REVIEW / Thoracic imaging

Diagnosis and treatment of pulmonary arteriovenous malformations in hereditary hemorrhagic telangiectasia: An overview


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KEYWORDS
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Abstract Hereditary hemorrhagic telangiectasia (HHT) or Rendu-Osler-Weber disease is an autosomic dominant disorder, which is characterized by the development of multiple arteriovenous malformations in either the skin, mucous membranes, and/or visceral organs. Pulmonary arteriovenous malformations (PAVMs) may either rupture, and lead to life-threatening hemoptysis/hemothorax or be responsible for a right-to-left shunting leading to paradoxical embolism, causing stroke or cerebral abscess. PAVMs patients should systematically be screened as the spontaneous complication rate is high, by reaching almost 50%. Neurological complications rate is considerably higher in patients presenting with diffuse pulmonary involvement. PAVM diagnosis is mainly based upon transthoracic contrast echocardiography and CT scanner examination. The latter also allows the planification of treatments to adopt, which consists of percutaneous embolization, having replaced surgery in most of the cases. The anchor technique consists of percutaneous coil embolization of the afferent pulmonary arteries of the PAVM, by firstly placing a coil into a small afferent arterial branch closely upstream the PAVM. Enhanced contrast CT scanner is the key follow-up examination that depicts the PAVM enlargement, indicating the various mechanisms of PAVM reperfusion. When performed by experienced operators as the prime treatment, percutaneous embolization of PAVMs, is a safe, efficient and sustained therapy in the great majority of HHT patients.

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Introduction

Hereditary hemorrhagic telangiectasia (HHT) or Rendu-Osler-Weber disease is an autosomic dominant disorder, which occurs in approximately 10 to 20 individuals per 100,000, characterized by the development of multiple arteriovenous malformations of the skin, mucous membranes, and visceral organs including cerebral, spinal, hepatic, pancreatic and pulmonary arteriovenous malformations (PAVMs) [1–6].

Two main types of HHT disease represent 80% of the cases and are due to the mutations in the ENG gene (encoding for endoglin) and activin type-II-like receptor kinase 1 (ACVRL1) gene (encoding for the activin receptor like kinase (ALK1)), causing respectively HHT type 1 and HHT type 2 diseases. The ENG and ACVRL1 genes code for proteins of the TGF beta receptor, which is involved in the proper blood vessel development [7]. The mutation of the SMAD4 gene, which is involved in TGF beta signal transduction, may also cause HHT disease, in association with juvenile polyposis [8].

HHT should be precisely clinically diagnosed on the basis of the Curaçao criteria. Three criteria are thus needed among the following:

- multiple mucocutaneous telangiectases;
- spontaneous and recurrent epistaxis;
- visceral involvement;
- a family first degree history of HHT [9].

Penetrance is age-related and is almost complete by the age of 40-years-old [1]. Mild to moderate epistaxis is the most common symptom of HHT. Cerebral vascular malformations have been reported in 3.7% out of 321 HHT patients, and included ten cases of AVM, one dural arteriovenous fistula and one cavernomatous malformation [10]. A substantial proportion of patients presenting with HHT disease may be affected by liver involvement, which can cause abdominal pain, abnormal serum liver function tests results, pseudocirrhosis, biliary necrosis and even high cardiac output failure [11]. Pancreatic involvement has been reported in 31% of the cases, mainly in association with ALK1 mutation patients [12].

Pulmonary arteriovenous malformations represent the most common pattern of the pulmonary involvement to be encountered in HHT patients. ALK1-type related pulmonary arterial hypertension may also be observed [13]. Paradoxical embolism due to de novo in situ curoic thrombus originating from the PAVM (tight stenosis related venous stasis, turbulent flow within a large sac) or septic, curoic thrombus or air embole originating upstream the PAVMs is one of the main causes of morbidity, responsible for stroke and cerebral abscess events [14]. PAVMs may also rupture, causing life threatening hemoptysis or hemotherax [15].

