Pulmonary arteriovenous malformation: case report

Malformação arteriovenosa pulmonar: relato de caso

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Abstract

This is a clinical case study involving the respiratory system of a patient in the state of Bahia. The aim of this paper is to report the occurrence of Pulmonary Arteriovenous Malformation (PAVM) of an adult patient from Jequié, Bahia, Brazil. The PAVM is a rare condition, presenting it has twice the incidence in females and is an important differential diagnosis in patients with persistent complaints of dyspnea and hypoxemia. For improving the quality of life of patient with PAVM the few known studies describe invasive procedures like embolotherapy, surgery and pulmonary transplantation to correct the malformation. And so, the present case report aims to contribute to the medical and scientific community through. The description of this rare condition in order to allow a better understanding and monitoring of about PAVM.

Descriptors: Therapeutic embolization; Bronchopulmonary sequestration; Dyspnea

Resumo

Trata-se de um estudo de caso clínico acometendo o sistema respiratório de um paciente no interior do estado da Bahia. O objetivo deste estudo é relatar a ocorrência de Malformações arteriorvenosa Pulmonar (MAVP) de um paciente adulto, do município de Jequié, Bahia, Brasil. A MAVP é uma condição relativamente rara, apresentando uma incidência duas vezes maior no sexo feminino e é um importante diagnóstico diferencial em pacientes com queixas persistentes de dispnéia e hipoxemia. Com vistas à melhora da qualidade de vida de pacientes com MAVP os poucos estudos conhecidos retratam procedimentos invasivos como emboloterapia, cirurgia para correção da malformação e transplante pulmonar. Deste modo, o relato de caso pretende contribuir para o meio médico e científico mediante a descrição desta rara patologia a fim de permitir uma melhor compreensão e acompanhamento a cerca da MAVP.

Descritores: Embolização terapêutica; Sequestro broncopulmonar; Dispnéia

Introduction

Pulmonary Arteriovenous Malformation (PAVM) first described in 1897 by Churton[1]. Arises from the replacement of capillaries by abnormal thin-walled vessels those stand between the arterial and venous circulations, promoting direct communication, free of capillaries between the movement pulmonary and systemic circulation.

The PAVMs have an impact on world 2-3 cases per 100 000 inhabitants[1]. Approximately 10% of cases are identified in childhood, observing a progressive increase in incidence up to 5-6th decades of life[1].

Have cause embryonic, clinical manifestations and severity depends on the degree of lung involvement and location in the thoracic cavity[2].

PAVM is unusual and this was demonstrated in 15.000 consecutive autopsies performed from 1953 in only three cases were detected PAVM[3]. Possibly small arteriovenous communications are no longer detected in the routine of study greatly hinders the finding of low prevalence in this study, thus confirming the difficulty of diagnosing this rare condition[4].

Report of 194 cases of PAVM, in 45 years, indicated an annual incidence of approximately 4,3 cases[4]. PAVM a proves to be twice as prevalent in females[5].

Morphologically the PAVM still presents diverse distribution, size and amount of malformations. A study of 350 patients was demonstrated that 36% of patients had multiple lesions and 25% with bilateral distribution. More than half of the lesions are located in the lower portions of the lungs and over 80% involved the pleura[6].

It highlights the importance of this study, since it allows the monitoring of a patient with a rare disease in the reality of medical practice, allowing you to reach greater understanding of the course of the disease underdiagnosed by health professionals.

The objective was to report a clinical case of PAVM in adult patient in Jequié – Bahia, as well as analyzing their changing clinical picture.

The information contained in this study were obtained through review of medical records, interviews with patients and recording of diagnostic imaging methods for which the patient underwent with basis on literature reviews.

This study meets the requirements of Resolution 466/12, when the human study, the National Health Council – CNS on CAAE 27972614.7.0000.0055 protocol.

Case report

Patient, male, 49 years old, assistant mason, catholic, natural Jequié-Bahia, reported dyspnea on moderate exertion longtime medical appointments.

