CASE REPORTS

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Pulmonary arteriovenous malformation: A case report

Plućna arteriovenska malformacija

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Abstract

Introduction. Pulmonary arteriovenous malformation (PAVM) is pathological communication between pulmonary artery and pulmonary vein, in way that it shunts normal alveolar capillary membrane resulting in inadequate blood oxygenation in this part of the lung parenchyma Modern therapy of PAVMs includes surgical treatment or endovascular embolization. **Case report.** A 30-year-old female patient had signs of parestesia and weakness of the extremities on the left side of her body. On physical examination there was only cyanotic discoloration of her lips and clubbing fingers. On the chest x-ray, in the right hemithorax, in the inferior region of the lung, there was relatively homogeneous and well defined shadow, intensity of the soft tissue, which was about 35 mm. A multislice computed tomography pulmonary angiography was performed and showed, in lung parenchyma on both sides, many PAVMs, of which the largest (35 mm) was in inferior right region of the lung on crossing between apical and posterior basal lung segment with 7 mm diameter feeding artery and 9 mm diameter draining vein. The selective pulmonary angiography was performed by Seldingers technique. Through sheath, we placed a plug with a diameter of 10 mm. The plug was expanded and a complete occlusion of the final part of the feeding branch of this PAVM was achieved (confirmed by control angiography). In that way, the PAVM was fully shut off from the circulation. In 3 months follow-up, the patient was feeling well, without any recorded complication. **Conclusion.** Endovascular embolization is recommended as therapy of the first choice for all of PAVMs that have feeding artery greater than 2 mm. Endovascular embolization has high success rate with minimal complications.

Key words:
arteriovenous malformations; computed tomography angiography; embolization, therapeutic; endovascular procedures; multidetector computed tomography.

Apstrakt

Uvod. Plućna arteriovenska malformacija (PAVM) je patološka komunikacija između plućne arterije i plućne vene kojom se zaobilazi normalna alveo-kapilarna membrana, usled čega se krv u tom delu plućnog parenhima ne oksigeniše. Savremeni pristup lečenju PAVM uključuje hirurški tretman ili embolizaciju endovaskularnim putem. **Prikaz bolesnika.** Bolesnica stara 30 godina javila se zbog trnjenja i slabosti ekstremiteta leve strane. Objektivno, imala je cijanozu usana i batičaste prste. Na radiografiji srca i pluća, u desnom hemitoraksu, u donjem plućnom polju u projekciji medioklavikularne linije, nwwađena je relativno homogena i relativno jasno ograničena senka promera oko 35 mm, intenziteta mekih tkiva. Urađena je multislajsna kompjuterizovana tomografija sa pulmoangiografijom i u plućnom parenhimu, obostrano, utvrđen je veći broj PAVM, od kojih je najveća (promera oko 35 mm) bila u donjem desnom plućnom režnju na prelazu između apikalnog i posterobazalnog segmenta, sa uočljivom dovodnom arterijom prečnika oko 7 mm i odvodnom venom prečnika oko 9 mm. Primenom Seldingerove metode urađena je selektivna pulmoangiografija. Kroz uvodnik, uveden je „čepić" (eng. *plug*) prečnika 10 mm, „čepić" je ekspandiran, čime je indukovana potpuna okluzija završnog dela dovodne grane te PAVM (potvrđeno kontrolnom angiografijom). Na taj način, PAVM je u potpunosti bila isključena iz cirkulacije. Tokom tri meseca praćenja, bolesnica se osećala dobro i nisu zabeležene nikakve komplikacije. **Zaključak.**

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Endovaskularna embolizacija se preporučuje kao terapija prvog izbora za sve PAVM čija arterija hranilica ima dijametar veći od 2 mm. Endovaskularna embolizacija pokazuje visoku stopu uspešnosti sa minimalnom stopom komplikacija.

Ključne reči:
arteriovenske malformacije; angiografija, tomografska, kompjuterizovana; embolizacija, terapijska; endovaskularne procedure; tomografija, kompjuterizovana, multidetektorska.

