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Pulmonary arteriovenous malformation – does anyone still remember about this abnormality?

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Summary

Background:

Pulmonary arteriovenous malformation (PAVM) is a junction between medium-sized arteries and veins bypassing the capillary system. The junctions may have a very different macro- and microscopic structure; they may be multiple or single. Their important feature is shortening of blood flow route between the pulmonary artery and pulmonary veins. PAVM is a very rare pathology, occurring twice more often in females than males; it may coexist with Osler-Weber-Rendu disease and may be hereditary. Currently the diagnostic 'gold standard' for this pathology is CT-angiography and the treatment of choice is embolization or cardiothoracic surgery.

Case Report:

In this article we presented CT images of an incidentally diagnosed PAVM in a 33-year-old woman, visible as a rounded opacity on chest radiograph.

Conclusions:

A routine chest radiograph in two basic views (PA and lateral) demonstrated the presence of a rounded opacity in the posterior basal segment of the left lower lobe. However it was not sufficient for a certain diagnosis. The chest CT examination with unenhanced and enhanced scans allowed to differentiate the described lesion and to conclusively diagnose arteriovenous malformation. The patient remains under constant thoracosurgical follow-up.

MeSH Keywords:

Pulmonary Artery • Telangiectasia, Hereditary Hemorrhagic • Tomography Scanners, X-Ray Computed

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Background

Pulmonary arteriovenous malformation (PAVM) is a junction between medium-sized arteries and veins bypassing the capillary system, which affects the impairment of oxidation and nutrition processes. The junctions may have diverse macro- and microscopic structure, they may be multiple or single. Their important feature is shortening of blood flow route between the pulmonary artery and pulmonary veins.

This malformation is shown in a radiographic image as a rounded or acinous opacity, which in CT angiography corresponds to vascular lumen widening in the junction with a wide double vascular opacity reaching the pulmonary hilus. These are afferent and efferent vessels. The rounded opacity may be of different sizes and be located in any area

of lung fields, yet in 80% of cases it occurs in lower lung lobe, in 15% of cases it coexists with Osler-Weber-Rendu disease [1].

In some cases pulsation may be observed and demonstrated in an x-ray scopy or the difference in the opacity size in the Valsalva or Muller's maneuvers may be visualized (Muller's maneuver: deep expiration and an attempt at inspiration made with closed mouth and nose, the generated negative pressure grow in the chest increases heart and pulmonary vessels volume – the maneuver is also used to demonstrate the presence of hiatus hernia and diaphragm mobility).

For the first time pulmonary arteriovenous fistula was described individually in 1939 – this was achieved by Smith and Horton's team of the United States of America.

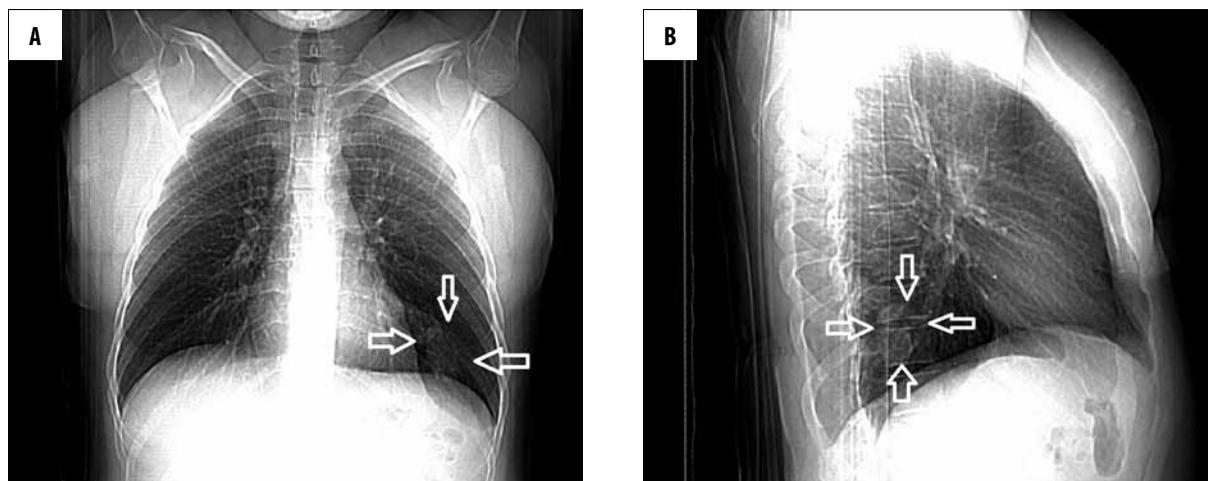


Figure 1. Chest CT topogram AP (A) and lateral (B) view – rounded opacity in the posterior basal segment of the left lung (arrows).

