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ORIGINAL ARTICLE / ÖZGÜN MAKALE

Results of surgical treatment of pulmonary artery sarcomas: Does histology affect survival?

Pulmoner arter sarkomlarının cerrahi tedavi sonuçları: Histoloji sağkalımı etkiler mi?

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ABSTRACT

Background: In this study, we aimed to describe our experience with primary pulmonary artery sarcoma in patients who underwent pulmonary endarterectomy and to evaluate clinical features, treatment, outcomes, and survival rates according to the histological subtypes of this malignant disease.

Methods: Between March 2011 and May 2022, a total of 13 patients (7 males, 6 females; mean age: 52.6±13.0 years; range, 30 to 69 years) who underwent pulmonary endarterectomy and diagnosed with a pulmonary artery sarcoma were retrospectively analyzed. The diagnosis was confirmed histopathologically in all patients. Data including demographics, clinical characteristics, intra- and postoperative complications, length of hospital stay, morbidity, mortality, and short-term and long-term outcomes were recorded. Operative mortality was defined as death in the hospital or within 30 days of surgery.

Results: Mortality was observed in one patient due to massive hemoptysis. Morbidity developed in two patients due to acute respiratory distress. Pulmonary vascular resistance improved significantly from 508 dyn/s/cm^{-5} to 191 dyn/s/cm^{-5} ($p<0.004$). All patients received chemotherapy following surgery. Median follow-up was 14 months. Median survival for the entire series was 18 months. One-year and three-year survival rates were 60.6% and 30.3%, respectively. Median survival for leiomyosarcomas (n=6) was seven months, while it was 44 months for intimal sarcomas ($p=0.004$). Three-year survival was 66.7% for intimal sarcomas and 0% for leiomyosarcomas.

Conclusion: Pulmonary artery sarcoma may mimic chronic thromboembolic pulmonary hypertension. Patients with a suspected diagnosis of pulmonary artery sarcoma should be referred to expert pulmonary endarterectomy centers for surgery where a multidisciplinary team is available. Pulmonary endarterectomy has both diagnostic and therapeutic value and may improve survival and quality of life. Patients with intimal sarcoma have longer survival compared to those with leiomyosarcoma.

Keywords: Malignancy, pulmonary artery sarcoma, pulmonary endarterectomy, sarcoma.

ÖZ

Amaç: Bu çalışmada pulmoner endarterektomi yapılan hastalarda primer pulmoner arter sarkomuna ilişkin deneyimlerimiz sunuldu ve bu malign hastalığın histolojik alt tiplerine göre klinik özellikleri, tedavisi, sonuçları ve sağkalım oranları değerlendirildi.

Çalışma planı: Mart 2011 - Mayıs 2022 tarihleri arasında pulmoner endarterektomi yapılan ve pulmoner arter sarkomu tanısı konan toplam 13 hasta (7 erkek, 6 kadın; ort. yaş: 52.6±13.0 yıl; dağılım, 30-69 yıl) retrospektif olarak incelendi. Tanı tüm hastalarda histopatolojik olarak doğrulandı. Demografik özellikler, klinik özellikler, ameliyat sırası ve sonrası komplikasyonlar, hastanede kalış süresi, morbidite, mortalite ve kısa dönem ve uzun dönem sonuçlar kaydedildi. Cerrahi mortalite, hastanede veya ameliyattan sonra 30 gün içinde gerçekleşen ölüm olarak tanımlandı.

Bulgular: Bir hastada masif hemoptizi nedeniyle mortalite gözlendi. İki hastada akut solunum sıkıntısına bağlı morbidite gelişti. Pulmoner vasküler direnç, anlamlı ölçüde 508 dyn/s/cm^{-5}'den 191 dyn/s/cm^{-5}'e geriledi ($p<0.004$). Ameliyattan sonra tüm hastalar kemoterapi aldı. Medyan takip süresi 14 ay idi. Medyan sağkalım 18 ay idi. Bir ve üç yıllık sağkalım sırasıyla %60.6 ve %30.3 idi. Medyan sağkalım leiomiyosarkom (n=6) için yedi ay iken, intimal sarkom için 44 ay idi ($p=0.004$). Üç yıllık sağkalım intimal sarkom için %66.7 ve leiomiyosarkom için %0 idi.