Reports ICU stay of General Hospital Prado Valadares (HGPV), diagnosed with respiratory failure, sepsis, and decreased level of consciousness. Refers long hospitalization in ICU with difficult weaning due significant hypoxemia, unresponsive to oxygen therapy and clubbing, suggesting an ancient hypoxemia. The patient recovered without echocardiography.

Performed computed tomography (CT) of the chest with a diagnosis of PAVM in the right lung base. In the same period was diagnosed ischemic stroke, evolved with cognitive deficit and hemiparesis on the left metameres. Remained hypoxic and cyanotic after hospital discharge, but without medical supervision, only sought reevaluation pulmonologist, after consultation with an endocrinologist who referred him. PAVM confirmed in the right lung on chest CT (Figure A and B). Maintain patient dyspnea on ordinary exertion, O_2 saturation at rest at around 77%, sequel to the left hemiparesis in metameres overall poor health, diabetes mellitus decompensated, central cyanosis and clubbing due to chronic hypoxemia; denied, however, new event of stroke.

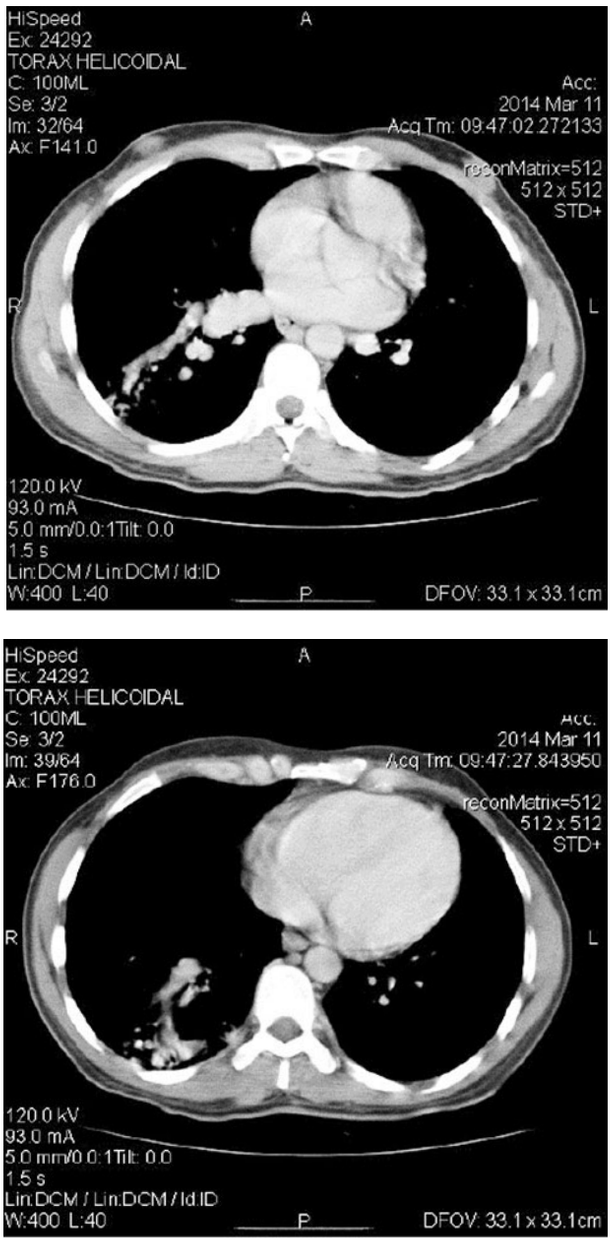

Figure 1. Hyperdense images (A and B) in a pulmonary parenchyma region, evidencing tortuous dilatations in the lower right lobe, consisting of arterial and venous pulmonary branches, ranging from the periphery to the pulmonary wire

Discussion

Most deaths caused by PAVM are due to complications such as stroke, brain abscess, hemoptysis and hemothorax. Although the mortality rate is uncertain by PAVM brief accompaniments that the reported cases show, it is possible to infer the morbidity significantly from known studies, due to the earliness with which clinical manifestations appear[4].