This overview article aims:

- firstly, to describe the progressive development and radiological characteristics of PAVMs, and their potential complications;
- secondly, to report the technical aspects and outcomes of percutaneous embolization to prevent and treat PAVMs related complications;
- thirdly, to show CT scanner/angiographic — pathological correlations.

Pulmonary arteriovenous malformations

Pulmonary arteriovenous malformation anatomy and classification

Pulmonary arteriovenous malformations are abnormal direct communications between pulmonary arteries and pulmonary veins without interposition of a capillary bed. Approximately 80 to 90% of patients presenting with PAVMs eventually present HHT, whereas the remaining are sporadic cases [16,14]. Conversely, 15 to 35% of HHT patients will present PAVMs [16,14]. The PAVMs have a natural tendency to increase in size over time. Various factors including puberty, pregnancy and also pulmonary arterial hypertension (PAH) may explain this phenomenon. Contrary to sporadic forms, HHT-PAVMs are often multiple, bilateral and preferentially located at the pulmonary bases [14,17].

The PAVM consists of three different anatomical components: one or more than one feeding artery(ies), an aneurismal sac and one or more draining vein (Fig. 1). The feeding artery may be long, (Fig. 1) or very short, thus making percutaneous embolization more challenging (Fig. 2). The aneurismal part of the PAVM may be either a sac or a serpiginous network (Figs. 1, 3, 4). Distinction between these two different patterns is crucial to plan the appropriate percutaneous treatment as the risk of accidental embolization device migration to the left heart is higher when considering a single aneurismal sac directly communicating with the left circulation.

Finally, the third portion of the PAVM consists of one or more draining veins whose diameters are larger than...
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Figure 2. Short solitary afferent artery feeding a simple large pulmonary arteriovenous malformation (PAVM) in the left lung in a 79-years-old woman presenting with multiple discrete PAVMs. Selective angiogram of the left latero basal artery in a 45 degrees right anterior oblique view before (a) and during embolization (b) discloses a very short subsegmental anterior artery (arrow) feeding the large aneurismal sac (arrowhead). The anchor technique permitted the coil blockage inside the subsegmental branch (large arrow). Coils are shown into the aneurismal sac (curved arrow).

the afferent feeding artery (Figs. 1 and 5). A simple PAVM (approximately 80% of the cases) is characterized by one or more afferent feeding arteries originating from a single segmental pulmonary artery (Fig. 1), while a complex PAVM has multiple afferent feeding arteries originating from several segmental arteries (Fig. 4) [14,18].

PAVMs development comprises three main steps as follows:

- a ground glass nodule corresponds to the initial enlargement of the postcapillary venules associated with an inflammatory cell infiltrate;
- small vessels that are visible within the ground glass nodule represent vascular branching and connection between the precapillary pulmonary artery and the postcapillary venules, throughout the capillary bed;
- enlargement of the draining vein;
- the definitive PAVM corresponds to a feeding pulmonary artery, an aneurysmal sac and an enlarged draining vein with concomitant disappearance of the ground glass pattern (Fig. 6) [19]. At this stage, the PAVM will evolve for its own account. Rarely, PAVMs will evolve to a giant form (Fig. 7).

Figure 3. Various patterns of aneurismal arterio venous connections. Thirty degrees left anterior angiogram of a subsegmental branch of the posterior segment of the left upper lobe (a) shows a serpiginous network channel (arrow) drained by two efferent veins. Thirty degrees right anterior oblique angiogram of a latero basal segmental artery (b) shows stenosis of the arteriovenous connection (large arrow). The risk of accidental distal device migration to the left circulation is higher in case of a large straight channel aneurismal sac. The presence of a stenosis of a pulmonary arteriovenous malformation (PAVM) may represent a local factor for spontaneous PAVM thrombosis.
Figure 4. Complex pulmonary arteriovenous malformation (PAVM) with afferent feeding arteries originating from different segmental pulmonary arteries. 82-years-old hypoxemic female with a persistent positive transthoracic echocardiography (grade 3) after percutaneous closure of a patent foramen ovale. Thirty degrees left anterior oblique volume rendering CT scanner shows a complex PAVM fed by two afferent arteries: a small lingular artery (arrow) and a large anterobasal artery (curved arrow). The CT scanner also shows a serpiginous aneurismal sac (arrowhead) and an enlarged draining vein (large arrow).