Sonuç: Pulmoner arter sarkomu, kronik tromboembolik pulmoner hipertansiyonu taklit edebilir. Pulmoner arter sarkom tanısından şüphelenilen hastalar, multidisipliner ekibin olduğu bir uzman pulmoner endarterektomi merkezine sevk edilmelidir. Pulmoner endarterektomi hem tanısal hem de terapötik değere sahiptir ve sağkalım ile yaşam kalitesini iyileştirebilir. Leiomiyosarkomlu hastalara kıyasla, intimal sarkomlu hastaların sağkalım süresi daha uzundur.

Anahtar sözcükler: Malignite, pulmoner arter sarkomu, pulmoner endarterektomi, sarkom.

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Pulmonary artery sarcomas (PASs) are extremely rare tumors with an incidence of 0.001 to 0.03%.[1] They originate from the intima of the pulmonary arteries and tend to spread within the pulmonary vasculature. Since the first description of pulmonary artery sarcoma by Mandelstam[2] in 1923,[1,3] only a few hundred cases have been described in the literature. It also has a fatal outcome mimicking chronic thromboembolic pulmonary hypertension (CTEPH), which eventually leads to right heart failure. Diagnosis is difficult due to its insidious clinical course and remarkable similarity to CTEPH in terms of symptoms and radiological findings. Moreover, PAS and CTEPH are classified as Group 4 pulmonary hypertension according to the World Symposium on Pulmonary Hypertension classification.[4] Pulmonary endarterectomy (PEA) is the only curative treatment for CTEPH.[5]

Despite novel imaging and interventional techniques, it remains difficult to diagnose PAS. Although, surgery still is the mainstay of management of patients with PAS, there is no widely accepted surgical approach until now. Surgery includes PEA, lobectomy, or pneumonectomy, based on the extension of the disease and patient clinical conditions. Surgery can provide a median survival of up to eight to 36 months; however, the prognosis of PAS is very poor with a median survival of 1.5 months without surgery.[1,3]

In the present study, we aimed to describe our experience with PAS in patients who underwent PEA, and evaluate clinical features, treatment, outcomes, and survival rates according to the histological subtypes of this malignant disease.

PATIENTS AND METHODS

This single-center, retrospective study was conducted at University of Health Sciences, Koşuyolu High Specialization Education and Research Hospital, Department of Thoracic and Cardiovascular Surgery between March 2011 and May 2022. A total of 871 consecutive patients underwent PEA in our institution during the study period. Of these patients, 13 (7 males, 6 females; mean age: 52.6±13.0 years; range, 30 to 69 years) who underwent surgery and were eventually diagnosed with PAS were included. We reported our first case in 2014.[6] The medical records of these patients were retrospectively reviewed from a prospective database and data including demographics, clinical characteristics, intra- and postoperative complications, length of hospital stay, morbidity, mortality, and short-term and long-term outcomes were recorded. Operative mortality was defined as death in the hospital or within 30 days of surgery.

The diagnosis of CTEPH was established by the presence of mismatched perfusion defects on ventilation-perfusion scan combined with evidence of pulmonary hypertension, despite adequate anticoagulation for at least three months. Pulmonary function tests, computed tomography pulmonary angiography (CTPA), six-minutes walk test (6MWT), and right heart catheterization were routinely performed as a standard preoperative evaluation. All patients were assessed by our multidisciplinary team of CTEPH experts including expert PEA surgeons, pulmonologists, cardiologists, radiologists, as well as rheumatologists., and all care was provided including detailed diagnostics and all forms of CTEPH therapy. In our center, the following tests are routinely performed during assessment for surgery: complete blood count, blood chemistry, urinalysis, and acute inflammatory markers. For patients with suspected PAS or systemic vasculitis, fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (PET/CT) was used. The definitive diagnosis was made based on a detailed histopathological examination. In nine patients, PET/CT was also performed, as malignancy was suspected. Our indications for PEA surgery can be listed as follows: patients with a mean pulmonary artery pressure (mPAP) of >20 mmHg, pulmonary vascular resistance (PVR) of 300 dyn/s/cm^{-5} or 3 Wood units, surgically accessible disease demonstrated in CTPA, and a World Health Organization (WHO)/ New York Heart Association (NYHA) functional class greater than II. Institutional standard protocols for management of perioperative anesthesia and postoperative care were followed for each patient. In-hospital complications were measured following surgery.

