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Iatrogenic exacerbation of pulmonary arteriovenous malformation in a patient with benign metastasizing leiomyoma

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Abstract

We present a case of a middle-aged woman with known benign metastasizing leiomyoma presenting with pleural effusion. After ultrasound-guided drainage of the largest cyst was performed, the patient became hypoxemic. Chest computerized tomography (CT) showed a large tortuous vessel adjacent to the biggest cyst that had been drained. A 10-fold increase in the diameter of this vessel was noted when compared to CT scan performed 24 h before the procedure. A 20% right-to-left shunt was observed on nuclear medicine shunt study. To our knowledge, this is the first reported case of metastasizing leiomyoma with coexistent pulmonary arteriovenous malformation.

Introduction

Uterine fibroids are the most common uterine tumor. Rarely, the fibroids can be associated with extra-uterine variant, such as benign metastasizing leiomyoma (BML) [1]. Typically, BML are noted years later in women with uterine fibroids. BML can be found in various distant locations, including lymph nodes, deep soft tissues, mesentery, bones, central nervous system, heart; however, the lungs are the most common sites. The dissemination route remains unknown and could be via a venous passage to the lung, lymphatic or iatrogenic [2]. The diagnosis is confirmed by the presence of histological findings consistent with leiomyoma. BML are mainly seen in reproductive-aged women, and are usually associated with an indolent course of slow-growing metastatic lesions and a regression of the metastases after menopause or anti-estrogen therapy [2].

Case Description

A 57-year-old woman was presented to the emergency room with a 1-week history of increasing shortness of breath. Her past medical history included a total abdominal hysterectomy performed 17 years ago after her second pregnancy due to a large uterine leiomyoma. Fifteen years ago she was diagnosed with BML, based on typical findings on the chest X-ray and lung biopsy. She underwent an oophorectomy and was treated with Megestrol, Femara, and Evista. Except for two episodes of pleural effusion requiring pleurocentesis respectively at the time of diagnosis and 2 months prior to presentation, the patient remained asymptomatic.

On arrival, the patient's vital signs were within the normal range and she required 2 L of oxygen to maintain saturation around 94%. A chest X-ray revealed a significant left-sided pleural effusion and her baseline bilateral multiple cystic lesions. An echocardiogram showed normal ejection

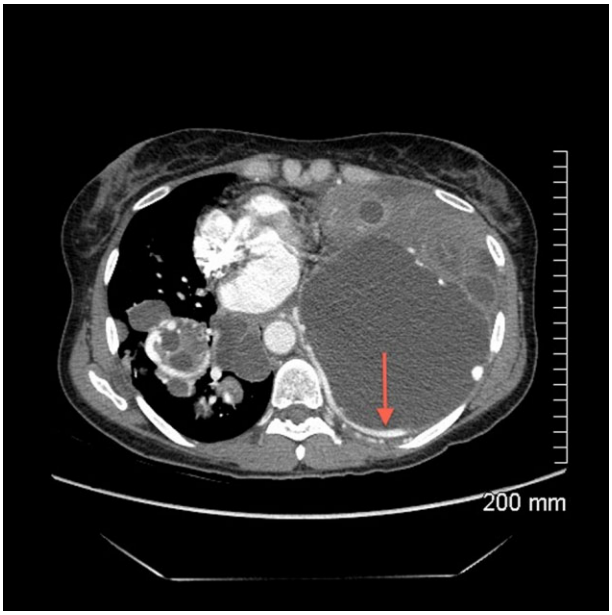

Figure 1. Computed tomography image showing the largest cyst 13 × 11.5 cm. Red arrow pointing at the 1–2 mm pulmonary arteriovenous malformation before the cyst was drained.

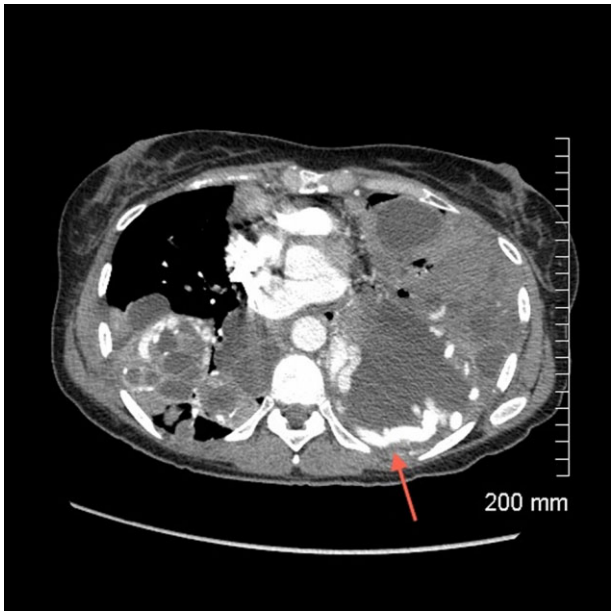

Figure 2. Computed tomography image showing expansion of pulmonary arteriovenous malformation after the drainage of the cyst. The pulmonary arteriovenous malformation now measures 11 to 12 mm in maximal diameter.

fraction and a pulmonary computerized tomography (CT)-angiography was negative for pulmonary embolism.

