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Surgical treatment of pulmonary arteriovenous malformations

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Background/aim: Pulmonary arteriovenous malformations (PAVMs) are direct communications between the branches of pulmonary arteries and veins. This study evaluates surgically treated cases of pulmonary arteriovenous malformations.

Materials and methods: We retrospectively examined 41 cases of PAVM that were operated in our clinic between 1995 and 2012. We obtained the clinical, radiological, and surgical data of the patients from their files.

Results: The 41 cases comprised 27 males and 14 females. Their mean age at diagnosis was 39.8 years (range: 9–71). The symptoms were hemoptysis in 28 cases, dyspnea in five, cough in three, and epistaxis in two; three patients were asymptomatic. Twenty-three right and 19 left posterolateral thoracotomies were performed, including one case which was operated bilaterally. Lower lobectomy was performed in 17 patients, lower lobectomy and lingulectomy in two, upper lobectomy in ten, middle lobectomy in two, segmentectomy in seven, and wedge resection in four. Postoperative histopathology was arteriovenous malformation in all cases.

Conclusion: PAVMs are rare clinical conditions. Surgery remains the first choice when embolization treatment cannot be performed or is not successful, in symptomatic and complicated patients with PAVM, and/or in cases where the PAVM diagnosis cannot be established.

Key words: Arteriovenous malformation, lung, surgery

1. Introduction

Pulmonary arteriovenous malformations (PAVMs) are direct communications between the branches of pulmonary arteries and veins, without an intervening pulmonary bed (1). In 1897, Churton first described PAVM in a 12-year-old boy (2). PAVM has been variously called pulmonary arteriovenous fistula, aneurysm, pulmonary angioma, arteriovenous angiomatosis, cavernous hemangiomas, and telangiectasia. PAVM is the preferred term, however, as it represents a developmental anomaly (1,3). Although it is the most common anomaly of the pulmonary vascular bed, PAVM remains an extremely rare clinical formation. The incidence in the general population is still not fully known, but it is estimated at 2–3 cases per 100,000. PAVMs are detected most commonly in the fifth and sixth decades; 10% are seen in infants and children (1). Patients with PAVM may be asymptomatic or may present with dyspnea, paradoxical thromboembolism, brain abscess, or massive hemoptysis. While more than 80% of PAVM cases are congenital, 47%–80% are also associated with hereditary hemorrhagic telangiectasia (HHT) or Rendu–Osler–Weber syndrome (1,3).

In 1940, Shenstone performed what is considered to be the first excisional procedure, a pneumonectomy for a large, centrally located lesion, which was reported by Hepburn and Dauphinee in 1942 (4). Since then, although surgery has been the only treatment approach for PAVM, following a report by Taylor et al. in 1978 of successful percutaneous transcatheter embolization in one case, embolization has gradually replaced surgery (1,3). Herein, we present a review of PAVM patients that underwent surgery in our clinic.

2. Materials and methods

We retrospectively reviewed the pathological records of surgical procedures performed in our department from 1995 to 2012, and we identified 41 patients with PAVM. Those cases were reviewed for demographic data, clinical presentations, diagnostic investigations, operative procedures, histopathological features, morbidity, and mortality.

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3. Results

Of the 41 patients who met our inclusion criteria, 27 were male and 14 female, with a mean age at diagnosis of 39.8 years (range: 9–71 years). Only one patient was under 18 years (9 years old, male). While 38 cases (92.6%) were symptomatic, only three cases (7.3%) were asymptomatic, and the lesions were detected incidentally on chest radiography. The symptoms were hemoptysis in 28 patients (68.3%), shortness of breath in five (12.1%), cough in three (7.3%), and recurrent epistaxis in two (4.8%). Patients presenting with hemoptysis had a mean age of 40.5 years (range: 22–67 years). Eleven of these patients had life-threatening massive hemoptysis; one of them was in the 22nd week of gestation and one other patient, who presented with recurrent hemoptysis, had a history of hemoptysis during pregnancy 11 years ago. Three patients who had received transcatheter coil embolization had recurrent hemoptysis at 1 year, 6 months, and 2 months of follow-up, respectively. Two patients had petechiae of the lingual and labial mucosa and telangiectasia in various parts of the body, four had digital clubbing, two had cyanosis, and one had mitral valve prolapse and pectus excavatum. Systolic murmur was detected in the thorax during inspiration in only three patients. In the family history of two patients with HHT, 13 and 11 family members, respectively, had similar lesions and suffered from recurrent epistaxis or hemoptysis.