Our surgical technique was described previously.[5] Briefly, median sternotomy and cardiopulmonary bypass was performed. Most tumors were observed in the dorsal region of the main pulmonary artery often with a polypoid or finger-like form, extending to the bifurcation of the main pulmonary artery. Following a proximal incision of the main pulmonary artery, a circumferential dissection plane was established between the gross tumoral mass with intimal and the medial layer of the pulmonary artery and extended to the distal segmental and subsegmental branches. A true endarterectomy plan was found and bilateral PEA was performed in all patients under deep hypothermia

(20°C) and total circulatory arrest with intermittent cross-clamping of the aorta. Prolene sutures were used to close arteriotomy lines. No additional materials such as pericardial patch or grafts were required for pulmonary artery reconstruction. Concomitant surgical procedures were performed during re-warming period. Early postoperative hemodynamic measurements were completed in the operating room, before the patients were transferred to intensive care unit (ICU). No surgical margin was defined, since no resection was performed. No lung resection was done for PAS.

During follow-up, all patients were seen in our PEA outpatients' clinic at one, three, and six months. Patients were assessed with a 6MWT and echocardiography and were, then, classified functionally according to the WHO/NYHA functional class at each follow-up visit.

All patients had a histopathological diagnosis after surgery, and all survivors received adjuvant chemotherapy after recovering from the postoperative course. Recurrence and disease progression were defined as evidence of a new lesion on PA and metastasis to another part of the body, respectively.

Statistical analysis

Statistical analysis was performed using IBM SPSS for Windows version 25.0 software (IBM Corp., Armonk, NY, USA). Continuous data were expressed in mean ± standard deviation (SD) or median (min-max), while categorical data were expressed in number and

Table 1. Baseline and demographic characteristics of patients

Characteristics	n	%	Mean±SD
Age (year)			52±13.0
Sex			
Female	8	61.5	
Male	5	38.5	
Duration from symptom to surgery (month)			9.6±2.1
Symptoms			
Shortness of breath	13	100	
Fatigue	13	100	
Cough	8	61.5	
Headache	4	30.4	
Hemoptysis	1	7.6	
Syncope	1	7.6	
NYHA class			
I	0	0	
II	2	13.4	
III	9	69.2	
IV	2	13.4	
6MWT (meter)			224±167.7
FEV1 (L)			2.6±0.9
FEV1 (%)			85±16.2
FEV1/FVC			87.5±17.6
sPAP (mmHg)			63.9±30.4
mPAP (mmHg)			30.9±16.1
Cardiac index (L/min/m^2)			2.5±1.0
Cardiac output (L/min/m^2)			3.8±1.4
PVR (dyn/s/cm^{-5})			508±324.7
18F-FDG PET/CT (SUV_{max}) (n=8)			5.6±2.1

SD: Standard deviation; NYHA: New York Heart Association; 6MWT: Six-minutes walk test; FEV1: Forced expiratory volume in 1 sec; FVC: Forced vital capacity; sPAP: Systolic pulmonary arterial pressure; mPAP: Mean pulmonary arterial pressure; PVR: Pulmonary vascular resistance; 18F-FDG PET/CT: Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography; SUV_{max}: Maximum standard uptake value.

