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# A case presented with fever enlightened by cardiac auscultation: Sarcoma originated in pulmonary artery

**Yavuz Fatih Yavuz*, Nazmi Toprak, Cemil Kavalci, Fevzi Yilmaz**

*Department of Emergency Medicine, University of Health Sciences, Antalya Education and Research Hospital, Antalya, Turkey*
**Corresponding author*

**Abstract:**

In the emergency department, there are many symptoms patients present. One of the major symptoms is fever which could be the only symptom, as our patient had. Not only do infections, drugs, trauma, etc., cause fever, but also undetermined cancer types do. In this case, we are presenting a 28-year-old male coming with a 3-week duration of fever and being admitted with the diagnosis of pulmonary artery intimal sarcoma as generally misconceived with pulmonary thromboembolism, to raise awareness of this fatal cancer.

**Keywords:**

Fever, pulmonary thromboembolism, sarcoma

## Introduction

Stromal sarcoma is a rare type of malignancy that might occur in the main arteries and their branches. The incidence was found about 0.001%–0.003%. Historically, the first presentation of intimal sarcoma to the literature was on autopsy by Mandelstamm. This pathology might be seen between 2 months and 89 years with the average age being about 45–54 years.[1]

The symptoms of sarcoma manifest great diversity but presenting with only fever is rare.[2] A comprehensive physical examination should be performed for this insidious presentation. Cardiac auscultation is an excellent bedside diagnostic tool a clinician can use to detect cardiovascular anatomy and physiological variations.

In this case, we are analyzing a process that started from fever and a suspected cardiopulmonary disease to a definitive diagnosis of primary pulmonary artery sarcoma (PPAS) to contribute to physicians' perceptions when approaching fever of unknown origin.

## Case Report

A 28-year-old previously healthy male applied to our emergency department with a 3-week duration of fever. There were not any other symptoms. He was kickboxing regularly. There were not any known medical conditions or allergies. No illicit drug use or any kind of smoking was claimed.

In history, the patient reported that he had been having occasional coughs with a fever over the last 2 months. The patient went to another emergency department for these complaints. After the follow-up, he was

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**ORCID:**
*YFY: 0000-0002-9234-6222*
*NT: 0000-0002-6210-1478*
*CK: 0000-0003-2529-2946*
*FY: 0000-0002-3675-7457*

**Address for correspondence:**
Dr. Yavuz Fatih Yavuz, 3807th Street, Meltem District, Muratpaşa, Antalya, Turkey.
E-mail: yavuzfatihyavuz@gmail.com

directed to infection's disease outpatient clinic. Blood and urine cultures were negative, and he was sent to the neurology outpatient clinic. The patient presented with these symptoms to different health-care facilities for a total of 5 times, but no definitive diagnosis was made.

In admission to our emergency department, he was alert, talkative, and breathing spontaneously and his Glasgow Coma Scale was 15. His body temperature was 39.4°C, blood pressure was 154/97 mmHg, pulse rate was 93, and breathing rate was 25/min with $SpO_2$: 97%. His detailed physical examination was otherwise normal except a 4/6 crescendo–decrescendo holosystolic murmur was heard on the pulmonary valve area augmenting with deep inspiration. His electrocardiogram was in sinus rhythm, and his pulse rate was 105 bpm. A transthoracic echo was ordered due to fever and cardiac murmur and detected a moderate degree of tricuspid insufficiency, 65 ± 10 mmHg pulmonary artery pressure, and a 31 mm × 25 mm mobile thrombus or a solid lesion. His ejection fraction was 65%. A computed tomography angiography (CTA) scan was ordered to detect pathology related to the pulmonary system. A CTA scan was analyzed as filling defects compatible with a massive pulmonary embolism in the bifurcation of the pulmonary artery [Figure 1]. The patient was admitted to our intensive care unit, and then transported to the pulmonary department for admission. Thrombolytic therapy was advised by the pulmonary department and commenced but stopped immediately after 5 min due to hypotension. The only abnormal blood results were white blood cells 19,000/$mm^3$, dominated by neutrophils; C-reactive protein: 106 mg/L. The blood and urine cultures were obtained and negative.

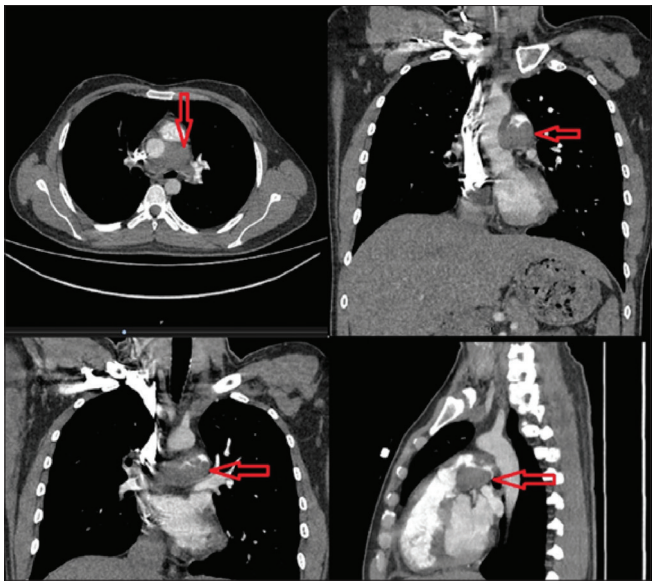

**Figure 1:** A computed tomography angiography scan with different axes displayed a massive pulmonary thromboembolism; note that red arrow shows nearly complete obliteration of the main pulmonary artery

