Pulmonary Varix Inside a Bulla

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Introduction

Pulmonary varices are uncommon vascular abnormalities that are usually asymptomatic and so they are normally diagnosed by chance from a chest x-ray. They often present as a pulmonary nodule and can be either congenital or acquired. If acquired, they are associated with pulmonary venous hypertension, usually as a result of mitral valve disease. Pulmonary arteriography provides a definitive diagnosis, although the use of new noninvasive imaging techniques is spreading. Treatment is not normally required unless serious complications arise. We present the case of a pulmonary varix located within a pulmonary bulla. This form of presentation has not been previously reported.

Key words: Pulmonary varix. Pulmonary nodule. Hemoptysis. Pulmonary bulla

Case Description

A 69-year-old woman was referred for study of a pulmonary nodule. She reported no addictions and the only information of interest in her medical history was diagnosis of type 2 diabetes mellitus after admission to hospital for hyperglycemic coma. Four months before admission, she had suffered an episode of self-limiting hemoptysis. A chest x-ray revealed a pulmonary nodule 2 cm in diameter with smooth and well-defined edges in the right upper lobe. The nodule had been observed in an x-ray taken 6 months earlier, but it was smaller. No noteworthy findings were reported in the physical examination. The laboratory analysis and electrocardiogram done on admission showed no abnormalities. Fiberoptic bronchoscopy revealed no macroscopic abnormalities. No abnormal findings were documented in the microbiological and cytological tests of the bronchial aspirate. In a new x-ray examination 1 month later, no pulmonary nodule was observed, although computed tomography examination of the chest revealed a nodular structure with a maximum diameter of 1.5 cm in the right upper lobe. The extent of contrast uptake by this structure was significant (similar to that of the pulmonary artery and vein), leading to the suspicion that the lesion was of vascular origin. The lesion was located within a bulla measuring 2 cm in diameter (Figure 1). Lung function tests gave no significantly abnormal values: forced vital capacity (FVC) was 2.06 L (85% of predicted), forced expiratory volume in 1 second (FEV₁) was 1.94 L (109% of predicted), FEV₁/FVC was 94%, and overall lung capacity was 4.30 L (93% of predicted). PaO₂ was 87 mm Hg, PaCO₂ was 38 mm Hg, and pH was 7.41 in arterial gas measurements taken while breathing air. In arterial gasometry while breathing pure oxygen, pH was 7.44, PaO₂ was 601 mm Hg, and PaCO₂ was 35 mm Hg. The pulmonary shunt fraction
(Qs/Qt) calculated from these values was 4%. Pulmonary arteriography revealed no abnormalities either in the pulmonary arteries or their branches, or in the arteriolar circulation. However, the venous phase of the study revealed saccular dilation of a venous branch of the apical segment of the right upper lobe (Figure 2). No shunting with systemic circulation was seen in the thoracic aortography; therefore mixed irrigation of the dilation was ruled out. Selective bronchial arteriography also failed to show significant abnormalities. With echocardiography, any type of valve disease or any other type of heart disease was ruled out. Given the lack of effect of this arteriovenous malformation on clinical and hemodynamic variables and on blood gases, it was decided not to refer the patient for surgery and she was discharged. No subsequent complications were reported.

Discussion

Pulmonary varix is an uncommon arteriovenous malformation consisting of abnormal dilation of a segment of the pulmonary vein. Abnormalities are classed into 3 groups according to structure: saccular (localized, ovular form), tortuous (extensive and irregular), or confluent (localized at the confluence of the pulmonary veins). The latter two types of varix, and in particular the confluent type, have been associated with pulmonary venous hypertension. In a review by Uyama et al., it was found that 62% of varices associated with valve disease were of the confluent type and 19% of the tortuous type. No cases of saccular varices associated with pulmonary venous hypertension were reported, and so local factors were thought to be important in the origin of this type of vascular abnormality. This case report is consistent with those findings, given that the patient’s varix was saccular and there was no associated valve disease. The fact that the lesion was located within a bulla suggests that the bulla itself contributed to the formation of the varix as a local factor. A varix in the wall of a bulla has not been reported previously.

Pulmonary varices are usually asymptomatic and so are only detected by chance in chest x-rays. The most common site is the right lower lobe. In rare instances, as in our patient, the varix can be associated with hemoptysis. Complications such as varix thrombosis and secondary systemic embolization and varix rupture in the pleural or bronchial cavities have been reported in isolated cases. Surgical complications associated with unnecessary diagnostic thoracotomies have also been reported, though these are now rarer because of advances in imaging techniques. Other rare manifestations of the condition have been described, such as dysphagia or the presence of middle lobe syndrome secondary to extrinsic compression.

The size of the varix measured in a chest x-ray will often vary because of Valsalva or Mueller maneuvers. This explains the variation in diameter and even the disappearance of the lesion as in this case report.

Diagnosis of pulmonary varix is established from the results of pulmonary angiography. The following criteria must be met: a) normal arterial phase with no abnormalities or capillary shunting; b) presence of venous dilation filling with contrast at the same time as the other veins; c) direct flow from the varices into the left atrium; d) drainage of the varix occurring later than drainage of normal pulmonary veins; and e) in the case of tortuous pulmonary veins, involvement principally of the proximal part of the vein. In the last decade, new techniques have been introduced that allow diagnosis of these abnormalities. Echocardiography is useful for studying varices close to the heart and the technique has the advantage of allowing assessment of valve disease in the same diagnostic procedure. Normally, transesophageal echocardiography is used, but diagnosis has also been made with transthoracic echocardiography. Likewise, nuclear magnetic resonance and helical computed tomography have been employed.

In most cases, treatment is not indicated and surgical resection of the varix is reserved for patients with serious
complications. Radiological monitoring is essential, although the lesion does not normally progress over time. When the lesion is associated with valve disease, valve surgery may reduce the size of the varices.

In conclusion, faced with the presence of a pulmonary nodule whose size changes over time, pulmonary arteriovenous malformation should be suspected, and on rare occasions, a pulmonary varix may be present. Diagnosis should make use of advanced imaging techniques. If doubts about diagnosis of the vascular origin of the disease remain, the technique of choice for diagnosis is pulmonary arteriography, which can distinguish arteriovenous malformation from pulmonary varix and other abnormalities. If diagnosis of pulmonary varix is confirmed, it is advisable to wait and only consider surgery in patients at risk of serious complications.

REFERENCES