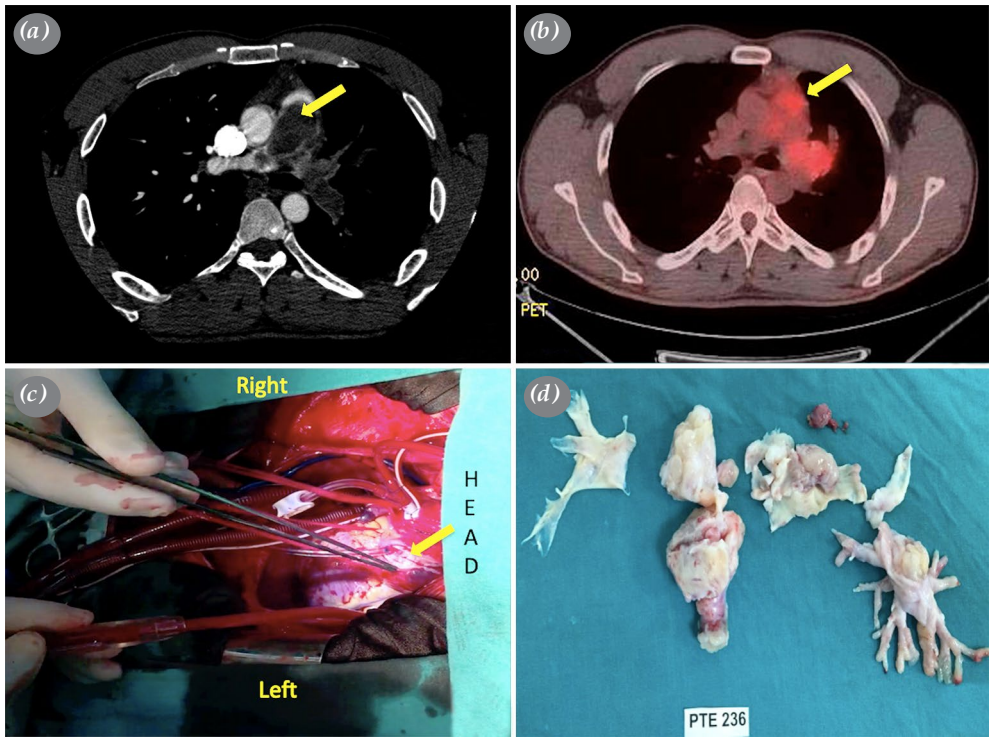

Figure 1. Intra- and postoperative images. **(a)** CTPA showing PAS lesions in the pulmonary arteries (arrow). **(b)** Image from 18F-FDG PET/CT showing hypermetabolic lesions in the pulmonary arteries (arrow). **(c)** Intraoperative image showing PAS inside right pulmonary artery. Forceps holds the pulmonary arterial wall and arrow shows the lesion after arteriotomy. **(d)** Postoperative specimen after PEA.

CTPA: Computed tomography pulmonary angiography; PAS: Pulmonary artery sarcoma; 18F-FDG PET/CT: Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography.

frequency. Pre- and postoperative measurements were tested by the Wilcoxon signed-rank test. Survival analysis was performed using the Kaplan-Meier method. The log-rank test was used to compare survival probabilities between two groups (i.e., those with the diagnosis of intimal sarcoma [IS] and those with leiomyosarcoma [LM]). A p value of <0.05 was considered statistically significant.

Table 2. Intra- and postoperative data of patients

Characteristics	n	%	Mean±SD
Cardiopulmonary bypass (min)			277±96.0
Aortic cross-clamp (min)			19.1±24.1
Total circulatory arrest (min)			28.8±13.3
ECMO (n)	3		
Mechanical ventilation time (days)			2.7±3.2
ICU (days)			5.2±4.1
LOS (days)			12.8±7.1
Postoperative mPAP (mmHg)			28.1±5.4
Postoperative PVR (dyn/s/cm^{-5})			191.5±74.7
Postoperative NYHA class I	10	83.3	

SD: Standard deviation; ECMO: Extracorporeal membrane oxygenation; ICU: Intensive care unit; LOS: Length of hospital stay; mPAP: Mean pulmonary arterial pressure; PVR: Pulmonary vascular resistance; NYHA: New York Heart Association.

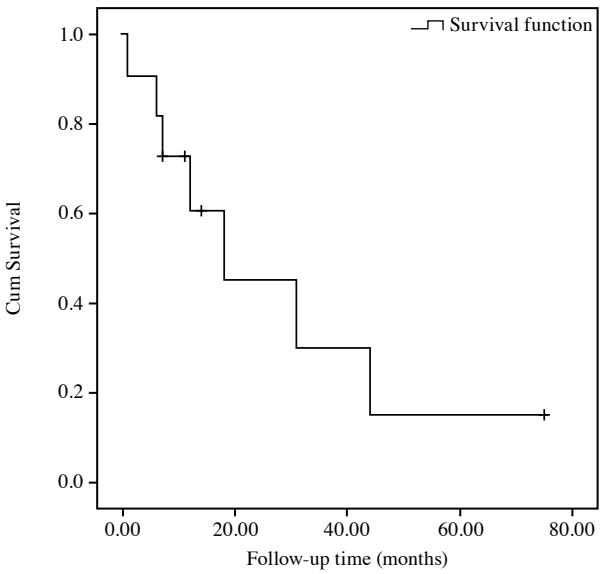

Figure 2. The Kaplan-Meier overall survival curve.

RESULTS

Baseline and demographic characteristics of the patients are shown in Table 1. Shortness of breath and fatigue were the chief complaints in all patients. The functional capacity of the patients was WHO/NYHA Class III-IV in 11 and Class II in two. Five patients had an increased uptake on PET/CT (Figure 1). None of the patients had a history of any type of malignancy and no chemotherapy or radiotherapy was used before the surgery.