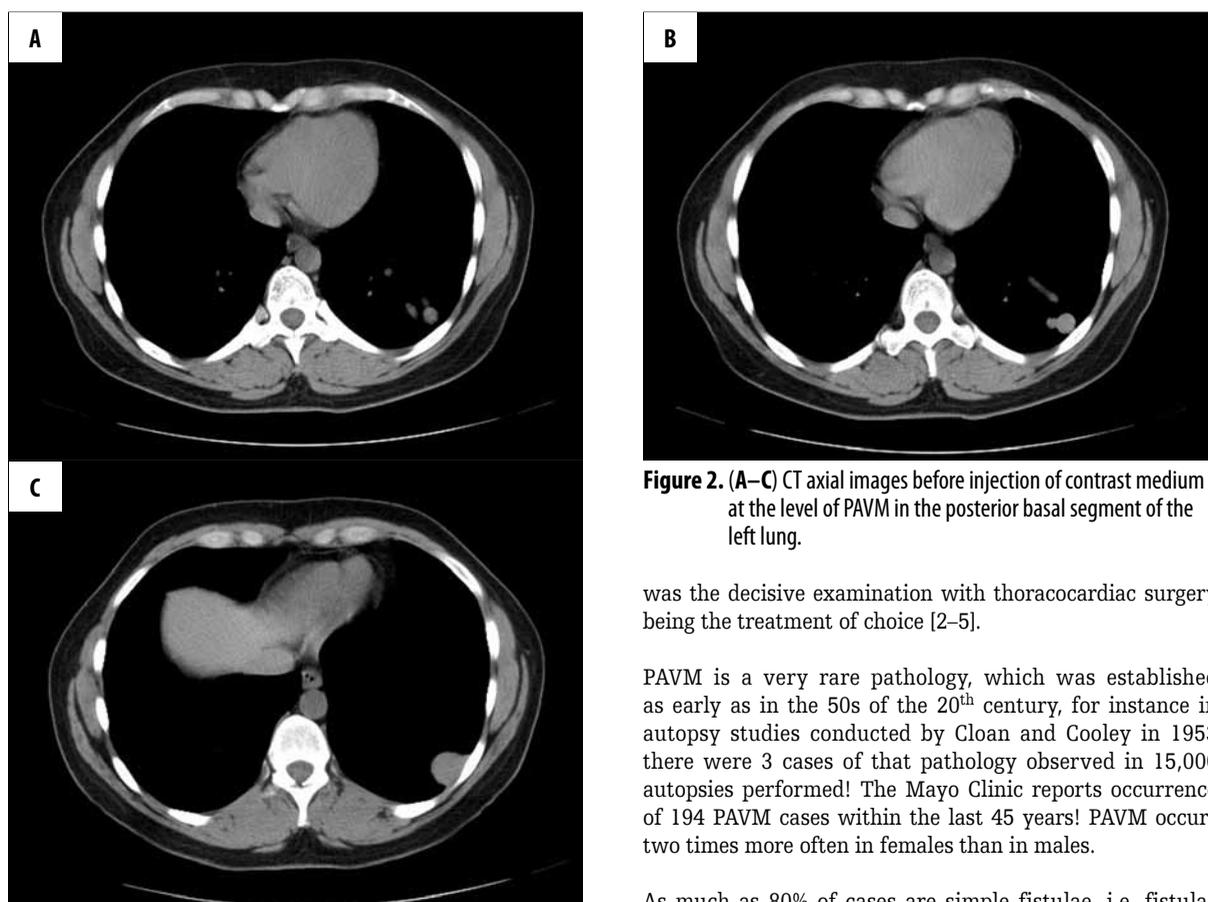


Figure 2. (A–C) CT axial images before injection of contrast medium at the level of PAVM in the posterior basal segment of the left lung.

