Interesting Image
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Significance of Clinico-radiological Correlation in a Patient with Pulmonary Intimal Sarcoma Simulating as Pulmonary Thromboembolism

Pulmoner Tromboembolizmi Taklit Eden Pulmoner İntimal Sarkomlu Bir Hastada Klinik-radyolojik Korelasyonun Önemi

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Abstract

Pulmonary intimal sarcoma (PAS) is a highly aggressive malignant mesenchymal tumor affecting the central pulmonary arteries. Similar clinical presentation and indeterminate laboratory parameters often result in misdiagnosis of this condition as pulmonary thromboembolism, which is a relatively common disease. Certain imaging features can however allow differentiation between these two diagnoses. We present one such case of PAS that was initially treated as pulmonary embolism; and briefly review the relevant imaging characteristics to avoid overlooking PAS especially in patients with an atypical clinical history for thromboembolism.

Keywords: Pulmonary intimal sarcoma, CT pulmonary angiography, pulmonary thromboembolism, wall eclipse sign

Öz

Pulmoner intimal sarkom (PAS), merkezi pulmoner arterleri etkileyen oldukça agresif bir malign mezenkimal tümördür. Benzer klinik tablo ve belirsiz laboratuvar parametreleri sıklıkla bu durumun, nispeten yaygın bir hastalık olan pulmoner tromboembolizm olarak yanlış teşhis edilmesine neden olur. Bununla birlikte, belirli görüntüleme özellikleri, bu iki durumun ayrımını sağlayabilir. Başlangıçta pulmoner emboli olarak tedavi edilen böyle bir PAS'li hastayı sunduk ve özellikle atipik klinik tromboembolizm öyküsü olan hastalarda PAS'yi gözden kaçırmamak için görüntüleme özelliklerini kısaca gözden geçirdik.

Anahtar kelimeler: Pulmoner intimal sarkom, BT pulmoner anjiyografi, pulmoner tromboembolizm, duvar tutulması belirtisi

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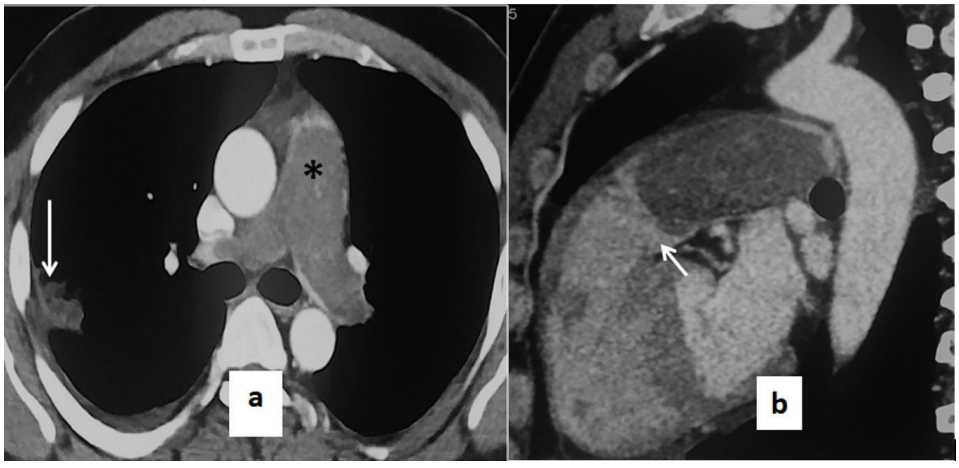

Figure 1. Pulmonary intimal sarcoma (PAS) is an aggressive malignant mesenchymal tumor originating from the subendothelial connective tissue lining of the central pulmonary arteries (i.e. main, right or left) (1,2). This rare condition is often misdiagnosed initially as pulmonary embolism and is followed by initiation of anticoagulation therapy which allows unbridled tumor growth (3,4). Although both diseases manifest as filling defects in pulmonary arteries, PAS is usually lobulated or nodular and forms an acute angle with the intimal surface with heterogeneous enhancement as compared to acute pulmonary embolism, which typically manifests as thin and thread-like defects (5,6). Vascular expansion with complete luminal occlusion is another distinctive feature of PAS and reflects the preferential intraluminal growth of the tumor.
Baseline computed tomography (CT) pulmonary angiography of a 35-year-old gentleman with complaints of non-productive cough and shortness of breath for 1 year with raised D-dimer levels (836 ng/mL, normal range <500 ng/mL) showed a lobulated heterogeneous intraluminal filling defect occluding the main pulmonary artery with extension into the right and left branches, mimicking a saddle embolus (asterisk in a). A small subpleural consolidation (arrow, a) seen in the right upper lobe represents pulmonary infarct. (b) Sagittal CT confirms the complete occlusion of the main pulmonary trunk with proximal extension of the filling defect into the region of the pulmonary valve (arrow). Although no obvious extravascular extension was seen, the lesion caused obscuration of the walls of the pulmonary arteries. With an initial diagnosis of massive pulmonary embolism, he was started on acenocoumarol but did not show any improvement.

