnose PAS (15). However, EBUS-TBNA increases the risk of bleeding and causes massive hemoptysis. PET-CT has been previously reported to be helpful in diagnostic workup (16). Ito *et al.* reported that the median maximum standardized uptake value of FDG is 7.95 in PAIS and 2.37 in pulmonary embolism (17). 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 PAIS and performed the surgery. PAIS should be suspected if there is a large intravascular filling defect and no clinical improvement on anticoagulant therapy. Suto *et al.* reported that some PAISs with low cellular densities and high mucous tissue proportions have SUVs similar to those in pulmonary thromboembolism (18). Therefore, when we re-evaluated the pathology of Case No. 8, pathologic examination of PAIS revealed low-grade components with myxoid change admixed with high-grade components.

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. In recent years, there have been conspicuous reports that pazopanib is effective for PAIS, and it is

possible that the prognosis of patients with PAIS has improved. Second, this study is a single center case series. Therefore, further retrospective research with a higher number of patients at other institutions and a longer follow-up period is necessary.

Conclusions

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 PAIS. In addition, intensive care such as performing frequent chest CT is required to detect recurrence, considering that there are numerous intrathoracic recurrences with 1 year after surgery.

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Footnote

Reporting Checklist: The authors have completed the STROBE reporting checklist. Available at <https://tcr.amegroups.com/article/view/10.21037/tcr-22-1945/rc>

Data Sharing Statement: Available at <https://tcr.amegroups.com>

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <https://tcr.amegroups.com/article/view/10.21037/tcr-22-1945/coif>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. This retrospective study was approved by the medical ethics committee of Juntendo University Hospital (No. 2020115), and was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The informed consent for this retrospective research was waived.

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