In our case, the patient has dyspnea longtime, which by intrinsic compensatory mechanisms, not associated with exposure to risk factors (smoking) allowed a quality of life without major repercussions of the respiratory system, since presented stroke at 49 years old.

In view of the possible diagnoses for reporting presented cases as PAVM pass unnoticed because of a higher prevalence of other disorders of cardiopulmonary such as asthma, tuberculosis, COPD, cystic fibrosis, among others.

A fairly common complication is attributed to PAVM stroke from embolic material that surpasses the ability to filter normally exercised by the pulmonary capillaries[7]. Accordingly, the piston tends to reach the central nervous system, which explains the high incidence of stroke in PAVM patients.

However, not all patients require immediate intervention. The decision to intervene should be based on the risks of serious complications, such as stroke or brain abscess, taking into consideration inter alia, possible complications of embolization procedures as transcutaneous catheter[8].

The indications for treatment have not been satisfactorily resolved by the lack of studies that evaluate the most appropriate interventions. The interventions[6] are from few studies and geared mainly on classical presentations[8].

- Patients who have one or more PAVMs with a diameter of blood flow (DVA) between 2 and 3mm in chest CT, regardless of symptoms, should undergo pulmonary angiography. At the time of pulmonary angiography or greater DVA 3mm 8 embolization is indicated.
- Symptomatic patients have indication whether embolization of the DVA. Symptoms may be due to hypoxemia, stroke and cerebral abscess[8].
- Asymptomatic patients with less than 2mm DVA must be accompanied with non-contrast CT, usually every three or five years[8].

Regarding therapy, studies known about the PAVM portray embolotherapy invasive procedures such as surgery to correct the malformation and pulmonary transplantation.

Therapeutic embolization occurs by occlusion of blood flow through insertion of embolic materials, such as metal alloys or disposable balloons. The angiographic diagnosis of additional malformations is usually performed in the same procedure embolotherapy, and recommended that multiple malformations are embolized in a single session[9].

In a study[10] that assessed short-term outcomes, the embolotherapy showed promise in 39 patients for 43

months, 80% reported improvement in the complaints of dyspnea and stabilization of the partial pressure of oxygen (PaO_2). Thus, the great advantage of embolotherapy would improve quality of life and the reduction of serious complications, such as strokes and brain abscesses.

Surgical intervention is configured as a treatment option, not always the procedure of choice and is reserved for cases of allergy to contrast or rare cases refractory to PAVM s embolotherapy. Surgical techniques include link PAVM aired for vascular malformation excision, lobectomy and pneumonectomy. In surgeries to treat PAVM risks are similar to morbidity and mortality from other thoracic surgical procedures[8].

Lung transplantation is considered the last resort in the treatment and is indicated in embolotherapy or failure of surgery, especially in patients with bilateral malformations[8].

Prevention of complications involves the care in the administration of intravenous medications because of embolic risk; one should prevent the passage of bubbles, preferring the use of filters for infusion of drugs. The underwater diving apparatus must be avoided at greater risk of emboli formation. In air travel, curiously, complications are not common[11].

Considering that the patient had a stroke and severe hypoxemia, the most recommended would be the intervention embolotherapy as the aforementioned classic criteria.

However, in deference to the principle of autonomy widespread in more informed health laws, the embolotherapy cannot be adopted in patient as contrary to his will. Thus, we adopted a conservative approach of monitoring, greater losses to be avoiding their quality of life.

Conclusion

The case report and publications analyzed demonstrate that although PAVM is a rare and underdiagnosed condition, the ability of medical professionals to realize its manifestations, distinguishing it from other possible diagnoses most prevalent allows interfere significantly on morbidity and quality of life the patient. In the present case report behavioral measures were adopted, where lack specific pharmacological measures for the case, the medical staff may have guidelines like avoiding smoking, smoke exposure, avoidance of strenuous efforts being valid and the multidisciplinary approach, aiming to improve hypoxemia.

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