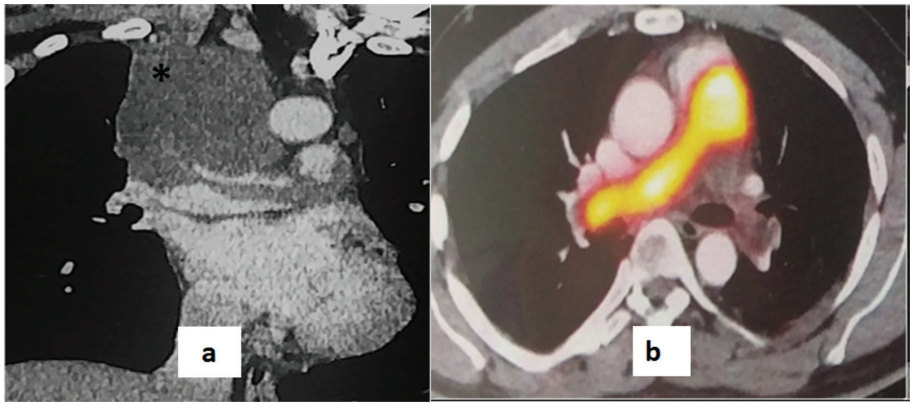

Figure 2. Coronal CT pulmonary angiography (a) performed after 7 months demonstrates the extension of the mass into the paratracheal location (asterisk) with the involvement of the superior vena cava. Such atypical features, including vascular expansion, complete occlusion of the pulmonary artery, and frank extravascular extension raised the suspicion of PAS. The patient underwent positron emission tomography (PET)/CT that showed avid radiotracer uptake (b) in the mass, which is not seen in embolism. The patient then underwent endobronchial ultrasound (EBUS) guided fine needle aspiration cytology (FNAC) of the lesion.
The most specific sign aiding in the detection of PAS is overt extravascular extension, which occurs in later stages (4). "Wall eclipse sign" is diagnostic of PAS and incorporates three criteria: (i) a low-density mass completely occupying the central pulmonary arteries, (ii) proximal protrusion of the mass into the right ventricular outflow tract, and (iii) obscuration of the one or both walls of the involved artery. All three are absent in thromboembolism (7). An acute angle of contact, presence of intratumoural vessels, absence of vessel tapering, and webs are helpful features for differentiating PAS from chronic thromboembolism (4,6). Although PAS does not show enhancement on CT, it however demonstrates avid uptake on ^{18}F-fluorodeoxyglucose PET, which reflects its increased metabolic activity; combined PET/CT is the most definitive modality of distinguishing PAS from non-neoplastic thromboembolic disease (5).