In reports concerning its pathology made available in the 40s and 50s of the 20th century most patients did not experience any clinical symptoms and in case of some of the patients cyanosis, polycythemia and clubbed fingers were observed. There was observed more frequent occurrence of the pathology in people with Osler-Weber-Rendu disease, hereditary form, more frequently in the left lower lobe (more than 50% of cases).

Differential diagnosis procedures allowed for neoplastic proliferation, tuberculosis, hamartoma and angiography

was the decisive examination with thoracocardiac surgery being the treatment of choice [2–5].

PAVM is a very rare pathology, which was established as early as in the 50s of the 20th century, for instance in autopsy studies conducted by Cloan and Cooley in 1953 there were 3 cases of that pathology observed in 15,000 autopsies performed! The Mayo Clinic reports occurrence of 194 PAVM cases within the last 45 years! PAVM occurs two times more often in females than in males.

As much as 80% of cases are simple fistulae, i.e. fistulae fed by single arteries and with single venous drainage; 20% of cases are multiple fistulae with more than one feeding artery and venous drainage respectively.

Single fistulae occur in 35–74% of cases, multiple fistulae occur in 35–65% of cases and more frequently result in complications in the form of infections or bleedings.

The described PAVM-related complications also include intracranial hemorrhage, cerebral abscess, pleural hematoma, hemoptysis. The complications occur more frequently in the 5th and 6th decades of life [1,5–8].

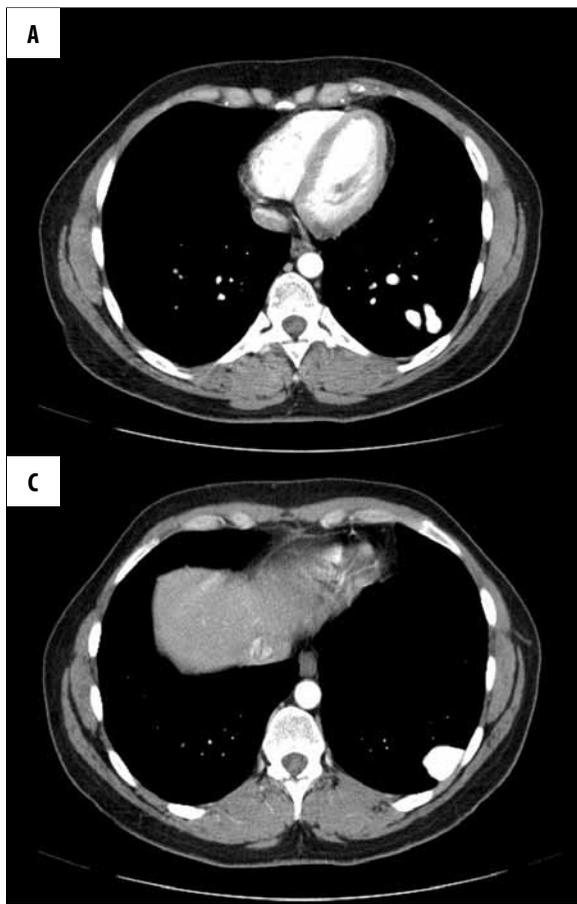


Figure 3. (A–C) CT axial images after injection of contrast medium, arterial phase, at the level of PAVM in the posterior basal segment of the left lung.