Introduction

Pulmonary arteriovenous malformation (PAVM) is pathological communication between pulmonary artery and pulmonary vein, in way that it shunts normal alveolar capillary membrane. Therefore, the blood in this part of the lung parenchyma is not oxygenized which leads to hypoxia and symptoms including adynamia, tiredness, dyspnea in physical activity, hemoptysis, palpitations, cough, paradoxal systemic embolism and chest pain [1, 2]. Smaller size PAVMs usually are asymptomatic and are incidental findings or can be discovered in case of complications. Modern therapy of PAVMs includes surgical treatment or endovascular embolization [3].

We presented a patient with PAVM successfully treated with endovascular approach.

Case report

A 30-year-old female patient was admitted to the Clinic for Neurology with signs of parestesia and weakness of the extremities on the left side of her body. On physical examination, there was only cyanotic discoloration of her lips and clubbing fingers. Neurological exam was normal. All laboratory results were in referent ranges.

Chest x-ray was performed in standing and posterior-anterior (PA) position. In the right hemithorax, in the inferior region of the lung on medioclavicular line projection, there was relatively homogeneous, relatively well defined shadow, intensity of the soft tissue, which was about 35 mm. Also, on the same side, there was voluminous hilus and prominent hilo-basal pulmonary vascularity (Figure 1). Based on the localization and the appearance of the shadow, and with anamnestic data that before 15 years the patient had been subjected to the surgery for PAVM, we suspected appearance of the new PAVM. We conducted multislice computed tomography (MSCT) pulmonary angiography which revealed many PAVMs in lung parenchyma on both sides, of which the largest one with diameter of 35 mm was in inferior right region of the lung on crossing between apical and posterior basal lung segment with 7 mm diameter feeding artery and 9 mm diameter draining vein (Figures 2−4).

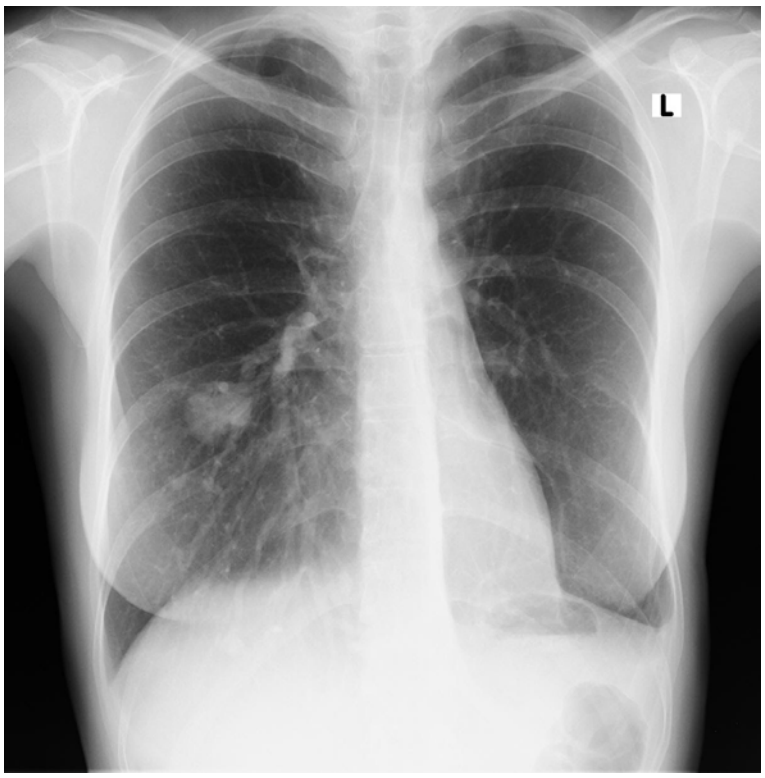


Fig. 1 - Chest x-ray in standing and posterior anterior (PA) position shows relatively homogeneous, relatively well defined shadow, intensity of the soft tissue, size about 35 mm in the right hemithorax, in the inferior region of the lung on medioclavicular line projection. Also, on the same side, there is voluminous hilus and prominent hilobasal pulmonary vascularity.