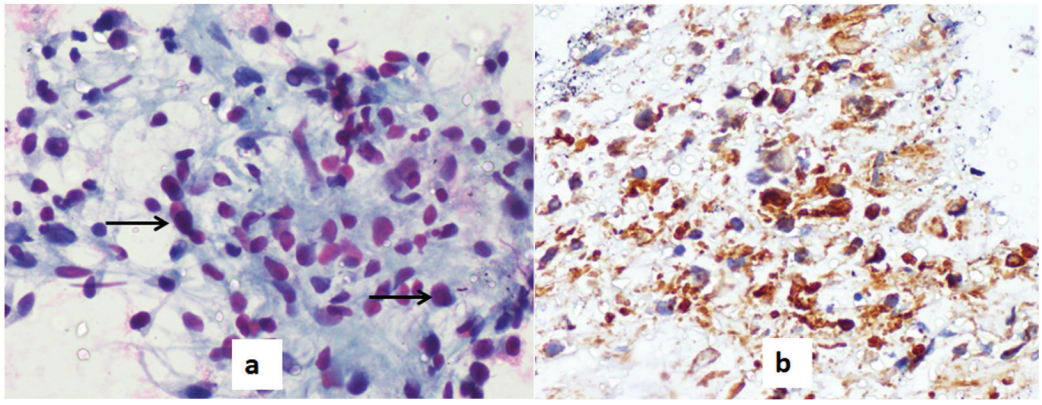

Figure 3. (a) EBUS-guided FNAC of the mass shows tumor cell fragments with pleomorphic oval to spindle cells showing high nucleo- cytoplasmic ratio (arrows in a) on high power photomicrograph (magnification 200x) with papanicolaou stain; (b) the tumor cells also showed strong positivity for smooth muscle actin (brown cells, same magnification), which was consistent with the diagnosis of PAS.

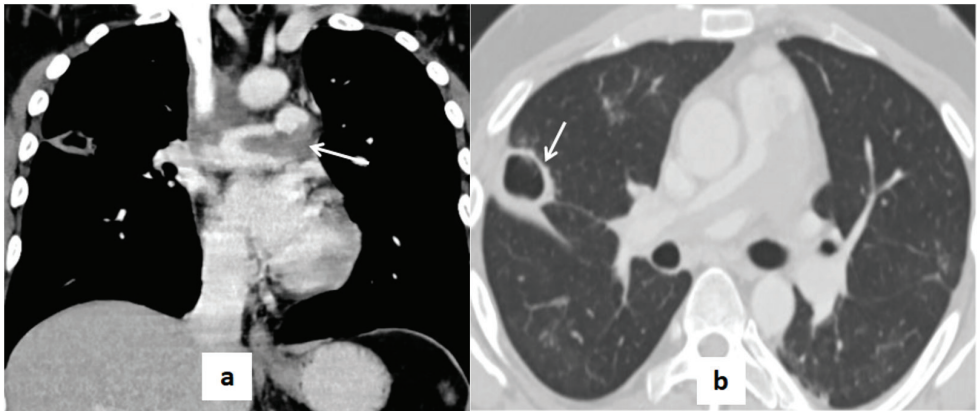

Figure 4. Coronal contrast-enhanced CT image post institution of cytoreductive chemotherapy revealed a reduction in the size of the mass (arrow in a) and cavitation in the pulmonary infarct (arrow in b) representing a partial response. However, the patient developed severe thrombocytopenia as a complication of chemotherapy with pulmonary hemorrhage and succumbed to the illness.

Ethics

Informed Consent: Obtained from the patient's attendant.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.R., D.J., Concept: E.D., Design: E.D., Data Collection or Processing: S.C., Analysis or Interpretation: S.C., E.D., Literature Search: S.C., Writing: S.C., E.D., S.R., D.J.

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References

1. Scheidl S, Taghavi S, Reiter U, Tröster N, Kovacs G, Rienmüller R, Lang S, Klepetko W, Olschewski H. Intimal sarcoma of the pulmonary valve. Ann Thorac Surg 2010;89:e25-e27.
2. Burke A, Tavora FR, Maleszewski JJ, Fraizer AA. Tumors of the Heart and Great Vessels. In: AFIP Atlas of Tumor Pathology. 4th ed. Silver Spring, Md: American Registry of Pathology, 2015.
3. Mussot S, Ghigna MR, Mercier O, Fabre D, Fadel E, Le Cesne A, Simonneau G, Dartevelle P. Retrospective institutional study of 31 patients treated for pulmonary artery sarcoma. Eur J Cardiothorac Surg 2013;43:787-793.
4. Ropp AM, Burke AP, Kligerman SJ, Leb JS, Frazier AA. Intimal sarcoma of the great vessels. Radiographics 2021;41:361-379.
5. 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 J Radiol 2018;19:792-802.
6. Cervilla-Muñoz E, Galeano-Valle F, Del-Toro-Cervera J, Calleja-Cartón E, Demelo-Rodríguez P. Differential diagnosis and treatment approach to pulmonary artery sarcoma: a case report and literature review. ERJ Open Res 2020;6:00124-2020.
7. Gan HL, Zhang JQ, Huang XY, Yu W. The wall eclipsing sign on pulmonary artery computed tomography angiography is pathognomonic for pulmonary artery sarcoma. PLoS One 2013;8:e83200.