Sri Lanka Journal of Medicine **SLJM**

Case Report

Citation: Wettasinghe MC[1], Perera S[2], Wickramasinghe ND[3], 2023. Pulmonary artery filling defects - Not all are pulmonary emboli. Sri Lanka Journal of Medicine, pp 82-84.
DOI: https://doi.org/10.4038/sljm.v32i2.418

Pulmonary Artery Filling Defects - Not all are Pulmonary Emboli

MC Wettasinghe[1], S Perera[2], ND Wickramasinghe[3]

[1] Base Hospital Teldeniya, Teldeniya, Sri Lanka
[2] National Hospital Kandy, Sri Lanka
[3] Department of Community Medicine, Faculty of Medicine and Allied Sciences, Rajarata University of Sri Lanka

Correspondence:
MC Wettasinghe
E-mail: chamimw003@yahoo.com
https://orcid.org/0000-0001-9183-2395

ABSTRACT

Differentiating pulmonary artery sarcoma from pulmonary artery emboli is challenging since the clinical presentation and imaging features of both conditions are often similar. Timely differentiation of the two conditions is important as delayed diagnosis often results in the progression of pulmonary artery sarcoma leading to poor prognosis and unnecessary anticoagulant therapy. We describe a case of a middle-aged male, who was initially diagnosed with pulmonary embolism, but later diagnosed with a pulmonary artery sarcoma. The case report emphasizes the importance of early imaging findings, which may raise the suspicion prompting further assessment of alternative diagnoses such as pulmonary artery sarcoma.

Keywords: *Pulmonary artery sarcoma, pulmonary embolism, Case report, CT Pulmonary Angiogram*

INTRODUCTION

Pulmonary artery sarcoma (PAS) is a rare aggressive tumour that can mimic pulmonary embolism (PE) in both clinical presentation and imaging findings [1,2,3]. There are often delays in the diagnosis of PAS due to its resemblance to PE, leading to delays in treatment and high mortality [4]. Further, those with PAS would undergo unnecessary anticoagulant therapy. The true magnitude of the incidence of PAS is difficult to ascertain as most cases are misdiagnosed as PE and autopsies are not performed on those who die due to presumed PE [4]. Current knowledge on PAS, especially related to their clinical presentations and imaging features, is largely based on case reports and case series due to its rarity and being misdiagnosed as PE [3,5].

Approximately, 400 cases of PAS have been reported in the literature [6]. A complete surgical resection is said to be the best option for survival. Yet, these tumours have high recurrence rates and overall PAS has a very poor prognosis [4].

We describe a rare case of a middle-aged male, who was initially diagnosed with PE, but later correctly diagnosed with a PAS. The case report emphasizes the importance of early imaging findings, which can raise the suspicion of atypical PE prompting further assessment of alternative diagnoses such as PAS.

Received: 2023-05-07 **Accepted revised version: 2023-12-27** **Published: 2023-12-31**

CASE REPORT

A 59-year-old previously well-male patient, who was under investigation for occult malignancy, was referred for a CT abdomen and chest. He was diagnosed with right pulmonary artery embolus 2 months back and since then was on anticoagulant therapy.

His initial presentation was progressive dyspnoea, which worsened over a period of one month. With the clinical suspicion of PE, he had had a CT pulmonary angiogram (CTPA), which showed an embolus filling the right main pulmonary artery. His full blood count, C-reactive protein level and ESR were within the normal limit. Further, the D-dimer level remained normal. He started on thrombolytics and discharged on oral anticoagulants. His lower limb venous duplex scan showed no deep vein thrombosis.

While on treatment, he had been experiencing exertional dyspnoea without significant deterioration. Since he had no known risk factors for PE, he was investigated for causes of unprovoked PE and to rule out occult malignancy.

The contrast-enhanced CT chest revealed a large filling defect within the right main pulmonary artery and extending into the main pulmonary trunk. There was distension of the right pulmonary artery and also linear enhancing areas were seen within the hypoattenuating filling defect/mass (Figure 1). This protrusion of the mass into the right ventricular outflow tract was identified as the 'wall eclipsing sign'. When compared with his previous CT images, which were taken 2 months earlier (Figure 2), there was progression of the suspected embolus. However, there were no peripheral emboli on both occasions. The only subtle atypical feature seen in the initial CTPA study was mild distension of the pulmonary artery with the embolus. Imaging findings led to the diagnosis of pulmonary artery neoplasm instead of an embolus and the patient was referred to a specialized vascular centre. Unfortunately, a contrast-enhanced CT chest done at the tertiary care unit revealed extra-luminal extension of the mass and it was not amenable for any surgical intervention; thus, the tumour was not biopsied due to the high risk of complications. The patient was referred for palliative chemotherapy.

DISCUSSION

PAS is a rare malignant tumour arising from the mesenchymal cells of the pulmonary artery [5]. PAS is commonly misdiagnosed as PE, and this leads to delays in the diagnosis and also to unnecessary anticoagulation therapy [7]. Further, the clinical presentation of PAS is non-specific and basic laboratory investigations are not specific in differentiating PAS from PE [3,8]. Thus, PAS is commonly misdiagnosed as PE in the initial imaging studies and by the time of the diagnosis of PAS, the disease may become advanced [3]. Studies have suggested some clinical features that would alert the clinical team in suspecting alternative diagnosis of PE. These include gradual onset of symptoms, presence of cough and haemoptysis, presence of systemic symptoms and asthenia, lack of improvement with adequate anticoagulation and absence of deep venous thrombosis [4].