Figure 5. Simple pulmonary arteriovenous malformation (PAVM) presenting with an enlarged draining vein. Twenty degrees right anterior oblique angiogram of a left anterior basal artery shows a simple PAVM presenting a fusiform aneurism of the draining vein (arrow). The pattern of draining veins is presumably related to the high flow output within the PAVM and/or to the pulmonary hypertension. Moreover, angiogram displays dynamic variations of the filling of draining veins according to the cardiac cycle. In case of a high output cardiac failure with pulmonary venous hypertension, a retrograde filling of a successfully embolized PAVM can be observed (Fig. 21).

Figure 6. Spectrum of pulmonary arteriovenous malformation (PAVM) disease development on CT scanner. CT scanner first step shows isolated nodular or ill defined ground-glass attenuation. It corresponds to the initial venous telangiectatic stage. This initial telangiectatic stage is followed by the pulmonary venous enlargement, which correspond to microscopic arteriovenous connections. In next stages, vascular branching is depicted on CT: the feeding artery lies within the ground-glass nodule, while draining veins are visible in the periphery of the nodule. Finally the development of a true PAVM occurs with an aneurismal sac and concomitant disappearance of the nodule or ground glass densities.

Pulmonary arteriovenous malformation repartition

The repartition type of the PAVM in HHT patient lungs may be either discrete or diffuse (5–7%). Diffuse pattern of PAVMs is variably defined as PAVMs involving every segmental or every subsegmental artery of at least one lobe [20]. Diffuse involvement has also minimally been defined as one segment diffusely involved [21]. A more clinically relevant classification has been proposed, as follows: patients presenting with involvement of every subsegmental artery of at least one lobe (diffuse subsegmental form) (Fig. 8), patients presenting with involvement of every segmental artery of at least one lobe (diffuse segmental form) (Fig. 9), and patients presenting with a combination of subsegmental and segmental involvement of at least one lobe (diffuse mixed segmental/subsegmental form) [22]. Symptoms that include hypoxemia, hemoptysis, and neurological symptoms are more frequently correlated to diffuse patterns [20]. Paradoxical embolism is more frequently encountered in the diffuse subsegmental form [22].

Spontaneous pulmonary arteriovenous malformations complications

PAVMs may be responsible for a paradoxical clot embolism due to the absence of the capillary filter bed interposition between the arterial and venous circulation. As a matter of fact, stroke and cerebral abscess events are uncontrolled right-to-left shunt related complications due to clot/septic thrombus embolization through out the PAVM [14]. The right-to-left blood shunting may cause hypoxia...
and subsequent polycythaemia (Fig. 7) [23]. Spontaneous thrombosis of a PAVM is rare and may be symptomatic and responsible for embolism to the left circulation (Fig. 10). The thin-walled PAVM aneurismal sac may also rupture. Most of the PAVMs have a subpleural localisation and may be revealed by a combination of potential life threatening hemoptysis/hemothorax and neurological signs due to cough favoured systemic air embolism (when rupture occurs into lung parenchyma, alveolar haemorrhage triggers patient’s cough, increased intrathoracic pressure and subsequent paradoxical air embolism through the parietal PAVM tear) (Figs. 11 and 12). Ruptures may be favoured by pregnancy probably due to the increased blood volume as well as alterations in the vascular tone related to changing serum estrogen and progesterone concentrations [15, 24].

PAH may occur and be favoured by both pulmonary arterial vasoconstriction secondary to PAVMs induced chronic hypoxemia and high blood flow output due to intrahepatic shunts (arterioportal, portohepatic and/or arteriohepatic shunts) [25]. ALK1 and more rarely endoglin mutations may also be a proper cause of heritable PAH [13, 14, 26]. Also, PAH may contribute to the PAVM enlargement [3].

