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Case Report

Pulmonary Arteriovenous Fistula: A Clinical Case

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Abstract

The evolution of pulmonary arteriovenous malformations (PAVM) over time in children with hereditary hemorrhagic telangiectasia (HHT) is not well defined, this malformation of which cyanosis remains one of the main symptoms requires saving embolization which is the treatment of choice. Surgery is a treatment option that remains very heavy for a population that is often very young.

Keywords: Arteriovenous Malformations; Rendu-Osler; Embolization; Surgery

Introduction

Pulmonary arteriovenous fistula is a malformative communication between an artery and a pulmonary vein creating an extracardiac right left shunt resulting in impaired oxygenation in the pulmonary blood with risk of paradoxical embolism and brain abscesses. These pulmonary arteriovenous malformations are hereditary and are present in 47%-80% of patients with Hereditary Hemorrhagic Telangiectasia (HHT) also called Rendu- Osler-weber [1]. The prevalence of HHT is 1 in 5000 - 10,000 people [2]. Hereditary Hemorrhagic Telangiectasia is a constitutional vascular dysplasia combining hemorrhagic manifestations, mucocutaneous telangiectasias and visceral arteriovenous malformations. All of these hereditary disorders are transmitted in an autosomal dominant way following mutations in the ENG, ACVLI, GDF2 and SMAD4 genes [3].

Case Report

A 06-year-old patient who consults for chest pain. The interrogation revealed a history of Rendu-Osler disease, the clinical examination was of no particularity except a slight cyanosis of the lips. The chest X-ray objectified a poorly limited opacity right apical. Following the antecedents, a thoracic Angio scan was requested from the outset which made it possible to discover multiple right upper lobar systematized arteriovenous fistulas, with dilation of the homonymous pulmonary arteries and veins (Figure 1). As part of this disease a cardiac echodoppler and a cerebral CT were made returned without abnormalities. A right posterolateral thoracotomy in the 5th intercostal space (nonavailability of vedeo-assisted thoracoscopic surgery column in our department) made it possible to do a right upper pulmonary lobectomy (see Figure 2). The postoperative followup was simple, with removal of the chest drains on day 3 and discharge of the patient on day 4.

Figure 1: MAVP Scan Images.

Figure 2: Peroperative view of the MAVP.

Discussion

The first description of pulmonary arteriovenous malformations (PAVM) or pulmonary arteriovenous fistula was reported by Churton in 1897 [4]. Il these are abnormal vessels that provide direct communication without capillaries between the pulmonary and systemic circulations and therefore an anatomical left right shunt. Dilated and thin-walled structures are a source of hemorrhages. As a result, patients with MAVP may experience hypoxemia, orthodoxy, chest pain, hemoptysis and paradoxical embolizations responsible for strokes or brain abscesses [1,2]. Most MAVP are hereditary, with a rate between 80% and 95% in patients with hereditary hemorrhagic telangiectasia (HHT). HHT, first known as Rendu-Osler disease, affects 1 in 5,000 to 10,000 patients. Vascular homeostasis of capillary vessels is disturbed, allowing the formation of successive and progressive arteriovenous telangiectasias and fistulas. Approximately 50% of patients with HHT have cerebral, pulmonary or hepatic arteriovenous malformations [5]. This will give hemorrhagic manifestations (such as digestive hemorrhages, hemoptysis and especially recurrent epistaxis).the second most common etiology of single MAVP is sporadic as long as the HHT has not been eliminated. Among the rare causes of this malformation are: gestational trophoblastic disease, schistosomiasis, actinomycosis, and sometimes chest trauma. 10% of MAVP appear in children and adolescents, they tend to increase in size with age. Puberty and pregnancy have been described as factors that induce its growth. Spontaneous hemorrhage of PAVM is rare but is the leading cause of feto-maternal death (1%) during pregnancy [5].

Treatment of PAVM by embolization with transcutaneous catheterization in the third trimester of gestation is feasible and effective. Diagnostic tests aim to identify an intrapulmonary right left shunt (RLS). Chest X-ray is normal in 10-40% of patients, including symptomatic. The transthoracic contrast echocardiography (Bubble test) is the most sensitive examination and most of the time makes it possible to differentiate between an intracardiac or intrapulmonary RLS. A stirred saline solution (in order to obtain microbubbles of air) is injected peripherally. Intracardiac RLSs are characterized by the visualization of air microbubbles in the left heart chambers during the first cardiac cycles following their appearance in the right atrium. In patients with intrapulmonary SDG, this event often occurs later, after 3-8 cardiac cycles. The CT Chest Scan is indicated to characterize the precise location of PAVM suspected by contrast echocardiography. In this indication its anatomical resolution is higher than an MRI.

Percutaneous embolization of malformations is the treatment of choice. It is the diameter of the nourishing arteries (> 2-3 mm) that conditions the indication of embolization [3].

Long-term follow-up is important since recanalization and collateralization can occur. Surgical resection is rarely necessary and reserved for patients who are not candidates for embolization. Other aspects to be incorporated into the treatment of MAVP are the recommendation of physical exercise, iron supplementation, the indication of Osler's prevention and the prohibition of scuba diving. Physical exercise is recommended to improve compensatory mechanisms that facilitate oxygenation. Iron treatment is necessary since hell deficiency is associated with increased blood viscosity, an increase in factor VIII and a risk of thrombosis.

An indication of antibiotic prophylaxis is required for procedures, such as dental care that carries a risk of bacteremia and septic embolisms in patients with RLS [3]. Scuba diving is contraindicated given the increased risk of decompression accidents with paradoxical gas embolism. In the follow-up, the current international recommendations propose a CT postembolization chest scan after 6 to 12 months, then every 3 years (to detect possible recanalization).

In the case of our patient who has as antecedent the THH the diagnosis was made following cyanosis and chest pain the angioscanner objectified multiple arteriovenous fistulas of the right upper lobe, with an extension assessment namely cardiac echography and cerebral CT without abnormalities and given the extent of parenchymal lesions and the unavailability of percutaneous embolization in our country a surgical treatment (right upper lobectomy) presented the best indication therapeutic in this child. Early and long-term surgical followup was favorable.

Conclusion

The surgical indication is necessary in front of a PAVM of large size, proximal, and/or extensive parenchymal lesions, presenting a complex nourishing vascular network [1]. This was the case in the observation we describe, which led to a lobectomy.

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