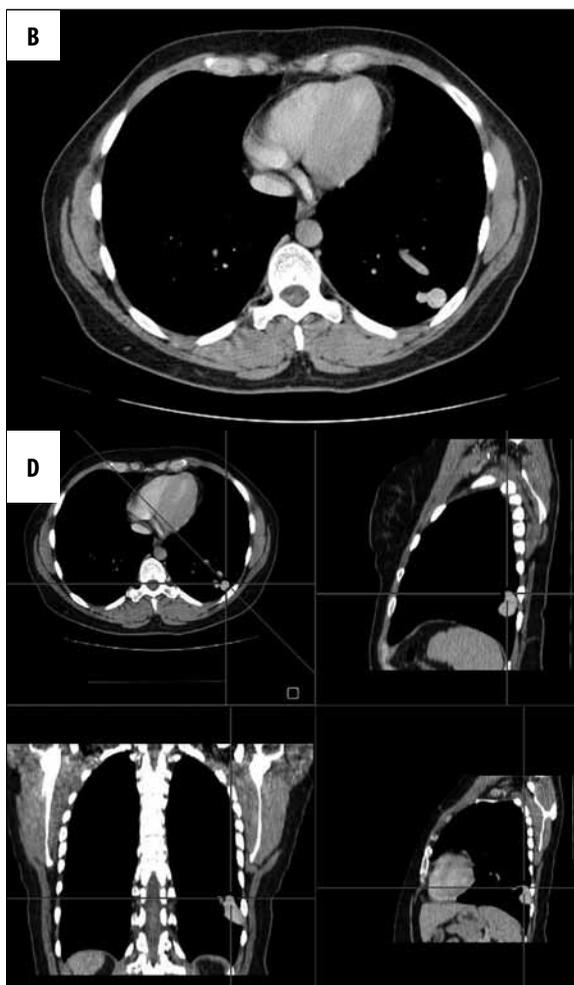
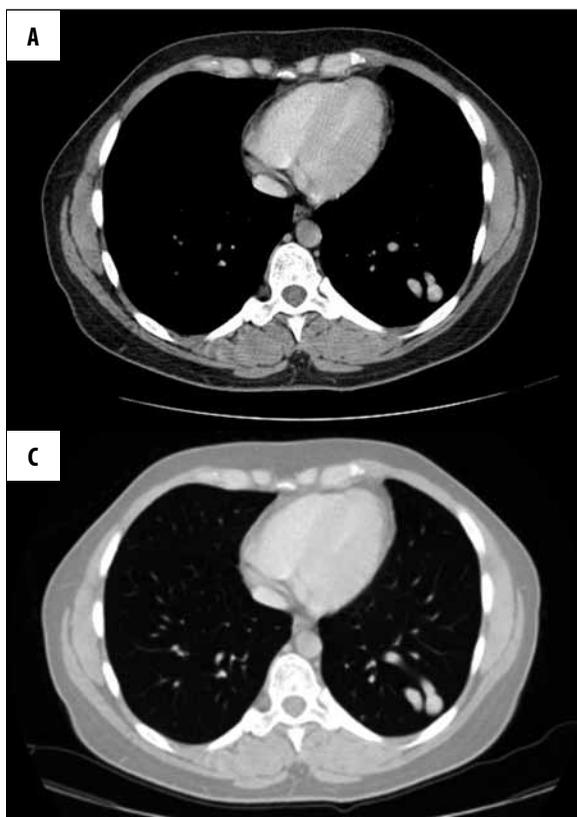


Figure 4. CT after injection of contrast medium, venous phase. Axial plane images at the level of PAVM in the posterior basal segment of the left lung mediastinal window (A, B), lung window (C). MPR reconstruction (D).

