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Case Report

A case report of primary pulmonary artery intimal sarcoma [☆]

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ABSTRACT

Primary pulmonary artery sarcoma is a rare tumor that mimics pulmonary embolism. Patients may present with cough, dyspnea, chest pain, and weight loss. The diagnosis is challenging. Herein, we report a case of 29-year-old female patient who had presented with dyspnea, fatigue for 2 weeks. Computed tomography pulmonary angiography scan suggests pulmonary embolism. We decided to perform surgical embolectomy. The histopathological results, however demonstrated primary pulmonary artery intimal sarcoma. The patient died 1-month post-surgery because of respiratory and circulatory failure.

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Introduction

Primary pulmonary artery sarcoma (PAS) is an extremely rare malignant neoplasm that was first reported in 1923 by Mandelstamm [1]. The exact incidence of PAS is difficult to determine because of its rarity and imitation with pulmonary embolism. The clinical symptoms, laboratory findings, and imaging features are non-specific which makes diagnosis difficult [2]. To date, there is no standard treatment for this disease. Surgery, radiation, chemotherapy, and targeted ther-

apy have all been applied so far [3]. This study emphasizes the clinical symptoms and survival outcomes of this disease.

Case report

A 29-year-old female patient was referred to our hospital for dyspnea, fatigue, and inability to perform daily activities. She did not have fever, cough, edema or focal neurologic signs. The symptoms had lasted for 2 weeks and then exacerbated

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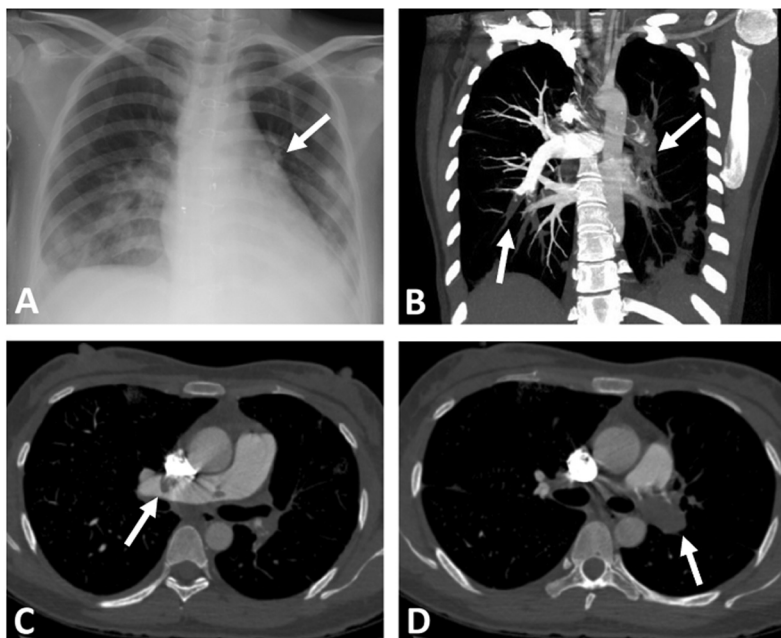


Fig. 1 – Chest X-ray showed enlarged pulmonary arteries (A, arrow) and bilateral opacity in the mild and lower lung zones. CTPA scan showed intraluminal hypoattenuating lesions arising from the right and left pulmonary arteries and extending into their branches (A, B and C, arrows). The lesions were poor enhancing and caused partial occlusion of the right PA and complete occlusion of the left PA and extended to distal both PA branches.

quickly. The patient also had felt chest tightness 3 months before, without any history of fever, anorexia, weight loss, dyspnea. She had no significant medical history or trauma, and her family history did not have any specific medical illness.

At the time of admission, the patient was conscious, and exhausted. Physical examination was notable for the symptoms of severe respiratory failure: respiration rate 28 breaths per minute, oxygen saturation of 85% in ambient air. Her other vital signs were as follows: Body temperature of 37°C, pulse rate of 110 beats per minute, and blood pressure of 120/70 mm Hg. Lung auscultation found both lungs ventilation was equal, there were bibasilar crackles without rhonchi sounds. Auscultation of the heart showed tachycardia, systolic sound over lower left sternal border. The patient was treated with face mask oxygen therapy and the oxygen saturation was raised to 94%. Neither symptoms of peripheral vein thrombosis nor hypercoagulable disorders were noticed.

