Hemoptysis in Hereditary Hemorrhagic Telangiectasia (HHT)

one symptom, several mechanisms

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INTRODUCTION

- The lung is a typical location of arteriovenous malformations (PAVMs) in HHT patients.
- Complications may occur in about 50% of patients with PAVMs, including cerebral stroke, cerebral abscess, hemoptysis, hemothorax, pulmonary hypertension.
- May be prevented by transcatheter embolization of PAVMs, when the diameter of the feeding artery is higher than 2.5 to 3 mm.
OBJECTIVES

• **Hemoptysis can occur before, during and after embolotherapy** in several mechanisms.

• This presentation illustrates different mechanisms and discusses the management of such episodes.
MATERIALS AND METHODS

• **During a 12-year period, 970 patients with HHT were evaluated** at our Center in Paris

• 425 patients had PAVMs and 246 of them were treated by embolization

• **Five percent of all patients presented hemoptysis**, before, during or after embolization

• All patients with hemoptysis underwent bronchoscopy, computed tomography of the thorax and/or angiography
HEMOPTYSIS BEFORE EMBOLIZATION
Spontaneous rupture of PAVMs

- Hemothorax and/or hemoptysis, possibly lethal, typically associated with neurological manifestations.
- Can occur at any age, even in childhood, and also with small PAVMs.
- Pregnancy increases the risk of rupture by increasing the size of PAVMs.
- Urgent embolization of ruptured PAVM is required.
HEMOPTYSIS DURING EMBOLIZATION
Risk factors

- Pulmonary Arterial Hypertension (PAH) [mean pulmonary arterial pressure (mPAP) > 25mmHg]
- Technically complicated catheterization (distal PAVM, tortuous vessels, recurrent course...)

- Sudden cough during embolization
- Well tolerated and responsible for small amount hemoptysis, except in severe PAH
Difficult catheterization:

Angiographic features before and during treatment:
Appearance of a ground glass opacity (green arrow) surrounding coils. Occlusion with steel and platinum coils stopped the bleeding.

Coronal CT scan:
ground glass opacity surrounding coils (yellow arrow)

Control CT scan 2 months later
Rupture associated with Pulmonary Arterial Hypertension

Pre-treatment CT scan and arteriography:
Large pulmonary arteries, intra-parenchymal aneurisms and tortuosity of pulmonary arteries (arrow) due to Pulmonary Arterial Hypertension.
The CT scan after procedure shows a ground-glass opacity surrounding coils in segment S9.

Arteriography before and after embolization:
The rupture was suspected because of sudden cough immediately associated with hemoptysis, which resolved after embolization.
HEMOPTYSIS AFTER EMBOLIZATION
HEMOPTYSIS AFTER EMBOLIZATION

• Occurs by reperfusion of embolized PAVMs
• Even in case of successful embolization
• Three main mechanisms
• Additional embolization is effective in all mechanisms
Reperfusion by recanalization (through previously deposited coils)

- Most common mechanism of reperfusion (80% of cases)
- Occurs in 20% of PAVMs treated with coils
- Is associated with:
  - increased feeding artery diameter (large PAVMs)
  - use of a small number of coils
  - use of too oversized coils
  - too proximal placement of the coil (more than 1cm from the aneurismal sac)
- May also occur when using vascular plug
25 y.o. pregnant woman already treated for PAVM who presented hemoptysis.

Pre-treatment CT reveals ground glass opacity (arrow) surrounding the previously embolized PAVM (curved arrow).

Arteriography:
Recanalization of PAVM due to misplacement of coils into the aneurismal sac and reperfusion by missed side branch.

Angiographic control after packing coils of both pedicles without any recanalization. Caesarian section at the 35th week.
Reperfusion by pulmonary-to-PAVMs anastomoses

- By development or appearance of vascular network towards the PAVM
- Occurs when coil’s placement is too proximal
- Post-procedural pulmonary angiography allows a better detection of accessory branches supplying the PAVM
15y.o., already treated during childhood for a complex PAVM referred to our center for hemoptysis. Angiography demonstrated reperfusion.
Arteriography (on the left) and CT scan (on the right) identify dysplastic anastomoses towards the PAVM (curved arrow) presumably responsible for hemoptysis (arrow).