Following surgery, the patients were kept intubated and transferred to the ICU, and both postoperative hemodynamic parameters and mean mPAP and PVR were closely monitored and calculated from the first postoperative day until transfer from the ICU to the ward. Intra- and postoperative data are summarized in Table 2. The 30-day mortality was observed in one patient (7.6%) who had massive hemoptysis following PEA.[6] Despite pacing on venous arterial extracorporeal membrane oxygenation (VA-ECMO) as a salvage therapy, the patient died on postoperative Day 10 due to right heart failure and reperfusion injury. In addition, two patients developed acute respiratory distress syndrome in the early postoperative period, and VA-ECMO was initiated. The patients were weaned from VA-ECMO on postoperative Day 5 and 9, respectively and were discharged without any problem.

The mPAP decreased from 30.9±16.1 (range, 18 to 75) mmHg to 28.1±5.4 (range, 21 to 35) mmHg after surgery (p=0.182). A significant difference was found in the mean PVR, which decreased from 508±324.7 (range, 168 to 1,200) dyn/s/cm^{-5} to 191.5±74.7 (range, 120 to 344) dyn/s/cm^{-5} after surgery (p=0.004). The median follow-up was 14 (range, 7 to 75) month. The median survival for the entire series was 18 (range, 6 to 82) months (95% confidence interval [CI]: 3.90-32.09). The one-year and three-year survival rates were 60.6% and 30.3%, respectively. While the median survival for LM (n=6) was seven months (95% CI: 4.85-9.14), patients with IS had a median survival of 44 months (95% CI: 23.19-64.80; (p=0.004).

Figure 2 shows the Kaplan-Meier overall survival curve. While six of the cases were diagnosed as LM, as they had histomorphological smooth muscle differentiation and positive immunoexpression of smooth muscle markers (such as desmin and smooth muscle actin), six other patients were classified as IS. One patient was also diagnosed as an undifferentiated sarcoma, as he had malignant pleomorphic spindle cells without any differentiation (Figure 3). According to the survival estimations, the median survival for LM was seven months, while the patients with IS had a median survival of 44 months. For LM and IS, the one-year survival rates were 20% and 100%, respectively. Figure 4 shows the

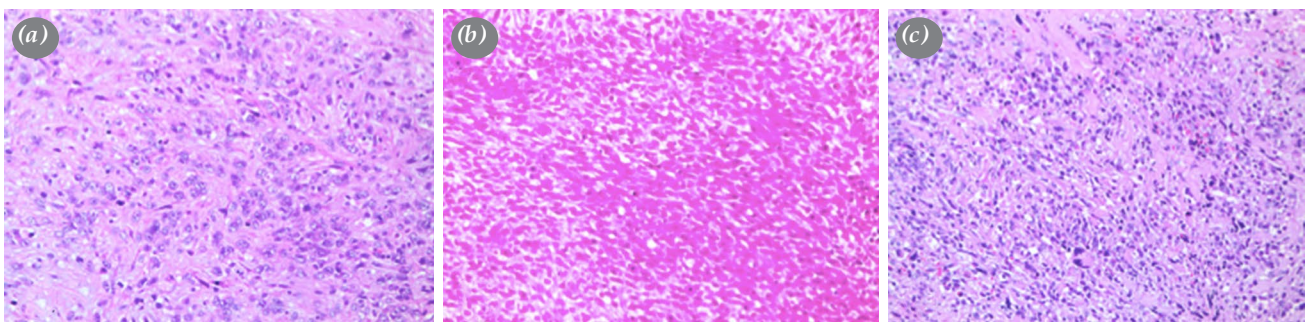

Figure 3. Histopathology images. **(a)** Intimal sarcoma. **(b)** Leiomyosarcoma. **(c)** Undifferentiated sarcoma (H&E, ×20).