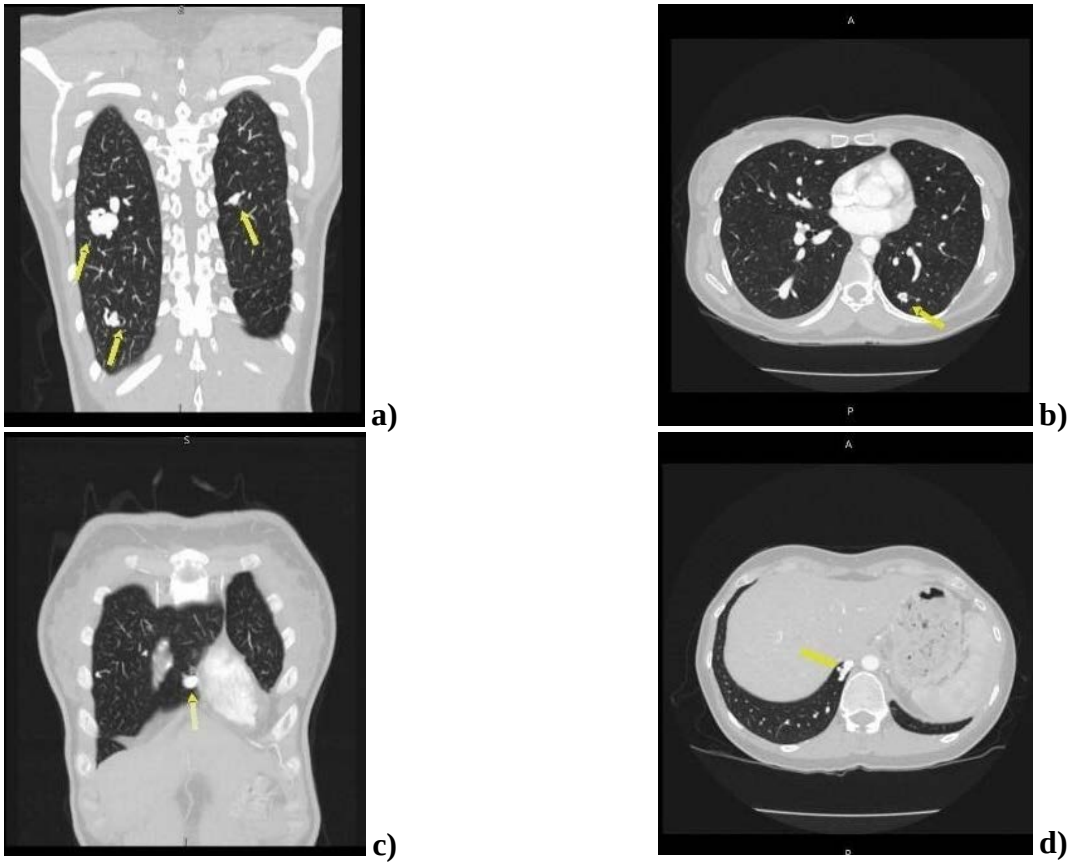


Fig. 2 (a–d) – Multislice computed tomography with pulmonary angiography (MSCT PA) finding shows many pulmonary arteriovenous malformations (PAVMs) in lung parenchyma on both sides.

