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Please cite this article **PULMONARY ARTERIOVENOUS MALFORMATION: CASE REPORT**

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UDC:

DOI: https://doi.org/10.2298/VSP190819029S

When the final article is assigned to volumes/issues of the Journal, the Article in Press version will be removed and the final version appear in the associated published volumes/issues of the Journal. The date the article was made available online first will be carried over.
PULMONARY ARTERIOVENOUS MALFORMATION: CASE REPORT
PLUĆNA PATOLOŠKA ARTERIOVENSKA KOMUNIKACIJA: PRIKAZ SLUČAJA

Igor Sekulić*, Dragan Sagić†, Siniša Rusović†, Dragan Dulović*, Viktor Pasovski†,
Jelena Bošković-Sekulić‡, Nemanja Rančić*, Jelena Stevanović*, Ranko Raičević‡

*Institut za radiologiju, Vojnomedicinska akademija, Beograd, Srbija
†Klinika za radiologiju, Institut za kardiovaskulare bolesti Dedinje, Beograd, Srbija
‡Klinika za neurologiju, Vojnomedicinska akademija, Beograd, Srbija
‖Urgentni centar, Klinički centar Kragujevac, Kragujevac, Srbija

*Institute for Radiology, Military Medical Academy, Belgrade, Serbia
†Clinic for Radiology, “Dedinje” Cardiovascular Institute, Belgrade, Serbia
‡Clinic for Neurology, Military Medical Academy, Belgrade, Serbia
‖Center for Urgent medicine, Clinical Center Kragujevac, Kragujevac, Serbia

Corresponding author: Ranko Raičević, MD, PhD, Clinic for Neurology, Military Medical Academy, Belgrade; E-mail: rankoraicevic@yahoo.com
Introduction

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

In our paper we showed one case with PAVM which was successfully treated with endovascular approach.

Case report

A 30-year-old female patient was admitted to the neurology service 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 range.

Chest x-ray was performed in standing and 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 (Image 1). Based on the localization and the appearance of the shadow, and with anamnestic data that before 15 years patient has gone under surgery for PAVM, we suspected that now there is the case of new PAVM. Further we conducted multislice computed tomography (MSCT) pulmonary angiography where in lung parenchyma on both sides there were many PAVMs, of which the largest of 35 mm was in inferior right region of the lung on crossing between apical and poster basal lung segment with 7 mm diameter feeding artery and 9 mm diameter draining vein (Image 2-4).

After, medical team, made of neurologist, thoracic surgeon and radiologist, decided that the largest among PAVMs should be taken care of by endovascular approach; intervention was conducted in Department for Interventional vascular radiology, Institute of radiology, Military Medical Academy with colleagues from “Dedinje” Cardiovascular Institute, Belgrade

With Seldingers technique, right transfemoral access is obtained and a 6F introducer sheath (Merit Medical) is placed. A guide wire (150 mm / 0.035 In; Merit Medical) and 6F Pigtail catheter (Cordis) is then advanced into right femoral vein towards inferior vena cava to right atrium and ventricle, with electrocardiography or EKG monitoring, further in truncus pulmonis, from where we entered in main right branch of the truncus pulmonis. Than the selective pulmonary angiography was performed (Image 5). The PAVM was noticed with feeding and draining blood vessels, as it was described on MSCT pulmonary angiography. With 7F multipurpose (MP) catheter (Cordis) we entered the feeding branches of the PAVM and performed supraselective angiography (Image 6). 7F long peripheral sheath, Shapeless (Arrow, Terumo) was placed over 260cm Hidrostiff wire (Merit Medical), with tip in feeding branch of PAVM. Through this sheath we placed the 10 mm diameter Plug (Amplatzer Vascular Plug II, AGA Medical Corporation). The Plug was expanded and there was a complete occlusion of the final part of the feeding branch of
this PAVM. After performing control angiography (Image 7) we noticed that Plug is in correct place and there was total occlusion feeding branch of the PAVM, which was now fully shut off of the circulation.

Discussion

Pathologic pulmonary arteriovenous malformation is the direct communication between the branches of pulmonary artery and pulmonary vein in way that there is a shunts of normal lung capillaries which leads to chronic hypoxia. The incidence of PAVM is 2-3 cases in 100,000 people. In more than 80% of cases it is congenital anomaly (together with hereditary hemorrhagic telangiectasia or Osler-Weber-Rendu syndrome), and the rare cases are after trauma of the thoracic cavity, thoracic surgery, long-term hepatic cirrhosis, metastatic disease, stenosis of the mitral valve, infections and systemic amyloidosis. Also, it can appear in pregnancy. Based on literature, 33% of patients with PAVM had earlier stroke, 18% transitory ischemic attack, 23% cerebral abscess, 3% haemothorax, and 59% symptoms of dyspnea or intolerance on physical activity. They are stable in 75% of the cases or slow growth, and only in small number of cases PAVM can induce higher rate of morbidity and mortality because of no treatment. Complications are brain abscess, stroke, hemoptysis and haemothorax, hypoxia, polycythemia, endocarditis, transitory ischemic attack, migraine and congestive heart weakness. Risk of neurological complications is higher in diffuse type PAVM, large shunt and feeding branch diameter more than 3 mm and in untreated forms of PAVM in comparison to treated forms.

Standard thoracic surgical techniques were previously available as the only treatment method (for example: ligation, local excision, segmentectomy, lobectomy, or pneumonectomy). In some cases, staged bilateral thoracotomies are performed or video-assisted thoracoscopic resection. Surgical resection is rare method of treatment and is reserved for the cases with lesions that are resistance 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 the PAVM and is conducted since 1980s. 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 the 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.

Typically patients with hereditary telangiectasia are examined; they undergo screening radiographic procedures PAVM is discovered in about 15% of cases. There was 90% chance of discovering hereditary telangiectasia with patients that had been initially diagnosed with PAVM and who were send on further investigation. Because of that it is important that every patient with suspected PAVM undergo 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, EKG (searching for the block of the left branch and hypertrophy of right ventricle), complete blood count, coagulation status and other laboratory analyses, it is also important to have patient informed confirm. 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 are simple PAVM which have only one feeding artery (80-90% of all PAVMs), complex with two or more feeding branches (10-20%) and rarely diffuse PAVM (5%).

Some of the tests which can indicate the PAVM 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’s not visible on MSCT, than there is possibility that PAVM is microscopic 2.

Main indications for treating PAVMs with endovascular approach are: PAVM with diameter of feeding artery greater than 2 mm, which can be treated with endovascular approach, symptomatic PAVM no matter what size it is 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 PAVM, which leads to a complete occlusion of the feeding arteries and blood flow to the PPAVK 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 right after treatment 16, 17. In 83% of the cases in patients with PAVM, treated lesion stays occluded, but in 17-20% of cases there is danger chance of reperfusion or formation of new PAVM 15, 18, 19.

Conclusion

Malformations like PAVM are associated with higher morbidity and mortality if not treated. That’s 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. Although there is different material for the embolization, there is significant risk of recanalization of treated PAVM or there can be more lesions developing. Because of all that was said, after embolization long term follow-ups are recommended.

References

Image 1. Chest x-ray in standing and PA position

Image 2. Multislice computed tomography pulmonary angiography
Image 3. Multislice computed tomography pulmonary angiography
Image 4. Multislice computed tomography pulmonary angiography (Maximum intensity projection or MIP sequences)
Image 5. Selective pulmonary angiography
Image 6. Superselective pulmonary angiography
Image 7. Control angiography
Received on August 19, 2019.
Accepted on March 13, 2020.
Online First March, 2020.