Selective catheterization (on the left) demonstrates another mechanism of reperfusion by recanalization of the arteries previously embolized. The CT scan feature (on the right) shows that the diameter of the artery beyond the coils is too large (3mm).
Reperfusion by systemic supply (1)

- Systemic supply present before embolization (30%) (steal syndrome around the PAVM) or after, due to a too proximal embolization.
- The risks of hemoptysis increases because the subsequent systemic supply is also connected to a normal pulmonary capillary bed.
- Massive hemoptysis may occur, without any prodrome and even if the PAVMs were accurately occluded.
Reperfusion by systemic supply (2)

- Embolization of systemic vessels is recommended in patients with recurrent post-embolization hemoptysis.
- Don’t use small sized micro-particles to avoid systemic emboli of particles.
15 y.o. boy, already embolized for multiple PAVMs. 4 years later, he presented bronchitis and small recurrent hemoptysis.

Chest CT scan identifies reperfusion of 2 PAVMs associated with development of systemic bronchial et non bronchial arteries.
Arteriography:
On the left:
Systemic pulmonary hypervascularization involving the first anterior intercostal branch of left internal thoracic artery (arrow)
Note the reperfusion of the PAVM (S2)(curved arrow)

On the right:
Systemic pulmonary hypervascularization involving left bronchial artery
Note the reperfusion of both (S2 and S9) PAVMs (curved arrow) and anterograde systemic-pulmonary shunt (arrow)
CT scan and arteriographic features of systemic supply to PAVMs

Left internal thoracic supply with retrograde systemic-pulmonary shunt (curved arrow). The enhancement of the anterior artery of left upper lobe is lower than that of the pulmonary artery, and identical to that of aorta.

Inferior phrenic supply

Right bronchial supply (arrow)
All 3 mechanisms of reperfusion may coexist

Chest CT scan:
Bleeding surrounding a complex PAVM of the right upper lobe (curved arrow)
Feeding artery already embolized (coils are too proximal) (green arrow) and bronchial arteries (yellow arrow) appear dilated. Recanalization? Systemic supply?

36 y.o., previously embolized for multiple PAVMs. She presented daily episodes of hemoptysis.
**Pulmonary arteriogram:**
2 pedicles to the PAVM, one inferior pedicle from a side branch (left) and a central recanalization of the superior pedicle (right).

**Broncho-systemic arteriogram:**
PAVM (curved arrow) is also reperfused by 2 bronchial arteries. Distal embolization of both pedicles with fibered coils was achieved. Control CT scan 2 months later revealed reperfusion by other systemic arteries leading to an upper lobectomy. The patient is presently asymptomatic (3 years of follow-up).
Spontaneous rupture of a previously non-embolized PAVM

• Small untreated PAVM can grow especially when a previous PAVM is treated (growth factor? redistribution of blood flow?...), in case of PAH and during pregnancy and puberty.
• Imaging and physiologic follow-up is recommended

Pulmonary infarction

- 10-15% of cases
- Often delayed (3 days up to 2 months after embolization)
- More frequent in patients with systemic collateral supply of PAVM
Don’t forget bronchial or tracheal telangiectasia or angioma:

- Responsible for small amount hemoptysis in patients treated or untreated.
- Only seen with fiberoptic bronchoscopy
CONCLUSION

• Hemoptysis is a rare but life-threatening presentation of HHT

• It can occur before, during and after embolization

• After embolization, there are three main mechanisms of hemoptysis potentially associated (recanalization, development of pulmonary anastomoses to PAVM and systemic supply)

• A careful analysis of pre-treatment CT scan and angiography may be helpful
How to prevent reperfusion?

- **Embolization should be as close as possible to the aneurismal sac** of the PAVM (1 cm before the sac), beyond any arterial branch to the normal lung.

- **The size of the coil is critical** (+20% oversizing).

- **Dense packing of the coils** (packing smaller coils into the first-placed coil = Scaffold technique) **is necessary to prevent recanalization**.

- Vascular plug is a good alternative but needs to fulfill the same requirements of distal embolization, oversizing and can be put in place together with additional coils.

- **Imaging follow-up is necessary** to screen any reperfusion or growth of non-embolized PAVM.
REFERENCES