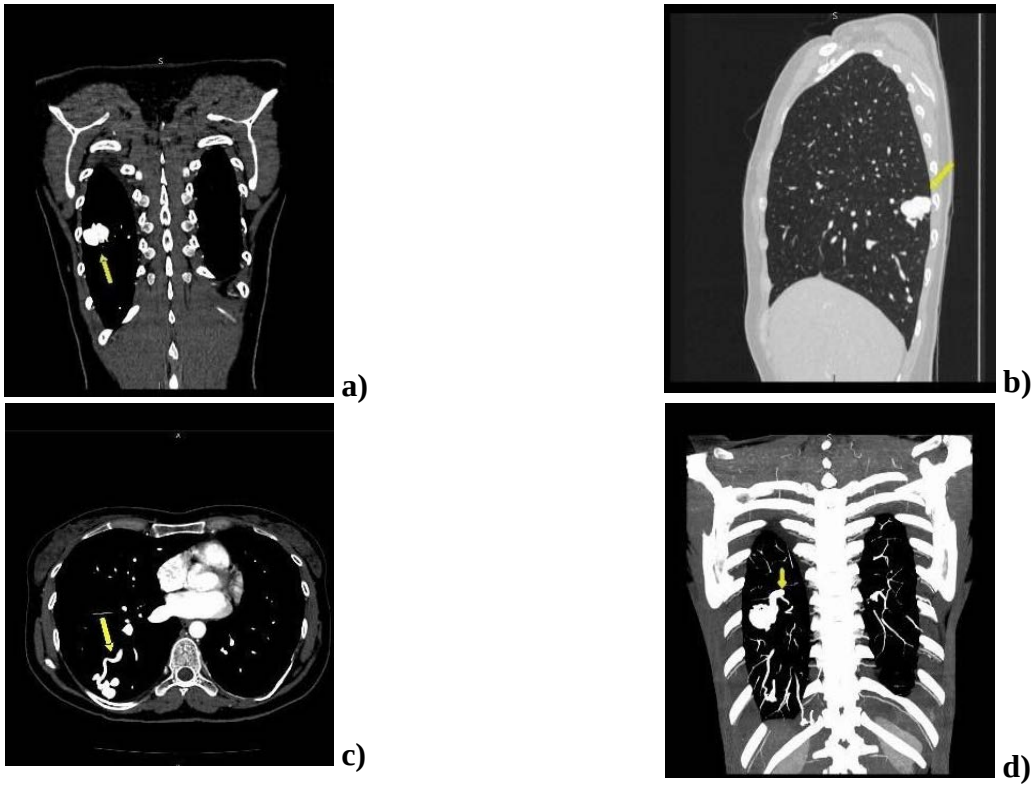
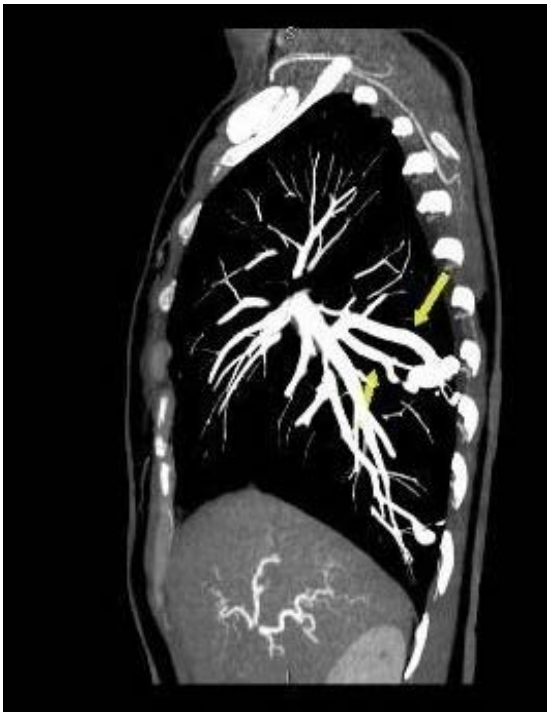
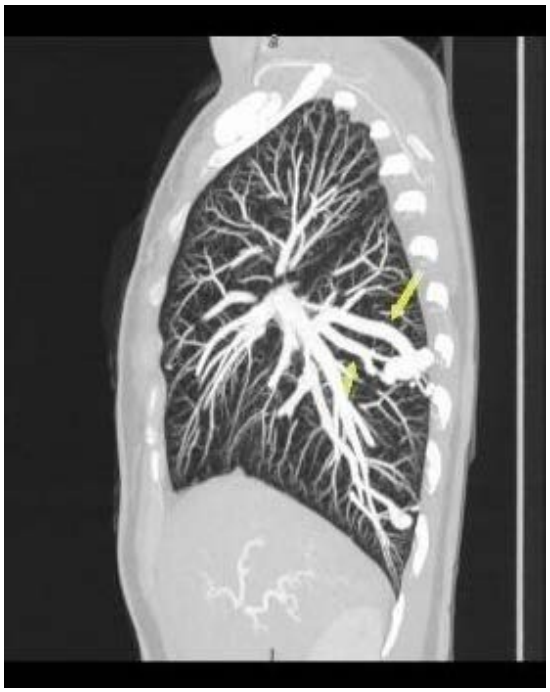


