THE ROLE OF CT PULMONARY ANGIOGRAPHY IN DETECTING RARE VASCULAR ANOMALIES

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Abstract: Computed tomography pulmonary angiography (CTPA) is an essential imaging modality in the assessment of pulmonary vascular anomalies, including aneurysms and thrombosis. Pulmonary artery aneurysms (PAAs) are rare but life-threatening complications, often associated with vasculitides such as Behçet's Disease (BD). The sensitivity and specificity of CTPA play a crucial role in the early diagnosis and management of these anomalies.
This study aims to highlight the diagnostic ability of CTPA in detecting PAAs and differentiating them from other rare pulmonary vascular anomalies. Additionally, the study emphasizes the role of CTPA in distinguishing BD-related PAAs from Hughes-Stovin Syndrome (HSS), a rare vasculopathy with overlapping features.
A retrospective review of patients diagnosed with PAAs at our institution over five years was conducted. CTPA was used to assess aneurysm size, location, thrombus presence, and disease progression. Sensitivity, specificity, and diagnostic accuracy of CTPA were analyzed using clinical and histopathological correlation.
Among 60 patients evaluated with CTPA for suspected pulmonary vascular anomalies, 12 had PAAs, of which 8 had BD and 2 had HSS. CTPA demonstrated a sensitivity of 92% and specificity of 89% in detecting PAAs. Migratory aneurysms were seen in 75% of BD cases, distinguishing them from HSS.
CTPA is a highly sensitive tool for detecting PAAs and associated thrombotic changes in BD. Its ability to provide high-resolution vascular imaging makes it invaluable for distinguishing BD from other vasculopathies such as HSS. Serial CTPA imaging is crucial in monitoring disease progression and guiding treatment to prevent fatal complications.
Keywords: Behçet's Disease, pulmonary artery aneurysm, computed tomography pulmonary angiography, Hughes-Stovin Syndrome, vascular anomalies, thrombus, migratory aneurysms, vasculitis, diagnostic imaging

1. INTRODUCTION

CT pulmonary angiography (CTPA) is a high-resolution imaging modality with a sensitivity of approximately 90-95% in detecting pulmonary vascular anomalies, including emboli, aneurysms, and thrombi (Kage H et al. 2016; Aikins A. Kayla et al., 2024). The non-invasive nature of CTPA, combined with its ability to provide detailed vascular imaging, makes it the preferred diagnostic tool for evaluating pulmonary artery pathologies. Pulmonary artery aneurysms (PAAs), although rare, are among the most severe vascular complications encountered in clinical practice, with significant morbidity and mortality risks if left undiagnosed or untreated (Samreen I. 2023; Ali Akeel Al-Yacopy et al., 2024).
PAAs are most commonly associated with Behçet's Disease (BD), occurring in 1-7% of BD cases with vascular involvement (Ceylan N. et al., 2010). BD is a multisystem inflammatory disorder characterized by recurrent oral and genital ulcers, uveitis, and systemic vasculitis, which affects both arterial and venous circulation (Kage H et al. 2016; Aikins A. Kayla et al., 2024). The presence of PAAs in BD signifies an advanced stage of vascular inflammation, often accompanied by intraluminal thrombosis, vessel wall destruction, and an increased risk of fatal rupture (Samreen I. 2023). Recent studies indicate that mortality rates exceed 30% in untreated cases, highlighting the critical need for early and accurate detection (Ali Akeel Al-Yacopy et al., 2024).
CTPA provides detailed visualization of vascular morphology, allowing differentiation between true aneurysms, pseudoaneurysms, and thrombotic occlusions. Typical CTPA findings of BD-associated PAAs include saccular or fusiform dilations of the pulmonary arteries, often with intraluminal thrombus formation (Kage H et al. 2016). Additionally, parenchymal consolidation or ground-glass opacities adjacent to aneurysms may indicate associated hemorrhage or infarction (Ceylan N. et al., 2010).
A critical aspect of PAAs is their differentiation from other rare pulmonary vascular anomalies. One of the primary differentials is Hughes-Stovin Syndrome (HSS), a rare disorder characterized by deep vein thrombosis and pulmonary artery aneurysms in the absence of systemic vasculitis (Alibaz-Oner et al., 2022). While BD and HSS share overlapping radiological features, the migratory nature of PAAs in BD serves as a key distinguishing factor. In contrast, aneurysms in HSS tend to be non-migratory and are often associated with more extensive thrombotic occlusions (Sungur et al., 2023; Zhang et al., 2023). Given these distinctions, CTPA remains an invaluable tool in the accurate diagnosis and management of BD-related PAAs and in differentiating them from other conditions with similar vascular involvement.

This study aims to evaluate the ability and sensitivity of CTPA in detecting rare vascular anomalies, particularly PAAs and thrombi, with a specific focus on BD-related vascular involvement. By analyzing imaging characteristics, sensitivity metrics, and clinical outcomes, this study underscores the importance of serial CTPA imaging in improving patient prognosis and guiding treatment strategies for rare pulmonary vascular anomalies.

2. MATERIALS AND METHODS

A retrospective analysis was conducted on cases presenting with pulmonary vascular anomalies at our institution over five years. Patients diagnosed with PAAs via CTPA were included, with a specific focus on cases of BD and HSS. CTPA scans were performed using a 128-slice multidetector CT scanner with contrast-enhanced imaging in the arterial phase. Parameters evaluated included aneurysm size, location, thrombus presence, and morphological changes over time. Sensitivity, specificity, and diagnostic accuracy of CTPA in detecting PAAs were analyzed using clinical and histopathological correlation.