Diagnosis of pulmonary arteriovenous malformation
Transhiobaric Contrast Echocardiography (TTCE) and Chest CT scanner examination are the two main tools permitting
Figure 8. Subsegmental diffuse pulmonary arteriovenous malformation of the middle lobe in a 31-years-old female patient presenting with a low PaO2 level (34 mm Hg). CT scanner (a) showing all peripheral subpleural subsegmental arterial branches dysplaying tiny arteriovenous connections (arrows). Selective angiogram of the left latero basal segment (b), in a right anterior projection showing diffuse subsegmental arterio venous connections (arrows).

PAVMs screening and evaluation in patients presenting with HHT [26]. Both TTCE and CT scanner examinations should be performed carefully to prevent intravenous gaseous or septic injection that may lead to iatrogenic paradoxical embolism. Isotopic and blood oxygen levels right-to-left shunt measurements are insufficiently sensitive to exclude PAVMs diagnosis [26,27]. Chest radiograph is an easy, fast, low radiation exposure examination used as a first line PAVM screening test although its sensitivity is low, particularly for the detection of small diameter PAVM. PAVMs appear as rounded well circumscribed lesions with branching afferent feeding and dilated efferent draining vessels (Fig. 13) [27]. Although contrast-enhanced magnetic resonance angiography (MRA) is not currently performed for PAVMs screening, recent studies have concluded that MRA was a contributive adjunct to pre-embolization planning [28–30]. The main advantage of MRA over the other techniques is to avoiding ionizing radiation exposure.

TTCE is a rapid simple and minimally invasive examination used as an initial PAVMs screening examination.

Figure 9. Axial MIP CT scanner shows diffuse segmental pulmonary arteriovenous malformation (PAVM) of the middle lobe. All the segmentar arteries of the middle lobe display arteriovenous malformations (arrows). Multiple discrete PAVMs are shown within the right lower lobe (arrowheads).

Figure 10. Asymptomatic partial thrombosis of the pulmonary arteriovenous malformation (PAVM) aneurismal sac. Contrast-enhanced CT scanner in a 34-years-old female patient without any clinical signs of systemic paradoxical cruric or septic embolism. Axial reformation (a) shows a thrombus within the aneurismal sac of a PAVM in the anterior segment of the right upper lobe (arrow). Anticoagulant therapy was planned for one month. Pre-embolization angiogram disclosed a tiny residual clot abutting the feeding artery (not shown). Embolization was successfully performed. Angiogram (b) showing an incidental clot within the aneurismal sac of a simple PAVM of the laterobasal segment of the left inferior lobe (arrow). Immediate embolization was performed without any immediate or delayed complication.
Diagnosis

Microbubbles

Interestingly, established echographic scanner could predict tissue hypervascularity [31]. After intravenous injection of agitated saline solution, echoic microbubbles can be easily depicted in the left cardiac cavities after two to five cardiac beats, thus suggesting the presence of a right-to-left pulmonary shunt (whereas microbubbles that are immediately visible after injection may suggest the presence of an intra cardiac shunt) (Fig. 13). Interestingly, a grading of positive TTCE results has been established according to the number of microbubbles that could be detected (Fig. 14) [31,32]. The reported diagnostic sensitivity of TTCE is high (up to 97%) and the negative predictive value is 99% in published series where chest CT scanner was used as the gold standard for PAVMs diagnosis [33].

Indeed, CT scanner is considered as the gold standard diagnosis tool as it provides an easy patient screening, a high anatomical resolution, a precise location (multiple discrete or diffuse form) and type definition (e.g. simple versus complex) of the PAVM. Furthermore, this tool is useful for percutaneous embolization planification and follow-up [34].