Chest X-ray showed enlarged pulmonary arteries and bilateral opacity in the mild and lower lung zones. Electrocardiogram recognized sinus tachycardia. Echocardiography showed severe pulmonary arterial hypertension (85 mm Hg), moderate tricuspid valve regurgitation due to right ventricular dilatation. There was no sign of peripheral venous thrombosis on vascular ultrasound. The C-creative protein and procalcitonin levels were within the normal range. D-dimer levels were 650 ng/mL. Computed tomography pulmonary angiography (CTPA) scan revealed complete occlusion of the left pulmonary artery (PA), partial occlusion of the right PA, extended to distal PA branches (Fig. 1). There were multiple ground-glass opacity areas in both lungs. The CTPA findings suggested pulmonary artery embolism. Abdomi-

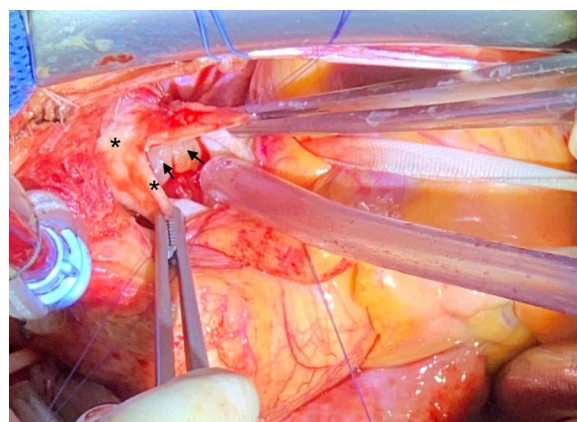


Fig. 2 – Main pulmonary artery (asterisk) was filled by myxomatous soft tissue (arrows).

nal computed tomography scan showed no abnormalities. This patient was indicated emergency surgical pulmonary embolectomy. During this procedure, we made an incision through the pulmonary trunk to the right pulmonary artery, and peripheral branches of the left pulmonary artery. After opening the artery, we detected lesions arising from the wall of pulmonary artery walls, occluding completely the left pulmonary artery, and partially the right one. Due to the lesion involving both pulmonary arteries, we could not completely remove them. Therefore, we decided to biopsy the tumor. The samples were white and friable suggesting a PA tumor than embolism (Fig. 2). On microscopic examination,