With the goal of alleviating her shortness of breath, the largest cystic lesion measuring 11.0 × 13.5 cm was targeted for ultrasound-guided drainage; 700 mL were removed and a pigtail catheter was left in place. During the procedure, the patient's oxygen requirements increased and she became hypotensive (84/55 mmHg). The procedure was stopped. A few hours later, the patient's saturation was 84% on a non-rebreather mask, her blood pressure was 90/60 mmHg, and she was tachycardic at 115 beats/min, with a PaO_2 of 57 mmHg. The drainage tube had collected 1800 mL in less than 24 h. A chest X-ray was negative for re-expansion pulmonary edema and pneumothorax. An urgent CT-angiography showed a prominent vessel surrounding the drained cystic lesion measuring 11 to 12 mm in maximal diameter, consistent with pulmonary arteriovenous malformation (PAVM). Comparison with previous CT revealed marked dilatation of this vessel, which only measured approximately 1–2 mm prior to drainage (Figs. 1, 2).

A 20% right-to-left shunt in the context of a pulmonary vein-to-artery fistula was revealed by 99mTc-MAA scintigraphy shunt study. The patient was transferred to interventional radiology where a pulmonary angiogram demonstrated the large left PAVM, as well as some smaller fistulas bilaterally. Embolizations of her left-sided PAVMs were performed resulting in an immediate return to her saturation baseline of 92% on room air (Fig. 3). A month later, she presented with shortness of breath and her saturation had declined to 86% on room air, prompting embolization of her right-sided PAVMs. A final pulmonary angiogram was performed showing complete resolution of the PAVMs.

Discussion

BML have been described as benign due to the low mitotic rate and other benign histological features. To date, approximately 150 BML cases have been described, most of them reporting lung involvement and slow-growing metastases. Most evidence supports the role of estrogen and progesterone receptors in the pathophysiology of BML. As BML is a rare entity, most information is case-report based. Estrogen and progesterone suppression (bilateral adnexectomy, GnRH analogues and progesterone antagonists) are the proposed treatment modalities.

Despite the presence of benign nodules, their localization or their size can become problematic. In this case, a large cyst was compressing a large arteriovenous fistula and a right-to-left shunt occurred once the cyst was drained. Prior to drainage of the cyst, the cyst itself was causing a tamponade effect by compressing the pre-existent PAVM. The drainage of the cyst therefore relieved the pressure on the vessel and resulted in the expansion of the PAVM. Further revision of the CT-angiography revealed an AV fistula, less extensive, also in the right lung. To our knowledge, this is the first case

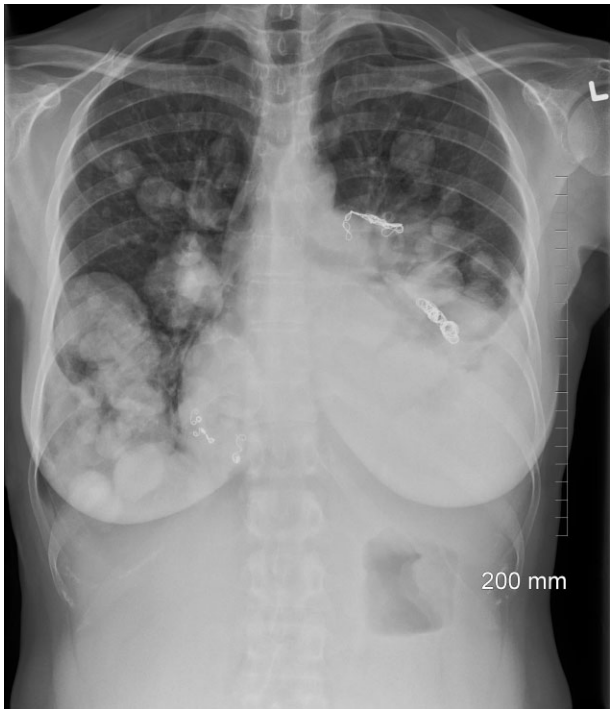

Figure 3. Chest X-ray after embolization.

reporting PAVMs in BML, and it remains unclear if this is related to the disease, or the result of a chronic process in the lung. Seventy percent of PAVM are congenital and associated with hereditary hemorrhagic telangiectasias [3]. Our patient had no other indication of this disease and her AVM was not seen on multiple previous imaging modalities. The most common cause of acquired PAVM is hepatopulmonary syndrome in the context of portal hypertension and has also been described in mitral stenosis, metastatic carcinoma or post-Fontane procedure [3]. Interestingly, it has also been very rarely associated with localized chronic chest infection, such as tuberculosis [4], schistosomiasis, and actinomycosis [5]. The hypothesis of neovascularization secondary to the chronic cyst seen in BML has been described in chronic chest infections. Additional cases are needed to support this hypothesis.

Currently, three treatment plans exist for PAVMs: percutaneous arterial embolization, open surgery, and lung transplant; the latter two are less preferable for the obvious reasons of invasiveness and availability. Embolization is preferred for almost all patients because of its high success rate, minimal invasiveness, and minimal loss of pulmonary parenchyma. Embolization has very high success rates, with case series reporting over 98% success rate regarding short-term outcomes [6]. Another case series in patients with bilateral PAVMs who were followed for an average of 43 months showed that embolization was completely or partially successful in 97% of patients, with improvement in dyspnea, NYHA function class and arterial oxygen tension (PaO_2) [7].

This case also suggests that interventional radiologists give careful consideration prior to performing image-guided drainages of chronic lung lesions when they are surrounded by slowly forming vascular structures. In the case at hand, the possibility of PAVMs had been entertained early in the patient's clinical history but had not been proven. The initial response had been to drain a very slowly growing benign cystic lesion in the lung which was thought to be responsible for the patient's subacute respiratory condition. The result, however, was a worsening in clinical respiratory symptoms. Subsequently, it was found that the vascular structures, later proven to be PAVMs, surrounding these chronic long-standing cystic lesions had had a "protective" tamponade effect. Whenever, then, long-standing disease processes in the lungs are encountered accompanied by surrounding neovascularization, interventional radiologists should consider the possibility of acquired PAVMs.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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