Fig. 3 (a–d) – Multislice computed tomography with pulmonary angiography (MSCT PA) shows the largest pulmonary arteriovenous malformation (PAVM) with diameter of 35 mm in inferior right region of the lung on crossing between apical and posterior basal lung segment.



a)



b)

Fig. 4 (a, b) – Multislice computed tomography pulmonary angiography (MSCT PA) findings: the largest pulmonary arteriovenous malformation (PAVM) with 7 mm diameter feeding artery and 9 mm diameter draining vein.

Medical team, made of a neurologist, thoracic surgeon and radiologist, decided that the largest among PAVMs should be taken care of by endovascular approach. Intervention was conducted in the Department for Interventional Vascular Radiology, Institute of Radiology, Military Medical Academy in Belgrade in collaboration with colleagues from "Dedinje" Cardiovascular Institute, Belgrade.

The right transfemoral access was obtained by Seldingers technique, and a 6F introducer sheath (Merit Medical) was placed. A guide wire (150 mm/ 0.035 In; Merit Medical) and 6F Pigtail catheter (Cordis) were then advanced into the right femoral vein towards the inferior vena cava to the right atrium and ventricle, with electrocardiography (ECG) monitoring, further in *truncus pulmonis*, from where we entered in the main right branch of the *truncus pulmonis*. Then, the selective pulmonary angiography was performed (Figure 5). The PAVM was noticed with feeding and draining blood vessels, as it was shown on MSCT pulmonary angiography.

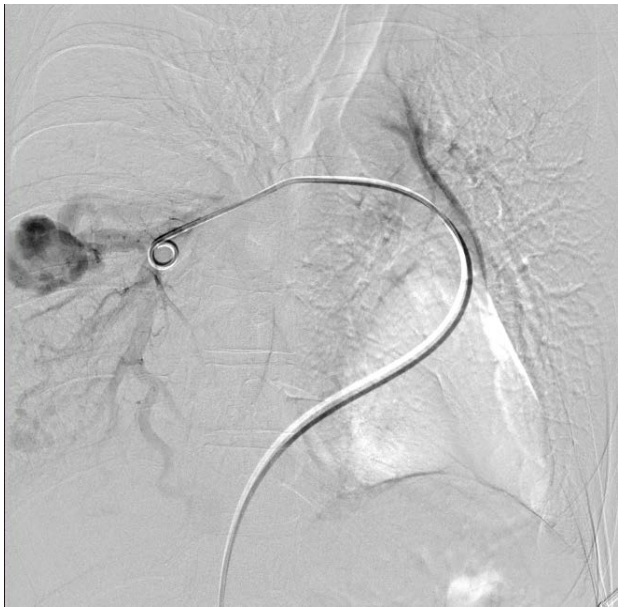


Fig. 5 – Selective pulmonary angiography finding: the pulmonary arteriovenous malformation (PAVM) was noticed with feeding and draining blood vessels.

We entered the feeding branches of the PAVM by using 7F multipurpose (MP) catheter (Cordis) and performed suprase-lective angiography (Figure 6). 7F long peripheral sheath, (Shapeless, Arrow, Terumo) was placed over 260 cm Hi-drostiff wire (Merit Medical), with tip in feeding branch of the PAVM. We placed the 10 mm diameter plug (Amplatzer Vascular Plug II, AGA Medical Corporation) through this sheath. The plug was expanded causing a complete occlusion of the final part of the feeding branch of this PAVM. Control angiography (Figure 7) showed that plug was in correct place with total occlusion of feeding branch of the PAVM, which was now fully shut off from the circulation.