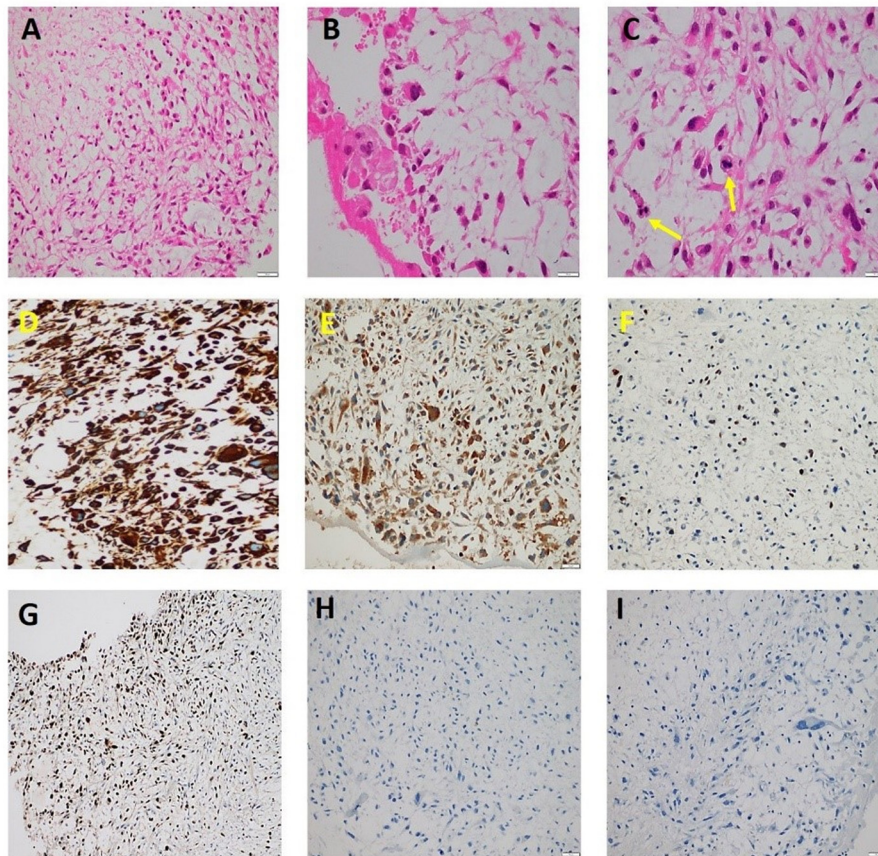


Fig 3 – Microscopic and immunohistochemical findings. (A) The necrotic area presented in the tumor (hematoxylin-eosin, x 200). The tumor consisted of large spindle and epithelioid cells lying discretely on a myxoid stroma (hematoxylin-eosin, x 400) with mitotic figures (C, yellow arrows). Immunohistochemical staining of the tumor cells showed strong positivity for Vimentin (D, x 200) and CD68 (E, x 200). Tumor cells also showed patchy positivity for MDM2 (F, x 200) and high Ki-67 proliferative index (G, x 100). The tumor cells were negative for CD31 and SMA (H and I, x 200) (Color version of the figure is available online.)

the tumor consisted of large spindle and epithelioid cells lying discretely on a myxoid stroma with several necrotic foci. The tumor cells had hyperchromatic nuclei, prominent nucleoli and abundant eosinophilic cytoplasm. The number of mitotic figures was high. Immunohistochemical staining showed strong and diffuse positivity of vimentin, CD68, focal expression of MDM2, and high Ki67 index. The tumor cells were negative for CD31, smooth muscle actin (SMA), S100, CD34, Desmin, MUC4, and CK. The histopathological results confirmed pulmonary artery intimal sarcoma (Fig. 3). The final diagnosis was primary pulmonary artery intimal sarcoma.

Post-surgery, intravenous heparin was used to reduce the incidence of thrombosis. Milrinone and dobutamine were also prescribed. Although the respiratory symptoms in the early days improved, 1 week later, they became worse. One month later, the patient died due to respiratory and circulatory failure.

Discussion

PAS can be classified into 2 types: Intimal sarcoma and luminal sarcoma, however, the luminal type is rarely seen, and

PAS mainly refers to intimal sarcoma [4]. Pulmonary artery intimal sarcoma is an extremely rare malignant neoplasm that originates from the intimal layer of the pulmonary arteries [5]. The etiology remains unknown. The age at diagnosis ranges from 24 to 74 years and the mean age is 55 [6]. There are no differences in prevalence and outcomes between males and females [6]. The pulmonary arteries and/or pulmonary trunk are the most common sites of PAS (60%), and 30% of patients have unilateral manifestation [7].

The clinical symptoms of PAS are variable and non-specific. They are often similar to PA embolism, leading to misdiagnosis. Most of the patients present with symptoms of acute or chronic pulmonary hypertension such as dyspnea, chest pain [8]. Other symptoms include syncope, cough, hemoptysis, palpitations, hoarseness, fever, night sweats, anorexia, weight loss [8,9,10]. However, some patients may be asymptomatic [6]. The average duration of symptoms is 10 months (range 2-36 months) [8].

The clinical laboratory findings of PAS patients are non-specific. Some patients have low total serum protein or elevated erythrocyte sedimentation rate, serum level of lactate dehydrogenase [9]. The D-dimer level is usually within normal ranges; however, elevated D-dimer values have been described in some cases [6,11].