Routine laboratory tests such as complete blood count and biochemical parameters were within normal limits. In two patients, preoperative arterial oxygen saturations were 74% and 77% on room air, and they increased to 92% and 98% postoperatively. In all cases, chest X-rays and chest computed tomography (CT) images were taken. CT imaging was contrast-enhanced in all cases except one. In the case files of four patients, there were reports of CTs, but the images were not available. All radiographs were reevaluated retrospectively by a radiologist. Radiological findings are shown in Table 1. With the results of these findings, PAVM was not suggested in 62.1% (n = 23) of the cases, findings were compatible with PAVM in 35.1% (n = 13) of the cases (Figures 1 and 2), and PAVM was suspected in one patient (representing 2.7%). Preoperative angiography was performed in six patients and three of them were detected as having PAVM. These patients were ineligible for transcatheter embolization. Hemothorax was detected in one of the patients with pleural effusion and an invasive procedure was not considered.

Table 1. Radiological findings.

Chest X-rays		
Pathological findings 75.6%		
10% were consistent with AVM		
Computed tomography		
Sizes of the measurable lesions: 1.5–9 cm (mean: 3.8 cm)		
Signs	Number of patients (n)	%
1. Signs of lesions	37	100%
Limited lesions	19	51.3%
Possible limited lesions	16	43.2%
Obliterating air-column lesions in centrum	2	5.5%
2. Collapsed and consolidated	18	48.6%
3. Parenchymal hemorrhage	23	62.1%
4. Cavitation	5	13.5%
5. Bronchiectasis	13	35%
6. Pleural pathology	8	21.6%
7. Malignant mass lesion	11	29.7%
Positron emission tomography		
Suspected malignancy	4	10.5%

(Standard uptake maximum values: 2.97–15.03)

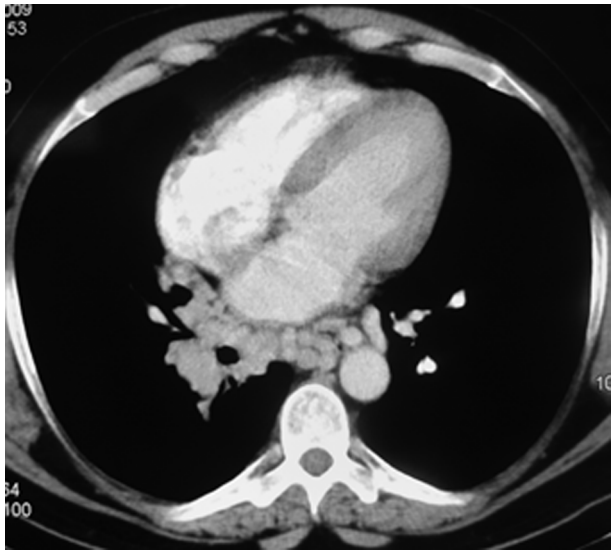

Figure 1. Hyperdense tubulonodular formations compatible with subcarinal, paraesophageal, and right infrahilar enlarged vascular structures.