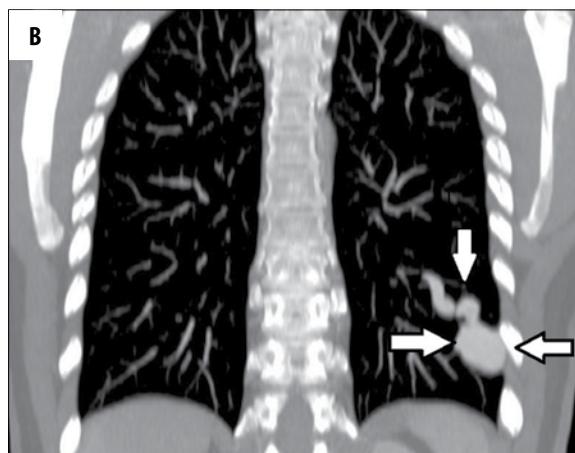
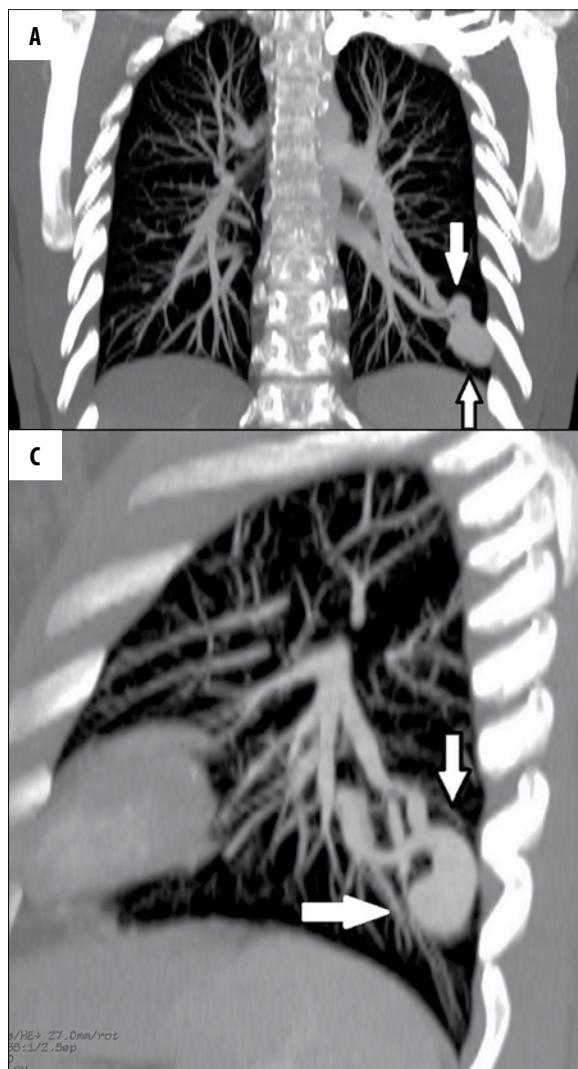


Figure 5. (A–C) CT after injection of contrast medium, arterial phase – MPR reconstruction. PAVM (arrows).

A multiphase CT scan of the chest demonstrated the presence of circumferential arteriovenous fistula located in the posterior basal segment of the left lower lobe, which corresponded to the rounded opacity visible in x-ray image in this location (Figures 2–6).

Discussion

Despite very rare occurrence of pulmonary arteriovenous malformation (PAVM) one should bear the abnormality in mind while performing differential diagnostic procedures of the rounded opacity in the lungs, particularly in case of patients with Osler-Weber-Rendu disease, cyanosis, hypoxemia and particularly in patients with whom we, the radiologists, meet more frequently and who experience recurrent respiratory tract infections, hemoptysis, pleural hematoma or intracranial hemorrhage, cerebral abscess [8–11].

It is difficult to make an unambiguous diagnosis having only a classic chest radiograph (x-ray scopy may be helpful – yet how many of us remember about and perform that procedure on a daily basis?), however a CT scan, and in particular a CT angiography, which is currently the “gold standard” in diagnosing the pathology, enables to make a certain diagnosis as to the nature of the pathology, even if the pathology is only read about in literature [1,5,9–11].

Making the correct diagnosis of PAVM is important due to the risk of complications, including the occurrence of the most severe ones, such as intrapulmonary and intracranial hemorrhages and the need to perform a surgical procedure: embolization and cardiothoracic surgery [8–11].

Conclusions

1. Routine chest radiograph in two basic views (PA and lateral) demonstrated presence of rounded opacity in the posterior basal segment of the left lower lobe, however it was not sufficient to state certain diagnosis, the differential diagnostics allowed for the presence of a pulmonary tumor, developmental anomaly.
2. The chest CT examination with unenhanced and enhanced scans allowed to differentiate the described lesion and to undoubtedly diagnose arteriovenous

Currently, the diagnostic ‘gold standard’ for this pathology is CT-angiography and the treatment of choice is embolization or cardiothoracic surgery [1,5–8].

Case Report

A 33-year-old female with no previous disease history underwent routine chest x-ray examination (in PA and lateral views) before being offered employment contract.

The performed radiographs demonstrated the presence of clear-bordered rounded opacity in the posterior basal segment of the left lower lobe (Figure 1).

Due to the above the performance of a chest CT scan examination was ordered in our laboratory for the purpose of evaluating the lesion differentiation.

The data supplied by the referring physician indicated that the basic laboratory tests performed in case of that patient were within normal limits, no x-ray examination of the chest had been performed so far and the patient had negative family history.