Echocardiogram, computed tomography pulmonary angiography and magnetic resonance imaging (MRI) are often used for the diagnosis of PAS. Other imaging studies include angiography and positron emission tomography (PET)-CT. On echocardiogram, we can see right-ventricular enlargement, tricuspid regurgitation, and pulmonary hypertension [9]. PA thrombosis, pulmonary stenosis, PA partial or complete obstruction or the mass attachment to the pulmonary valve or PA wall may be seen on echocardiography. Some features of PAS on CTPA include intraluminal defects in the pulmonary trunk, right or left PA, and may extend to the segmental arteries or heart [9]. Additionally, the tumor appears as rounded, bulged or lobulated surfaces, wall eclipse sign, low-density, heterogeneous attenuation from hemorrhage, enhancement, increased diameter of the PA, lung ischemia [5,9]. Compared to PAS, PA thromboembolism is often homogeneous without enhancement. Some findings such as heterogeneous attenuation, wall eclipse sign, lobulated surfaces, surface nodularity, and central location were significantly more common in PAS than in PA thromboembolism [9]. On MRI, PAS tumors are restricted on diffusion, and heterogeneous enhancement [5]. PAS patients show hypermetabolic FDG uptake on PET-CT with the median max SUV of PAS was 7.15 (range, 3.1-17.6), whereas these findings are not present in PA thromboembolism [5].

Pulmonary artery intimal sarcoma originates from the inner lining of the pulmonary arteries. Histologically, Pulmonary artery intimal sarcoma consists of spindle-shaped, or epithelioid cells and often looks like undifferentiated pleomorphic sarcoma (UPS), myxofibrosarcoma or epithelioid angiosarcoma [12]. However, immunohistochemistry showed that MDM2, vimentin are positive, the proliferation index of Ki67 is high [12]. The imaging features may suggest pulmonary artery intimal sarcoma, but a definitive diagnosis depends on the histopathology.

Up to the present, there is no standard treatment available due to its low incidence [11,13]. The management often includes multidisciplinary approaches such as surgery, chemotherapy, radiation [13,14]. Surgery is considered the mainstay of management of PAS. Some studies demonstrate chemotherapy may reduce the risk of metastatic, chemotherapy, and radiotherapy may improve survival rate compared to surgery alone [10].

PAS is an aggressive malignant tumor. Even with prompt diagnosis and early treatment, the prognosis remains poor. Approximately 50% of PAS patients have lung and mediastinal metastases, 16%-19% of PAS patients develop distant metastases at the time of diagnosis, and 60% of patients have recurrence [7,9]. The overall survival time is approximately 1.5 months without treatment [7]. Some studies with small samples reveal the median overall survival time is variable, ranging from 14.6 to 37 months after treatment [11].

In our case, due to the life-threatening pulmonary artery occlusion, emergency reperfusion was necessary. In the acute phase of high-risk or in selected cases of intermediate-risk PE, systemic thrombolysis is the first choice when hemodynamic deterioration on anticoagulation treatment. The greatest benefit of this therapy is observed when treatment is initiated within 48h of symptom onset, and thrombolysis can still be useful in patients who have had symptoms for 6-14 days [15]. In this patient, the symptoms had lasted for 3 weeks. So we

decided to surgical embolectomy. However, we cannot completely remove the tumor. Unfortunately, the patient died one month later.

Conclusion

Pulmonary artery sarcoma is extremely rare and the clinical symptoms and radiological features often mimics pulmonary embolism. In young female patients who presented with pulmonary occlusion without past diseases or hypercoagulable disorders with normal coagulation test, pulmonary artery sarcoma should be considered in the differential diagnosis.

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Author contributions

The authors confirm contribution to the paper as follows: study conception and design (Nguyen Lan Hieu, Le Hoan), data collection (Hoang Bui Hai, Vu Ngoc Tu), draft manuscript preparation (Thieu Thi Tra My, Tran Ngoc Minh, Nguyen Ngoc Cuong, Pham Thuan Manh). All authors reviewed the results and approved the final version of the manuscript

Name of department and institution in which the work was done

Cardiovascular center, Hanoi medical university hospital.

Patient consent

Patient's supervisor was informed that her documents (without personal information) including diagnosis and treatment information might be published for science purpose. She agreed and signed in the consent form.

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