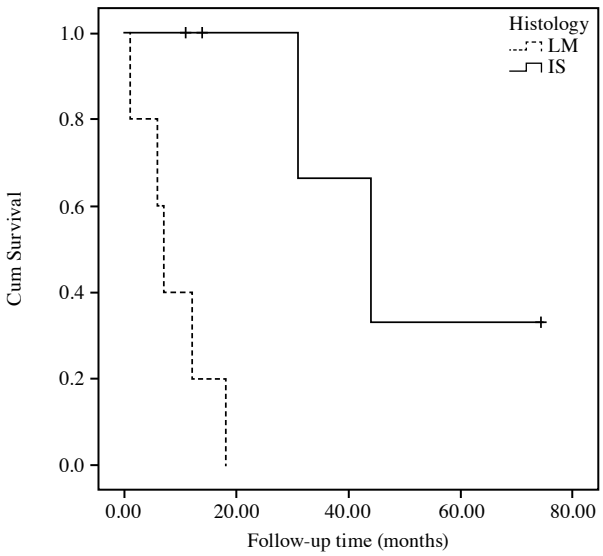

Figure 4. The Kaplan-Meier survival curve between histopathological types.
LM: Leiomyosarcoma; IS: Intimal sarcoma.

histology-specific survival curve. In addition, the three-year survival rate was 66.7% for IS and 0% for LM.

Apart from the patient who died in the postoperative 30-day period, six patients died during follow-up until the cut-off date of the study. Five of them died due to disease progression or recurrence, and one patient unfortunately died due to novel coronavirus disease 2019 (COVID-19). One patient required pneumonectomy for recurrence at four months after PEA and died at 44 months. The other six patients are still alive. The treatment and outcomes are shown in Table 3.

DISCUSSION

In the present study, we described 13 patients diagnosed with pulmonary artery sarcoma following PEA surgery. Clinical evidence of sarcoma was not observed preoperatively. Histologically, we identified LMs in six patients and ISs in seven patients. There was one in-hospital mortality in early period of our PEA experience. The mPAP and PVR values were reduced and symptomatic relief was achieved following PEA surgery. All patients received chemotherapy in postoperative period. The median survival for the entire series was 18 months. In this study, we observed that patients with diagnosis of IS had a better prognosis than that of LMs (44 *vs*. 7 months, respectively).

Pulmonary artery sarcoma is a rare condition with an incidence of 0.001 to 0.03% and its diagnosis is difficult due to its clinical similarity to acute or chronic pulmonary thromboembolism.[7,8] In our study, all patients with a possible diagnosis of CTEPH were treated, and all received effective anticoagulant therapy preoperatively for at least three months. The similarity between CTEPH and PAS complicates the

Table 3. Treatment and outcomes

Patient no	Operative procedure	Histological type	Adjuvant treatment	Recurrence/ progression/other	Survival (months)
1	PEA	Leiyomyosarcoma	None	None	Dead (n=0)
2	PEA	Leiyomyosarcoma	CT	Progression (brain)	Dead (n=7)
3	PEA+PFO closure	Intimal sarcoma	CT	Progression (liver, bone)	Dead (n=44)
4	PEA	Intimal sarcoma	CT	None	Alive (n=82)
5	PEA+CABG	Leiyomyosarcoma	CT	Recurrence	Dead (n=18)
6	PEA	Intimal sarcoma	CT	Recurrence	Dead (n=6)
7	PEA	Leiyomyosarcoma	CT	Progression (bone, brain)	Dead (n=12)
8	PEA	Intimal sarcoma	CT	Other (SARS-CoV-2)	Dead (n=31)
9	PEA	Intimal sarcoma	CT	None	Alive (n=19)
10	PEA	Intimal sarcoma	CT	None	Alive (n=16)
11	PEA	Undifferentiated sarcoma	CT	None	Alive (n=13)
12	PEA	Intimal sarcoma	CT	None	Alive (n=10)
13	PEA	Leiyomyosarcoma	CT	None	Alive (n=7)

PEA: Pulmonary endarterectomy; CT: Computed tomography; PFO: Patent foramen ovale; CABG: Coronary artery bypass grafting; SARS-CoV-2: Severe acute respiratory syndrome-coronavirus 2.