As a matter of fact, we usually perform a contrast-enhanced thoraco abdominal CT examination at our institution. This allows us to depict the PAVM feeding vessels and the aneurismal sac lumen that can be spontaneously thrombosed before treatment (Fig. 10), to detect the PAVMs systemic supply (Fig. 15), and the subphrenic specific visceral involvement (liver and/or pancreatic involvement)

Figure 11. Acute hemothorax revealing a ruptured subpleural pulmonary arteriovenous malformation (PAVM). A 56-years-old female patient presenting with acute hemothorax and systemic hypotension due to intra pleural rupture of a peripheral PAVM during transatlantic air flight travel. The diagnosis of hereditary hemorrhagic telangiectasia (HHT) was suspected on recurrent spontaneous epistaxis and multiple cutaneous telangiectases. Sagittal (a) contrast-enhanced CT scanner reformation shows the PAVM herniation into the pleural space (arrow), hematic pleural effusion (large arrow) and a large fresh clot adjacent to the PAVM (arrowhead). The diagnosis of HHT was confirmed by the presence of the pulmonary and hepatic involvement. Prior to embolization, pulmonary artery pressure level assessment indicated severe pulmonary hypertension (systolic 92, diastolic 35, mean, 56 mmHg), secondary to a severe HHT hepatic involvement. Percutaneous emergency embolization was successfully performed. Angiograms (b and c) show a tortuous afferent feeding artery and a tiny PAVM wall rupture (arrow). Coils were successfully anchored into the artery and the aneurismal sac (arrowhead). Pulmonary artery pressures remained unchanged after embolization. Indication for anti-angiogenic (Bevacizumab) treatment of the hepatic involvement of HHT was discussed.
Spontaneous pulmonary rupture of a pulmonary arteriovenous malformation (PAVM). This 35-years-old man presented a sudden cough immediately followed by hemoptysis and concomitant transient right hemiparesis. PAVM parenchymal rupture (hemoptysis) of a PAVM and a systemic cerebral gas embolism (that may be facilitated by cough) were suspected. Contrast-enhanced CT scanner examination revealed alveolar hemorrhage related ground glass attenuation (arrow) circumscribing the fissured PAVM aneurismal sac. Emergency embolization of the culprit PAVM was successfully performed, as well as embolization of three other non-complicated PAVMs. The patient remains asymptomatic ten years after the initial onset of symptoms.

In patients presenting without PAVM, identification of liver/pancreatic involvement provides the third visceral diagnostic item and subsequently helps to early clinical diagnosis and improved management of HHT disease. A steal phenomenon might be encountered on both angiographic and CT scanner examination due to the PAVMs high blood flow output (Fig. 16) in a low-resistance vascular system, and

to the positive pressure gradient directed from the feeding arteries to the lesion center. As such, the afferent feeding arteries, particularly the PAVM accessory feeders, may thus be overlooked on imaging studies (Fig. 16).

A systemic supply has been reported in 29% of 70 pre-embolization work-up PAVMs patients (Fig. 15). A relationship between the steal phenomenon (which decreases the local pulmonary arterial vascularization) and the development of a systemic supply to PAVMs has not been established to date.

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Treatment of pulmonary arteriovenous malformation

PAVMs should be treated before the complication may appear, owing to the high complication rate reaching almost 50%, and even more during pregnancy [21]. In diffuse clinical forms, the neurological morbidity (stroke and brain abscess) of untreated PAVMs is higher and reaches 70% [20]. This underlines the necessity of screening PAVMs by using non-invasive tests in HHT families, in both children and adults [15].

It is recommended that patients presenting with PAVMs should undergo antibiotic prophylaxis prior to dental and surgical interventions to reduce paradoxical embolic abscess risks [6].

The risk of cerebral complications is usually considered significant when the feeding artery of the PAVM exceeds 3 mm in diameter, and may also increase in patients with multiple PAVMs [35]. The 3 mm cut-off size is solely based on empirical data regarding patients who never experienced such cerebral ischemic events below that size [35,36].

However, Antibiotic prophylaxis remains recommended if TTCE has positive findings, even if CT scanner does not show any image suggestive for PAVM [6].