A magnetic resonance angiography scan was performed by the pulmonology department interpreted chronic massive pulmonary embolism. After inconclusive therapies, a positron emission tomography–computed tomography (PET-CT) scan was ordered 1 month later and displayed pathological uptake of this area [Figure 2a and b]. He underwent a pulmonary endarterectomy operation with bypass grafting. The extracted mass was identified as pulmonary artery intimal sarcoma by the pathology department. The patient's PET-CT scan after a 7-month follow-up with chemotherapies detected no pathological F-fluorodeoxyglucose uptake.

Our patient has been informed of the case above and confidentiality and signed an informed consent form voluntarily.

## Discussion

PPASs are rare types of malignancies that clinicians might be confused with due to their presentation as Pulmonary Embolism (PE). The disease incidence was detected as 0.001%–0.003% and up to 2021, Mandelstamm first reported it in 1923 on an autopsy, and only ~400 cases were reported since.[3-5] Making the differential diagnosis of the PPAS from PE was crucial as an extensive observational project showed that 47% of PPAS cases were diagnosed as PE.[6] In a study in 2017, Srivali *et al*. detected the main symptoms of the disease as dyspnea, cough, syncope/presyncope, and hemoptysis in a case series of nine patients who had PPAS. After the first CT scans with contrast, anti-coagulation therapy was initiated for seven patients.[2] Insufficiency of specificity of imaging techniques to this disease and the absence of the major symptoms of our patient made this case unique for PPAS.

Pulmonary embolism (PE) is the major subtype of venous thromboembolism. The mortal features of pulmonary thromboendarterectomy (PTE) made this disease the most fatal disease after stroke and coronary artery disease. The developing new treatment techniques

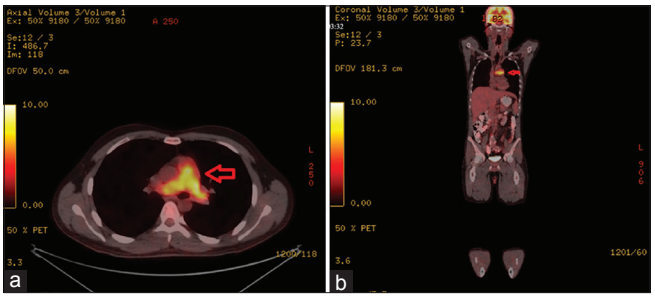


**Figure 2:** (a and b) A PET-computed tomography scan shows pathological F-fluorodeoxyglucose uptake in the same area (red arrow) after failed the pulmonary thromboendarterectomy therapy. PET: Positron emission tomography

provide immediate intervention to this pathology such as local thrombolysis; mechanical extraction devices; hemodynamic support devices; extracorporeal membrane oxygenation; and surgical embolectomy.[7] The patient's cardiopulmonary extra load causes the main symptoms such as dyspnea, syncope, presyncope, hemoptysis, and palpitations. However, if a pulmonary embolism diagnosis is present, fever must be a primary symptom of the disease in the existence of septic emboli, according to Ye *et al*.'s study.[8] This physiological reaction is generally the result of inflammation, autoimmune processes, or malignancy as such in our case.[9] The patient was investigated for an infection but no cause for the fever was found.

PTE risk factors must be assessed before an alternative diagnosis is made. Pleuritic pain, hemoptysis, and syncope should direct the physician to PTE instead of an alternative diagnosis.

A comprehensive physical examination of the cardiopulmonary system, especially cardiac auscultation, might detect a clue that helps to enlighten the main pathology. Although cardiac auscultation skills are facing the danger of disappearing, probably due to the inappropriate use of high-tech tools such as Doppler echocardiography, cardiac auscultation is still an important and cost-effective screening method for the physician.[10]

When a physician doubts that there might be a chronic pulmonary embolus in the absence of the symptoms of acute embolus, alternative diagnoses should be considered. It should not be forgotten that pulmonary arteries might be obliterated by different causes, and this results in pulmonary hypertension.[11] Because the patient had pulmonary thrombus, physicians can have anchoring bias and do not seek other alternative diagnoses. Sarcomas are a rare type of malignancy that a physician must keep in mind as an alternative diagnosis particularly when the patients come with vague symptoms such as fever.

## Conclusion

In our literature review, based on the initial symptomatology of the patients of PPAS, we found that they generally had progressive dyspnea and cough. We emphasize that if there is only unexplained fever with concurrent new murmur in unknown cardiopulmonary pathology, malignancies must be included in the differential diagnoses while considering acute/chronic pulmonary embolism. This interaction could be a life-saving moment even though the survival rate is low for those seeking the cure.

### Author contributions statement

This case has been prepared with the contributions (interpretation, design, discussion, etc.) equally of all the contributors above, and the submitted version has been accepted by the contributors. The participation and co-work of the contributors were approved by themselves.

### Conflicts of interest

None declared.

### Consent to participate

Our patient has been informed of the case above and confidentiality and signed an informed consent form voluntarily.

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