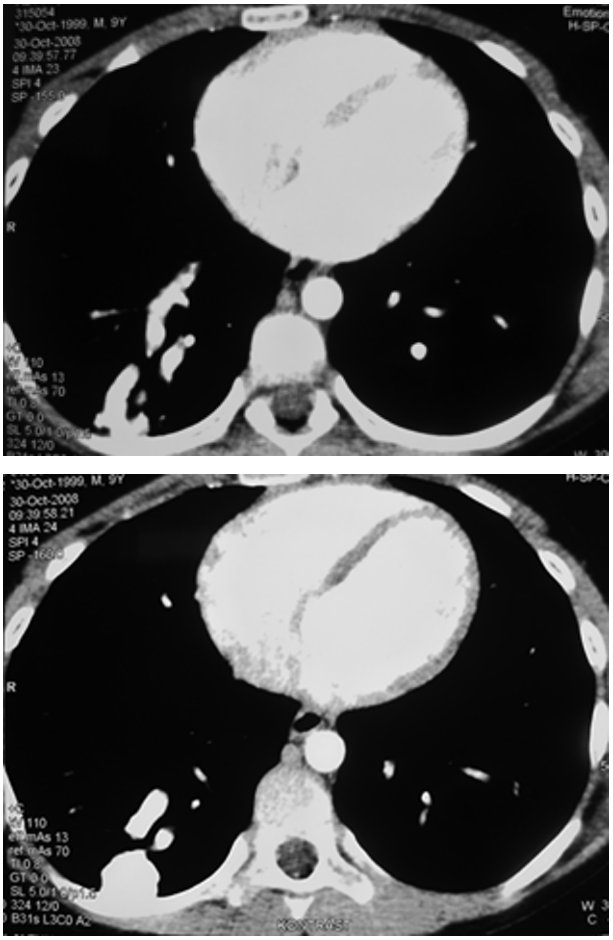

Figure 2. A, B) Dense contrasted tubular and nodular enhancing lesions consistent with typical PAVM; right lower lobe.

Bronchoscopy was performed in all cases. Rigid bronchoscopy was performed in the operating room on the patients operated on emergently, due to massive hemoptysis. Fiberoptic bronchoscopy was performed electively in the other cases. In 18 cases, there was active hemorrhage: hematoma in four, inactive hemorrhage with evidence of previous bleeding in four, endobronchial lesion in three, telangiectasia in two, mucosal irregularity in one, and vascular fistula in one. The other cases revealed normal findings. The biopsy results of the patients with endobronchial lesion indicated squamous metaplasia in one.

Although there were no neurological symptoms in 11 cases, contrast-enhanced brain CT or magnetic resonance imaging was performed. PAVM was observed in only two patients. The neurology clinic had followed these patients because they were asymptomatic.

Transthoracic needle biopsy was performed in three patients with suspected malignancy and pathology was reported nondiagnostic.

Cases followed with a presumptive diagnosis of PAVM were referred to the hematology and dermatology departments to investigate HHT. In the dermatological consultation, two patients with skin manifestations were evaluated as having HHT. While the eye examinations were normal in these cases, ear, nose, and throat examinations revealed telangiectasia on the nasopharynx, larynx, oral mucosa, and nasal mucosa. One of the patients suffered recurrent nose bleeding and the other suffered from coughing. In both cases, there was clubbing of the fingers. Chest X-rays showed that one had a lesion on the right-upper lobe and the other had bilateral lower-lobe lesions that were compatible with PAVM. Echocardiography indicated increased left ventricular diameter and volume, enlarged left ventricle, and pulmonary artery pressure of 45 mmHg in one case. Doppler ultrasonography of the liver indicated hepatic artery dilatation (10 mm) in one patient. The patient consulted with gastroenterology and follow-up was recommended. The patients who were diagnosed with PAVM via pulmonary angiography were ineligible for embolization procedure in another center.

Surgical resection alone was carried out for all patients. Emergency surgeries were performed in 11 cases; elective surgeries were performed when optimum conditions were established in the other cases. Including one case of bilateral procedure, 41 patients underwent 42 posterolateral thoracotomies (one rethoracotomy). Selective double-lumen intubation was performed in all cases. Table 2 shows the surgical procedures.

The macroscopic appearance of the lesions, in addition to pulsatile solitary lesions, indicated intralesional

Table 2. Surgical procedures.

	Right			Left				Total
	L	S	W	L	S	W	P	
Upper lobe lesion	5	1	1	5	4	1	1	18
Lower lobe lesion	11	1	1	7*		1		21
Middle lobe lesion	2		1					3

L: Lobectomy, S:segmentectomy, W:wedge resection, P: pneumonectomy, *:added lingulectomy.

hematoma in nine cases. The diameter of the lesions ranged from microscopic appearance to 9 cm. Histopathological examination revealed dense vascular structure in all cases, as well as parenchymal bleeding in 19 cases, intralesional hemorrhage and hematoma in nine cases, and bleeding accompanied by bronchiectasis in six cases.