Surgery has long been the treatment of reference until the seventies, including pneumonectomy, lobectomy, segmentectomy, wedge resection or vascular ligation (Fig. 7) [14]. Pulmonary transplantation has been performed in diffuse forms associated with respiratory insufficiency [37]. Percutaneous image-guided embolotherapy is nowadays preferable in most disease cases as this avoids major surgery, general anesthesia, and loss of pulmonary parenchyma function. The aim of transcatheter embolization is to occlude all the PAVM feeding arteries by a selective catheterization of pulmonary arteries by using a coaxial system, via a percutaneous femoral approach. CT scanner is the key examination tool for the planification of percutaneous embolization. Firstly, CT scanner determines the number and location of the PAVMs, secondly their precise angioarchitecture characteristics, thirdly the presence of complications (e.g. thrombosis) [34]. The device diameter is chosen according to the measurement data of the afferent feeding artery

Figure 16. Pulmonary arteriovenous malformation (PAVM) related steal phenomenon. Frontal Mini MIP CT scanner (a) shows a subpleural peripheral low-density area distal to a simple PAVM thought to correspond to the vascular steal phenomenon (arrow). Right common A7-A8 arterial trunk selective angiogram (b) shows the main feeding arteries of the simple PAVMs of A7 (arrowhead) and A8. The A8 feeding artery looks very long owing to the reverse flow related absence of collateral opacification (arrow). The collateral reverse flow upstream the PAVM is favoured by the low-resistance high flow. After successful A7-A8 PAVMs embolization occlusion (c), A8 collateral flow (arrows) is reopacified, due to the steal phenomenon disappearance.
Results of pulmonary arteriovenous malformation transcatheter embolization and follow-up

Successful transcatheter embolization is assessed by complete retraction of the embolized PAVM (afferent artery located downstream the coils/aneurismal sac) on a follow-up control CT scanner (Figs. 18 and 19). However, TTCE still remains positive in 80–90% of successfully treated patients [40]. This may be due to the occult presence of tiny additional PAVMs that were not detected by both conventional angiography and CT scanner examinations or the presence of a tiny PAVMs feeding artery too small to be catheterized [40]. Transcatheter embolization of PAVMs is eventually efficient and durable in the majority of patients (83% of patients in Mager’s study) [41—43].

Immediate complications are rare in experienced hands and include device migration, gazeous embolism, stroke,

Figure 17. Therapeutic anchor technique. The therapeutic anchor technique consists of placing the first coil into a small normal afferent branch, closely upstream the pulmonary arteriovenous malformation (PAVM) (arrow). The first coil creates a scaffold which facilitates the placement of other coils, lowering the risk of migration of embolization devices through the PAVM. Progressive occlusion of the feeding artery is performed by using complementary coils, of decreasing diameters. Coil packing is mandatory to achieve a complete lumen occlusion of the feeding artery (arrowhead). Success of percutaneous embolization is assessed by the complete retraction of the aneurismal sac over time.

Figure 18. Successful transcatheter embolization. Axial CT scanner (a) shows a complex racemous sporadic pulmonary arteriovenous malformation (PAVM) of the apical segment of the right inferior lobe (arrow) in a cyanotic 18-years-old female with clubbing. Post-embolization axial CT scanner (b) shows coil occlusion of the afferent feeding artery and subsequent complete retraction of the PAVM (arrow).
pulmonary infarctions and hemoptysis (Figs. 20 and 21). Regarding mid- to long-term complications, reperfusion of the embolized PAVM may be due to the following mechanisms, as follows: reperfusion through or around the anchored coils (Figs. 22 and 23); development of pulmonary to PAVM neovasculature anastomoses (Fig. 23), reperfusion by systemic bronchial and/or non bronchial arteries (Fig. 24) [41–43]. In case of systemic bronchial artery supply, post-embolization systemic supply may be responsible for delayed hemoptysis.