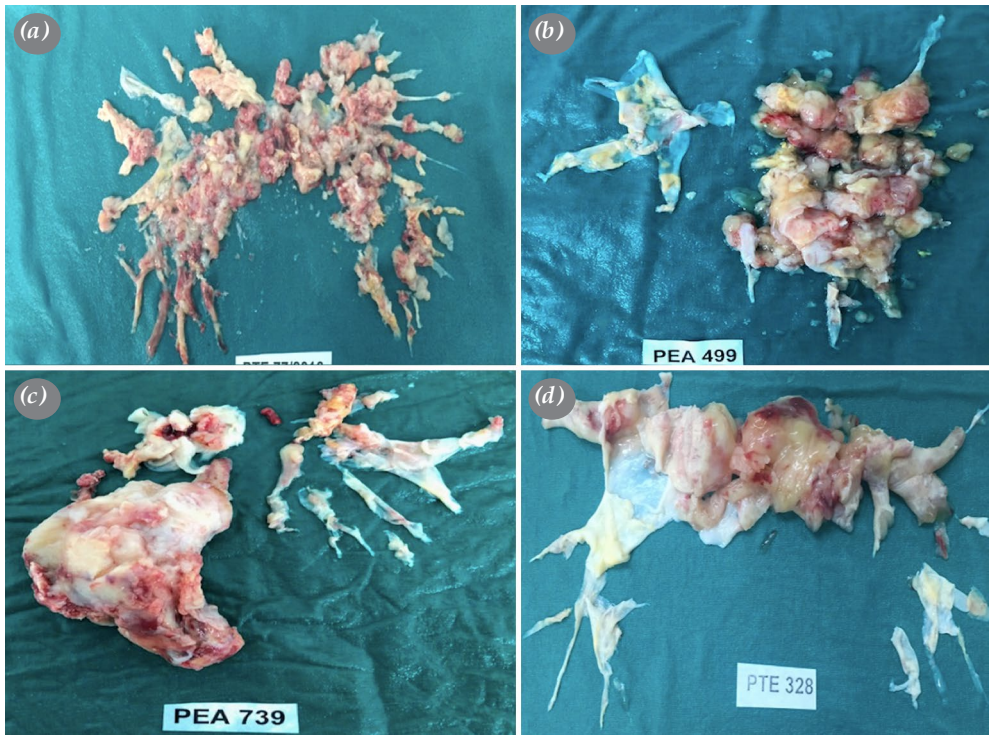

Figure 5. (a, b) Pulmonary endarterectomy specimens resected from some of the patients. **(c, d)** Gelous consistency. Rigid consistency.

diagnosis, management, and treatment. The tumor originating from the central pulmonary artery leads to progressive obstruction that impedes blood flow from the right ventricle to the pulmonary vascular bed. The CTPA is commonly used to distinguish between PAS and CTEPH, and although both show enhancement on CTPA, IS tends to present as a unilateral, central, lobulated, PA filling defect which increases in size along with the increasing diameter of the PAs and has acute angles with the vessel wall, in contrast to the pulmonary embolism. Moreover, IS shows heterogeneous densities due to areas of necrosis, hemorrhage, and ossification within the mass.[9] In addition, PET/CT can be used in the differential diagnosis of pulmonary sarcoma and CTEPH, as the standard pathology uptake values are different.[10] The CTPA is a routine practice in our center; however, nine patients underwent PET/CT in this study, five of whom had positive uptake. There was no significant difference between the histological types in terms of maximum standard uptake value (SUV_{max}) (p=0.629).

Pulmonary artery sarcoma is divided into two main types: intimal and intramural.[11] The former presents an intraluminal polypoid growth pattern.[12,13] On the other hand, the latter is different from IS and is classified separately as in soft tissue sarcoma (LM).[13] Since intimal PAS is more common than intramural PAS, PAS is often referred to as IS.[13] An IS resembles mucoid or gelatinous clots that fill vascular lumens, and the distal extension of the tumor may have firm fibrotic regions, be bony or gritty, and chondromyxoid foci may be present in mural lesions.[14] An IS may arise from the pluripotent mesenchymal cells of the intima artery, that is, the inner membrane of the pulmonary artery.[15] Therefore, PAS may have a variety of pathological types. Cox et al.[12] reported the pathological results of 138 cases of PAS, of which 43 were undifferentiated sarcomas. The typical histological features and immunohistochemical staining for PAS are important for classifying the different pathological types. Under the microscope, IS exhibits proliferation of spindle cells in a myxoid background.[16,17] The tumor may be also associated with large regional myxoid tissues and local necrosis, and typical spindle cells are arranged like a woven mat or striated, as in LM. Pulmonary artery IS is a malignant tumor that progresses rapidly. If the intervention is delayed, the prognosis is extremely poor and the recurrence rate is high.[18] Pulmonary artery LM is a rare disease. More recent reviews have suggested a predominance of two to one in women, with a median age at the time of diagnosis, ranging

between 49 and 52 years.[17] Median survival without surgery is one and a half months, and mortality is usually due to right-sided heart failure.

In the current study, survival probabilities were significantly different according to the histological type (p=0.004). The median survival time for LM was seven months, while IS had a median survival time of 44 months. In addition, the one-year survival rate was 20% for LM and 100% for IS.