One patient developed PAVM 15 years after hydatid cyst surgery, and six patients had lesions accompanied by bronchiectasis, suggesting that, in these cases, the lesions were secondary to these diseases. Therefore, PAVM was secondary to another disease in seven patients and congenital in 34 cases.

The postoperative follow-up period ranged from 6 months to 17 years (mean: 6 years). In the early postoperative period, left hemiplegia developed in one patient who had had a lower lobectomy. During follow up of this patient, a completion pneumonectomy was performed following consolidation of the upper lobe on radiologically. Myoplasty was performed in one patient who developed a bronchopleural fistula after postoperative empyema. One patient developed deep vein thrombosis, and one patient had recurrent hemoptysis. The source of bleeding could not be determined by angiography, but the hemoptysis improved with medical treatment. Five cases were lost to follow-up and all the other patients remained healthy.

4. Discussion

PAVM is a low-pressure, abnormal, vascular connection composed of an afferent pulmonary artery, distended efferent pulmonary veins, and an intervening thin-walled aneurysmal sac or tangle of dilated tortuous vascular channels. The PAVM may lie deep within the pulmonary parenchyma or immediately under the pleura (5). The incidence of PAVM in the general population is still not fully known, but is estimated to be 2–3 per 100,000 (1). In an autopsy study in 1953 at Johns Hopkins Hospital, only three cases of PAVM were detected in 15,000 consecutive autopsies. However, it was noted by the same investigators that small PAVMs could easily be missed in routine autopsy (3). Nakayama et al. identified eight patients with PAVM among 21,235 participants by low-dose thorax CT (6). More than 500 cases have been reported in the literature (1). At our clinic, 15,914 thoracic surgeries were performed between 1995 and 2012; therefore, PAVM frequency among thoracic surgeries was 26 in 10,000. Our hospital is a referral center for pulmonary disease and chest surgery patients, so this is a selected group of patients.

Females are affected twice as often as males, but there is a male predominance in newborns. Although PAVM occurs at any age, it is more frequently seen in the fifth and sixth decades. Around 10% of cases are identified in infancy or childhood (1,3). Contrary to the literature, our study showed male dominance, with a F/M ratio of 1:1.93. Only one of our patients (a 9-year-old male) was less than 18 years. The mean age of the patients was 39.8 years.

The etiology and natural course of PAVM is unknown. These lesions are considered to be congenital and, rarely, may be viewed as secondary to cases such as cardiac surgery (Glenn or Fontan procedures), hepatic cirrhosis, infections, amyloidosis, mitral stenosis, chest trauma, metastatic carcinoma, previous thoracic surgery, and bronchiectasis. Pregnancy is also associated with growth and increased complications of PAVM (1,7,8). It is known that there is a relationship between PAVM and HHT. Recent studies on the genetics of HHT have shown that genetic transitions observed in HHT may be applicable to PAVM etiology (3,5). In our study, PAVM was secondary in seven cases and congenital in 34 cases.

More than half of patients with PAVM are asymptomatic, and lesions are often detected by routine chest X-ray (7). In general, common symptoms are fatigue, dyspnea on exertion, cough, chest pain, hemoptysis, and recurrent epistaxis attacks. Although dyspnea is the most common symptom, it may not be seen until partial oxygen pressure drops to less than 60 mmHg (1,5). Risk factors are young age, lesions larger than 2 cm, and HHT disease (1,7). Epistaxis is the most common symptom in patients with HHT, characterized by spontaneous development or emergence with minor trauma (9). Hemoptysis rarely

occurs as a result of intrabronchial rupture or bleeding bronchial telangiectasis. Massive hemoptysis and massive hemothorax from rupture of PAVM into the pleural space have been reported, with fatal outcomes (10). Ference et al. reported an 8% incidence of fatal or massive hemoptysis or hemothorax (11). Pregnancy may be a risk for hemoptysis in patients with PAVM (1,10). Three patients (7.3%) in our series were asymptomatic. The most common reason patients seek medical attention is hemoptysis, which occurred in 68.3% of cases in this study; of these, 26.8% had life-threatening massive hemoptysis. One patient had hemoptysis while she was pregnant, two patients had epistaxis, and one patient had hemothorax.