CT scanner follow-up examination may highlight abnormal secondary PAVM reperfusion—indirectly by showing incomplete PAVM retraction—and directly by using I.V. contrast iodinated medium injection [44,45]. CT scanner imaging follow-up is mandatory every year in subsegmental diffuse PAVMs types, and every five years in the other cases, owing to the risk of late PAVM recanalization and regrowth of untreated PAVMs [5,41–43].

Figure 19. Pulmonary arteriovenous malformation (PAVM) incomplete retraction at follow-up corresponding to secondary recanalization. Work-up axial enhanced CT scanner (a) shows a simple peripheral PAVM in the anterior segment of the right lower lobe (arrow). Post-embolization CT scanner one year after embolization (b) shows persistent or recurrent contrast enhancement of the partially retracted PAVM (arrow). Central recanalization through the previously deposited coils was disclosed on angiography.

Figure 20. Acute pulmonary ischemia after percutaneous pulmonary arteriovenous malformation embolization. Axial CT scanner shows multiple areas of pleural base pulmonary infarction and left pleural effusion 10 days after bilateral embolization (arrows).

Figure 21. Post-embolization pulmonary arteriovenous malformation (PAVM) thrombosis. This 36-years-old man presenting with multiple PAVMs and liver involvement with hepatic artery enlargement (11 mm). Enhanced CT scanner was performed at stroke onset (hemiplegia) seven hours after PAVMs embolization. The feeding artery was completely occluded but a clot is shown within the aneurismal sac extending downstream reaching the draining vein (arrow). The enhancement of the aneurismal sac comes from a retrograde filling of the pulmonary vein related to the high cardiac output secondary to the liver involvement. On cerebral MR imaging, a non obstructive clot was detected within the middle cerebral artery, without cerebral infarction. Anticoagulant therapy was initiated and patient had an immediate full clinical recovery. In our practice, anticoagulation is initiated after PAVM embolization as soon as a retrograde filling of the embolized PAVM is depicted on control angiography.
Figure 22. Recanalization around the previously deposited coils two years after pulmonary arteriovenous malformation (PAVM) embolization. Imaging work-up before re treatment. Enhanced oblique MIP CT scanner (a) shows recanalization of a previously treated right laterobasal PAVM. A recurrent flow through previously deposited coils is present. The feeding artery of the PAVM is shown patent immediately beyond coils. Selective angiography (b) shows reopening channels around the anchored coils (arrow).

Figure 23. Pulmonary arteriovenous malformation (PAVM) recanalization through the anchored coils and pulmonary-to-PAVM anastomoses. Angiogram shows the recanalization of the afferent feeding artery (left A10) throughout previously thrombosed anchored coils (arrow) and the development of pulmonary-to-PAVM neovasculature anastomoses (large arrow). Both mechanisms explain the opacification of the draining vein (arrowhead).
Diagnosis

Reperfusion of pulmonary arteriovenous malformations (PAVMs) due to post-embolization systemic neovascularization development. High cardiac output failure in a 33-years-old female presenting with diffuse subsegmental form of PAVMs throughout both lungs five years after serial peripheral-to-proximal percutaneous embolization. Axial enhanced CT scanner (a) shows huge enlargement of systemic bronchial (arrow), intercostal and internal thoracic arteries (large arrow), and concomitant reperfusion of a left anterior PAVM (arrowhead). Axial enhanced CT scanner examination (b) shows huge enlargement of both inferior phrenic arteries (arrows) with retrograde filling of multiple peripheral embolized PAVMs (arrowheads). This reperfusion can be overlooked on angiograms because of the retrograde filling of feeding arteries via systemic supplies (false negative angiograms).

Conclusion

PAVMs are one of the main causes of morbidity in patients presenting with HHT disease, owing to the risks of rupture as well as paradoxical embolism exposing to stroke and/or cerebral abscess. PAVMs should be carefully screened in the patient and own family by using transthoracic contrast echocardiography and CT scanner examination. Percutaneous embolization has become the treatment of choice of PAVM, due to its safety and efficiency in experienced hands.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