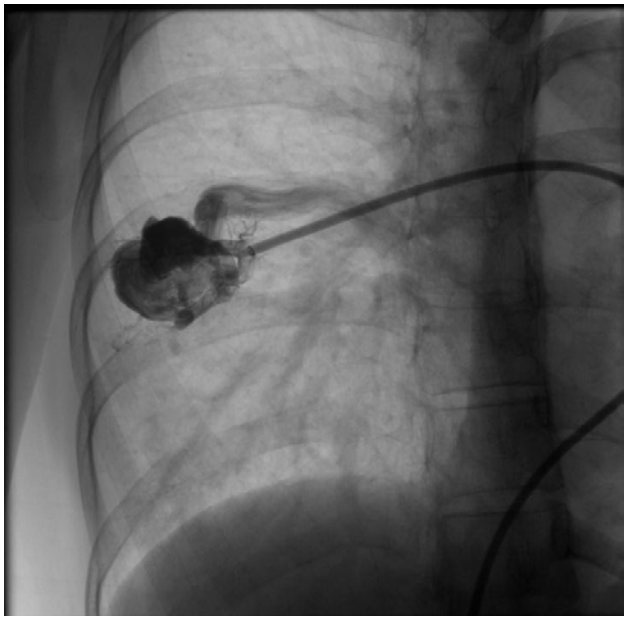


Fig. 6 – Superselective pulmonary angiography after entering the feeding branches of the pulmonary arteriovenous malformation (PAVM).

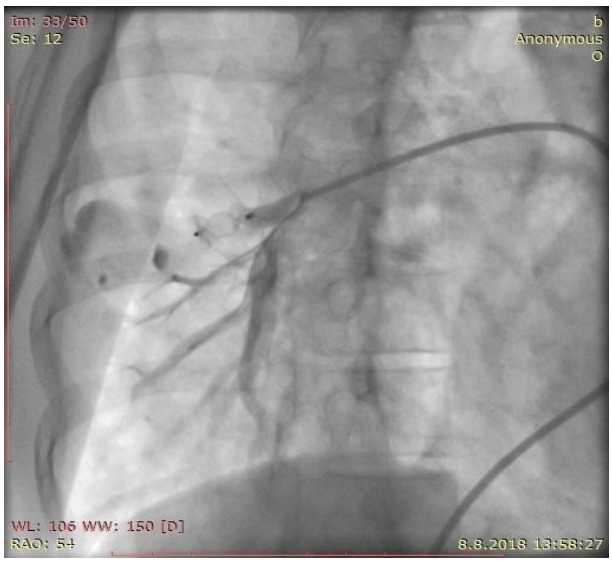


Fig. 7 – Control angiography after expanding the 10 mm diameter plug (Amplatzer Vascular Plug II, AGA Medical Corporation) in final part of the feeding branch of the pulmonary arteriovenous malformation (PAVM) shows that there is total occlusion of feeding branch of the PAVM.

In 3 months follow-up, the patient was feeling well without any recorded complication.

Discussion

Pathologic pulmonary arteriovenous malformation is the direct communication between the branches of pulmonary artery and pulmonary vein in way that there is a shunt of normal lung capillaries which leads to chronic hypoxia [1, 2, 4]. The incidence of PAVM is 2–3 cases in 100,000 people [1, 5, 6]. In more than 80% of cases, it is congenital anomaly (together with hereditary hemorrhagic telangiectasia or Osler-Weber-Rendu syndrome), and in rare cases, it was caused by trauma of the thoracic cavity, thoracic surgery, long-term hepatic cirrhosis, metastatic disease, stenosis of the mitral valve, infections and systemic amyloidosis [1-5, 7, 8]. Also, it can appear in pregnancy. Based on literature, 33% of patients with PAVM had earlier stroke, 18% had transitory ischemic attack, 23% had cerebral abscess, 3% had haemothorax, and 59% of patients had symptoms of dyspnea or intolerance on physical activity [9].