The majority of patients (75%) show no abnormal physical findings. Physical examination can reveal cyanosis, anemia, telangiectasia, clubbed fingers, and systolic murmur heard in the chest. The classic triad of PAVM is cyanosis–polycythemia–clubbed fingers, but these were observed in only 20% of the cases. The presence of cyanosis suggests that at least 25%–30% of the blood in pulmonary circulation goes with right-to-left shunting to the left atrium without oxygenation (1,12). In patients with HHT, the most common and often the only finding is skin telangiectasis (3). According to the International Clinical Diagnostics (Curaçao) Criteria, diagnosis of the disease is made via three or more of following criteria: self-recurrent bleeding episodes, mucocutaneous telangiectasis, visceral PAVMs, and first-degree relatives diagnosed with HHT (5,13). A Mayo Clinic study reported cyanosis, clubbing, and murmur incident rates of 29%, 19%, and 34%, respectively (5); in our study, these rates were 4.8%, 9.7%, and 7.3%, respectively. Only two of our cases were accompanied by HHT.

PAVMs are serpiginous-like masses related with vessels or nodular masses with clear margins on thoracic CT. The classic view of PAVM on chest radiographs is in the form of round or oval lesions of equal density, frequently lobulated but well circumscribed, mostly occurring in the lower lobes, and 1–5 cm in diameter. Lesions are the only indication in two-thirds of cases. In plain chest X-rays, PAVM may manifest in the form of the spread of hemorrhaging to parenchyma, or atelectasis due to bronchus compression. Microvascular telangiectasis can create a normal chest X-ray, or pulmonary vascular signs may increase vaguely (2,7,12). In dynamic examination, PAVMs typically enhanced contrast after the right ventricle or pulmonary artery, before the left atrium and ventricle. Sensitivity and specificity of PAVM detection in contrast-enhanced CT has been reported as 83% and 78%, respectively, which compares favorably with digital subtraction pulmonary angiography results of 70% and 100% (10). Remy et al. reported that the CT rate of PAVM diagnosis was 98.2%, and that of angiography was 59.6% (14). Contrast-enhanced chest CT can view multiple small PAVMs, whereas pulmonary angiography sometimes cannot (1). In our study, radiological investigations of patients who were surgically diagnosed with PAVM were reevaluated for this study. Unlike other studies, 62.1% of the lesions were not compatible with PAVM; they were mostly consolidation of bleeding and of a ground-glass appearance.

The current preferred treatment for the majority of patients with PAVMs is percutaneous embolotherapy using coils or balloons (15). In recent years, Amplatzer vascular plugs have also been used. Embolization is less invasive than surgery, and it is an easily applicable method. The success rate has been reported as 98% (16). Surgical approaches include wedge resection, segmentectomy, lobectomy, and pneumonectomy. In our study, surgery was preferred in cases where treatment of the embolism was unsuccessful or not possible, in patients with symptomatic and complicated PAVM, and/or when a differential diagnosis of PAVM was not possible. The surgical approach is anatomical resection in our clinic. The parenchyma-sparing approach has been preferred in small, peripherally located lesions in recent years and was performed in 26% of the operations in this study.

Approximately 20% of untreated patients die due to complications (5). The most frequently reported complications relate to the central nervous system and are seen in 30% of patients. Complications were often seen in patients whose feeding artery was more than 3 mm in diameter (17). Hemoptysis and hemothorax are rare but life-threatening complications. Stringer et al. reported that 30 of 140 patients with PAVM had serious complications or died. Dalton et al. reported that nine patients developed intrapleural rupture (12). The mortality rate has been reported as 50% in untreated cases, compared with less than 3% in treated cases (1,3). Neurological complications were not observed in this study, but 70.7% of the patients were admitted to our clinic with complications. Postoperative complications were seen in three cases, and mortality was not reported. Five cases were lost to follow-up and the other patients remain healthy.

In conclusion, PAVMs are rare clinical conditions. Surgery remains the first choice in cases where treatment of the embolization cannot be performed or has not been successful, in symptomatic and complicated patients with PAVM, and/or cases where the PAVM diagnosis cannot be established.

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