There is no established treatment protocol for PAS. Currently, the main treatment for PAS is resection of sarcoma. Compared to isolated tumor resection, PEA seemed to yield a better survival rate and alleviate symptoms.[7,8,17,18] Although there is no consensus on surgical treatment, PEA and pneumonectomy are the two most common surgical procedures. Grazioli et al.[18] compared PEA and pneumonectomy in the surgical treatment of 13 patients PAS and reported a median survival of 6.6 months and 26.8 months, respectively. On the other hand, Jamieson[19] reported that pneumonectomy was not always beneficial in the treatment of the disease, as most cases are bilateral. We performed PEA to perform complete tumor resection in all patients, and the surgical approach was performed according to our published technique.[5] Pulmonary artery sarcoma usually arises within the main pulmonary arteries or around the pulmonary valve and often invades adjacent structures, such as the right ventricular outflow tract. This proximal extent of the tumor makes PEA more technically challenging compared to PEA for CTEPH. In addition, the consistency of the tumor can vary, presenting as either gelatinous or rigid material, and both variations lead to difficulty in finding the correct plane of dissection (Figure 5).

Mussot et al.[8] reported 31 patients with PAS. In this study, 25 patients were treated with PEA and five patients with pneumonectomy. In addition, one patient required right pulmonary artery replacement. The 30-day mortality was 13% and one-, three-, and five-year survival rates were 63%, 29%, and 22%, respectively. In our study, only one patient had 30-day mortality (7.6%), and the median survival for the entire series was 18 months. Our one-year survival rate is 60.6%, consistent with previous studies.

Even after complete surgery, the prognosis of IS is poor due to frequent relapses, more often local than distant. A significant symptomatic improvement among PAS patients with endarterectomy has been well established and accepted. On the other hand, the knowledge on the benefit of adjuvant and/or neo-adjuvant therapy is still controversial. Some researchers believe that chemotherapy or radiotherapy has no effect on long-term survival, whereas the others suggest that it can prolong survival.[1,8,20] Adjuvant therapy is often used either alone or in combination with radiotherapy. As in lung cancer, if lung resection is planed surgical approach, neoadjuvant treatment with radiotherapy and/or chemotherapy may be considered to decrease tumor size and improve resectability. Xu et al.[21] reported a meta-analysis including 275 cases. Although surgical resections were different, patients who received adjuvant and/or neo-adjuvant therapy had longer survival rates. In addition, patients with complete resection or without metastasis had longer postoperative overall survival compared to incomplete resection or with metastasis. It has been reported that chemotherapy prolongs the postoperative survival in patients who had incomplete resection and metastasis [18] While Mussot et al.[8] found no significant difference between the patients with and without postoperative chemotherapy in their series, Blackmon et al.[22] reported better outcomes in the patients treated with a combination of chemotherapy and radiotherapy. Wang et al.,[23] also reported similar results. We preferred cisplatin as the chemotherapeutic agent for adjuvant therapy in all our patients in the study, and none of them received radiotherapy postoperatively, since no lung resection was performed. Even if treated, the postoperative survival rates are still poor, with a median overall survival ≤18 months.

Due to the shorter follow-up and the limited number of patients in our series, we cannot definitively prove that patients with IS had longer survival compared to LM. Nonetheless, although this is a retrospective study, our data represent a valuable contribution to the literature, considering how rare this disease is and how little experience there is with its treatment.

In conclusion, pulmonary artery sarcoma is a very rare vascular tumor with a poor prognosis. Its remarkable clinical and radiological similarity to chronic thromboembolic pulmonary hypertension makes diagnosis difficult. Although pulmonary endarterectomy is the mainstay of treatment, aggressive adjuvant therapy after surgery is necessary to improve survival and quality of life. Therefore, all patients suspected of pulmonary artery sarcoma should be referred to a referral center to be examined by an experienced multidisciplinary team. Although, both intimal sarcoma and leiomyosarcoma have a poor prognosis, the intimal sarcoma subtype seems to have

a better short-term outcome. Further well-designed, large-scale, prospective studies are needed to develop an established treatment algorithm in this patient population.

Ethics Committee Approval: The study protocol was approved by the Kartal Kosuyolu High Specialization Training and Research Hospital Clinical Research Ethics Committee (date: 09.02.2021, no: 2016-KAEK-112-475). The study was conducted in accordance with the principles of the Declaration of Helsinki.

Patient Consent for Publication: A written informed consent was obtained from each patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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