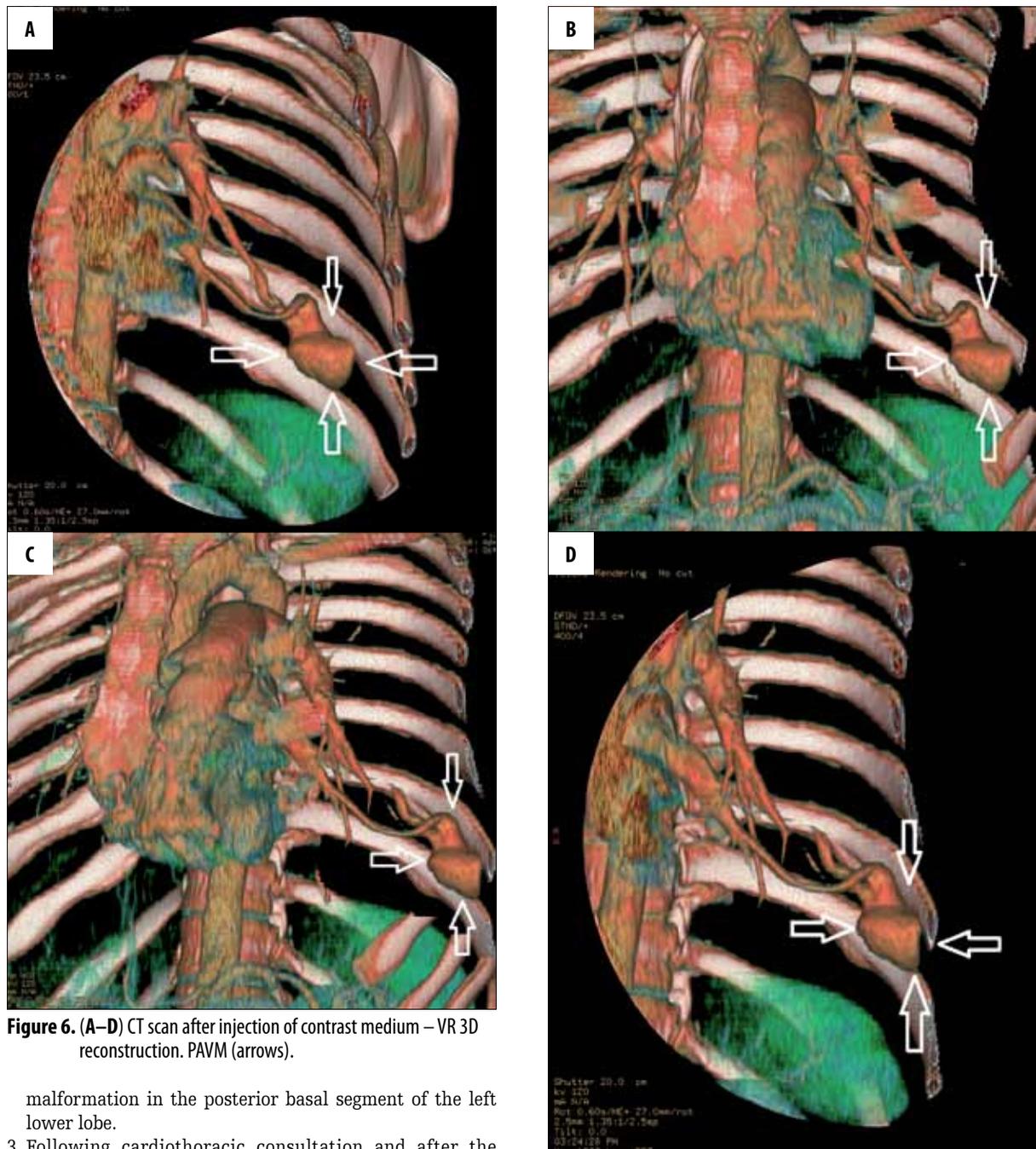


Figure 6. (A–D) CT scan after injection of contrast medium – VR 3D reconstruction. PAVM (arrows).

malformation in the posterior basal segment of the left lower lobe.

3. Following cardiothoracic consultation and after the patient was provided with information on the type of the pathology observed in the performed imaging examinations the patient gave no consent to further medical

treatment. The patient remains in constant thoracic surgery follow-up.

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