Further, there are reported specific imaging findings that are useful in differentiating PAS from PE [8]. It is important to identify these subtle findings, which can aid in early diagnosis of PAS. On CTPA, the PAS occupies the entire vascular lumen and expands the pulmonary artery. There might be areas of contrast enhancement within the mass and sometimes extra-luminal extension. PE usually has multiple filling defects with peripheral emboli [8], while PAS is seen within the main or proximal pulmonary arteries [2]. The wall eclipsing sign is said to be pathognomonic for PAS and is defined as having a low-density mass occupying the entire lumen of the pulmonary artery with protrusion of the proximal part towards the right ventricular outflow tract and eclipsing one or both walls of the pulmonary artery [9]. This sign was seen in the second CT thorax of our patient (Figure 1). Our patient did not have peripheral thrombi at the initial CTPA, but the mass was occupying the entire lumen. Further, the proximal end of the mass was not convex but was concave. There was no extra-luminal extension or enhancing areas within the mass. Progression of the luminal filling defect or suspected embolus was seen despite anticoagulant therapy on the subsequent contrast-enhanced (arterial phase) CT with enhancing areas, which aided in the diagnosis of PAS in our patient.

Further, in some previously reported cases, the pathological diagnosis was not obtained due to the high risk of complications and the diagnosis was based purely on imaging findings, as in our case [1].

Although rare, having a very high degree of suspicion on atypical appearances of the embolus may help in hastening the diagnosis of PAS over PE.

This case highlights the importance of identifying the key imaging features that help to differentiate PE from a pulmonary artery neoplastic lesion. Knowledge of the imaging features would help in suspecting the diagnosis at the earliest encounter, since the early diagnosis of the PAS may provide the option of surgical resection, which is the favourable treatment for PAS.

Author Declaration

Authors' contributions:
Study concept and design: M.C.W. and S.P.; Acquisition and interpretation of data: M.C.W. and S.P; Drafting of the manuscript: M.C.W. and S.P
All authors read and approved the final version of the manuscript.

Conflicts of interest:
The authors declare that there is no financial or non-financial conflict of interest.

Funding statement:
Self-funded

Ethics statement: Informed verbal consent was obtained from the patient.

Consent for publication:
Informed verbal consent was obtained from the patient. The case report only contains CT images and the case report, and the images included do not contain any personal identifiable information.

Statement on data availability:
All data generated during this study are included in this published article.

REFERENCES

1. Kim C, Kim MY, Kang JW, Song JS, Lee KY, Kim SS. Pulmonary artery intimal sarcoma versus pulmonary artery thromboembolism: CT and clinical findings. Korean journal of radiology. 2018;19(4):792-802.
2. Srivali N, Yi ES, Ryu JH. Pulmonary artery sarcoma mimicking pulmonary embolism: a case series. QJM: An International Journal of Medicine. 2017 May 1;110(5):283-6.
3. Ushioda R, Kitahara H, Ise H, Koichi Y, Wakabayashi N, Tanaka C, Nakanishi S, Ishikawa N, Kamiya H. A case of pulmonary artery sarcoma that was initially misdiagnosed as pulmonary embolism. Journal of Surgical Case Reports. 2019 Mar;2019(3): rjz078.
4. Al-Mehisen R, Al-Halees Z, Alnemri K, Al-Hemayed W, Al-Mohaissen M. Primary pulmonary artery sarcoma: A rare and overlooked differential diagnosis of pulmonary embolism. Clues to diagnosis. International Journal of Surgery Case Reports. 2019 Jan 1; 65:15-9.
5. Bandyopadhyay D, Panchabhai TS, Bajaj NS, Patil PD, Bunte MC. Primary pulmonary artery sarcoma: a close associate of pulmonary embolism—20-year observational analysis. Journal of thoracic disease. 2016 Sep;8(9):2592.
6. Bai X, Ruan L. A case report of primary pulmonary artery intimal sarcoma. European Journal of Medical Research. 2021 Dec;26(1):1-6.
7. Yazgan, C., Ertürk, H., & Taskin, A. (2020). Unusual cause of filling defect in pulmonary artery: pulmonary artery sarcoma. The Pan African Medical Journal. 2020; 35, 41. https://doi.org/10.11604/pamj.2020.35.41.19563
8. Altshuler E, Lowther G, Jantz M. Primary Pulmonary Artery Sarcoma Confined to the Left Pulmonary Artery. J Investig Med High Impact Case Rep. 2021 Jan-Dec; 9:23247096211014687. doi: 10.1177/23247096211014687. PMID: 33969719; PMCID: PMC8113362.
9. Zhang, S., Zhang, Y., Liu, M., Tao, X., Xie, W., Wan, J., & Zhai, Z. (2021). Radiological, histopathological findings, and clinical outcome of pulmonary artery sarcoma. Pulmonary circulation, 11(1), 2045894020940537. https://doi.org/10.1177/2045894020940537
10. Cervilla-Muñoz, E., Galeano-Valle, F., Del-Toro-Cervera, J., Calleja-Cartón, E., & Demelo-Rodríguez, P. (2020). Differential diagnosis and treatment approach to pulmonary artery sarcoma: a case report and literature review. ERJ Open Research, 6(3).