PAVMs are stabile in 75% of the cases or slow growth, and only in small number of cases, most often because of no treatment, PAVMs can induce high rate of morbidity and mortality [10]. Complications of PAVMs are brain abscess, stroke, hemoptysis and haemothorax, hypoxia, polycythemia, endocarditis, transitory ischemic attack, migraine and congestive hearth weakness [1, 4, 6]. Risk of neurological complications is higher in diffuse type of PAVM, large shunt and feeding branch diameter more than 3 mm [11] and in untreated forms of PAVM in comparison to treated forms [4].

Standard thoracic surgical techniques were previously available as the only treatment method (for example: ligation, local excision, segmentectomy, lobectomy, or pneumonectomy) [12]. In some cases, staged bilateral thoracotomies or

video-assisted thoracoscopic resection are performed [13]. Surgical resection is rare method of PAVM treatment and is reserved for the cases with lesions that are resistant on endovascular therapy or when the endovascular treatment is not available. Every time, when it is possible, endovascular embolization is the gold standard in treating PAVM and is conducted since 1980s [9]. Nevertheless, for large, centrally localized lesions, lobectomy is still required. Surgery is a safe method of treatment of PAVMs in selected cases, i.e. when PAVM is solitary and large (> 2 cm diameter), and the risks of embolotherapy are high. Surgery remains 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 [14].

Typically, patients with hereditary telangiectasia are suspected for PAVM and they undergo screening radiographic procedures. In those patients, PAVM is discovered in about 15% of cases [4]. There is 90% of chances of discovering hereditary telangiectasia in patients that have been initially diagnosed with PAVM and they are send on further investigation. Because of that, it is important that every patient with suspected PAVM undergoes detailed and targeted diagnostic. It means that before visiting the interventional radiologist, some other investigations have to be done: multidisciplinary clinical evaluation, imaging for endovascular embolization (number of lesions, localization and type of PAVM, as well as measured diameter of feeding/s arteries), anesthetic assessment for the type of anesthesia, ECG (searching for the block of the left branch and hypertrophy of the right ventricle), complete blood count, coagulation status and other laboratory analyses. It is also important to have patient's informed consent [4]. Gold standard in diagnosing the PAVM is MSCT pulmonary angiography. It is important to identify localization and the type of PAVM, and the diameter of the feeding artery or the feeding branch of pulmonary artery. There is simple PAVMs which have only one feeding artery (80–90% of all PAVMs), complex ones with two or more feeding branches (10–20%) and, rarely, diffuse PAVMs (5%).

Some of the tests which can indicate the PAVM presence are lower oxygen saturation, conventional chest x-ray and transthoracic contrast echocardiography (TTCE). High sensitivity of the TTCE (98.6%) is very important in diagnostics. If this test is positive, the MSCT of the thorax is performed, where the PAVM can be seen, and if it is not visible on MSCT, than there is possibility that PAVM has microscopic dimension [2].

Main indications for treating PAVMs with endovascular approach are: PAVM with diameter of feeding artery greater than 2 mm, symptomatic PAVM with no matter what size it has and atypical lesion that is similar to PAVM on MSCT and presence of suggestive symptoms [4].

Different materials for embolization are available but for those purposes usually coils and plugs are used. These materials function by causing total occlusion of the distal part of feeding artery of PAVM, which leads to its complete occlusion and shunt off of PAVM from the circulation [4].

Embolization of PAVM is very successful method of treatment (success of > 99%) [1, 15]. Successful occlusion can be achieved with only one treatment in 85% of patients, and symptomatic relief can be expected immediately after the treatment [16, 17]. In 83% of the cases in patients with PAVM, treated lesion stays occluded, but in 17–20% of cases there is possibility of reperfusion or formation of new PAVM [15, 18, 19].

Conclusion

Malformations, like PAVM, are associated with high morbidity and mortality if not treated. That is why endovascular embolization is recommended as therapy of first choice for all of the lesions that have feeding artery greater than 2 mm. Endovascular embolization has high success rate with minimal complications. Despite, there is significant risk of recanalization of treated PAVM or more lesions developing. Because of that, long term follow-ups are recommended after embolization.

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