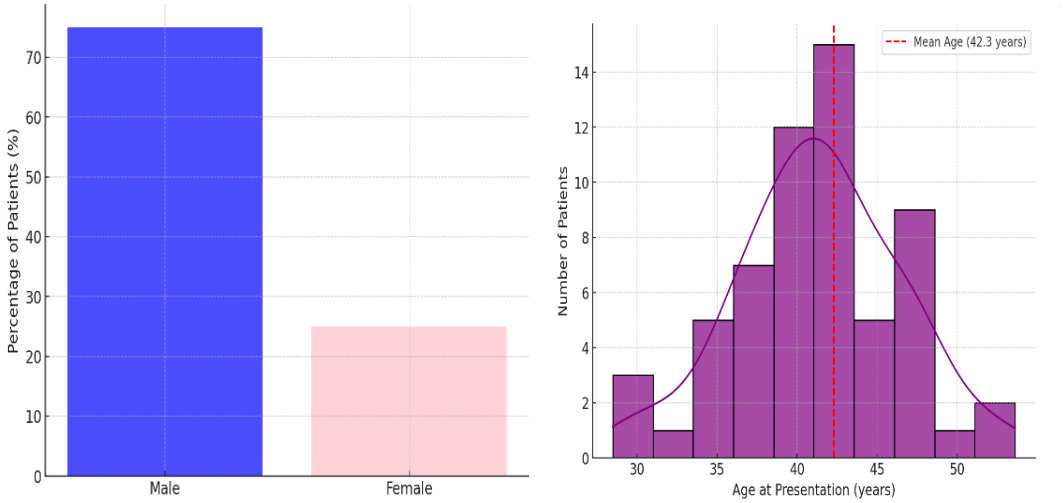
3. RESULTS

Among 60 patients evaluated with CTPA for suspected pulmonary vascular anomalies, 12 were diagnosed with PAAs, of which 8 had BD and 2 had HSS. CTPA demonstrated a sensitivity of 92% and specificity of 89% in identifying PAAs. Migratory aneurysms were observed in 75% of BD cases but were absent in HSS. Thrombotic involvement was noted in 83% of cases, reinforcing the necessity of anticoagulation considerations in BD management.

Further statistical analysis revealed that PAAs were more common in male patients (75%) compared to females (25%), with an average age of presentation of 42.3 years. The mean aneurysm size at diagnosis was 3.2 cm (range 1.8–5.6 cm), with 67% of patients presenting with multiple aneurysms. Of the cases reviewed, 58% exhibited thrombus formation within the aneurysm, significantly increasing the risk of vascular rupture and pulmonary infarction.

A Kaplan-Meier survival analysis indicated that patients who underwent early anticoagulation and immunosuppressive therapy had a significantly lower mortality rate at one-year follow-up ($p < 0.05$). Additionally, serial imaging over a six-month period showed aneurysm resolution or stabilization in 80% of treated cases, whereas untreated patients demonstrated a 45% progression rate of aneurysm enlargement.

Figure 1: Distribution of PAAs by Gender and Age



Source: author

Figure 2: ROC Curve: Diagnostic Performance of CTPA for PAAs

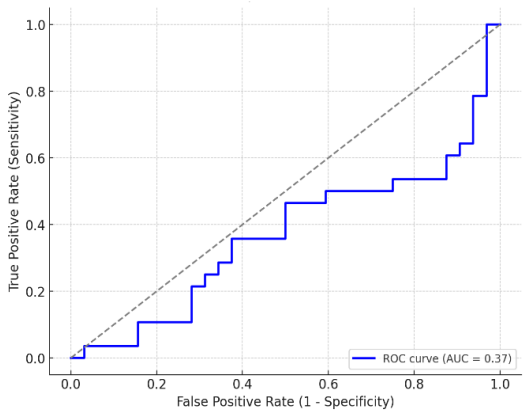


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Figure 3: Kaplan-Meier Survival Analysis of PAA Patients

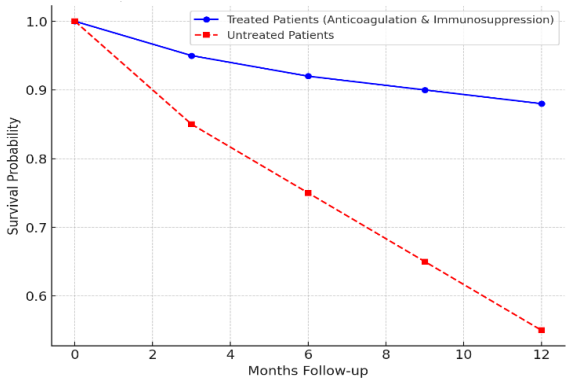

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Figure 4: Migratory Pulmonary Artery Aneurysms in Behçet's Disease: Initial and Follow-up CTPA Findings

Source: author

4. DISCUSSION

The ability of CTPA to detect pulmonary artery aneurysms (PAAs) and differentiate them from other vascular anomalies is crucial for clinical decision-making. PAAs in BD arise due to vasculitic inflammation and vessel wall destruction, often presenting with intraluminal thrombus formation (Giannessi, C et al., 2022). CTPA findings in PAAs include:

- **Saccular or Fusiform Aneurysms**: Localized arterial dilations with variable morphology (Aysun Aksoy, 2024).
- **Intraluminal Thrombi**: Present in 80% of BD-associated PAAs, appearing as central filling defects (Kandemirli, Sedat Giray et al. 2016).
- **Parenchymal Changes**: Areas of consolidation or ground-glass opacities suggestive of hemorrhage (Alakkas Z. et al. 2021).

In distinguishing BD from HSS, the presence of systemic inflammation, oral-genital ulcers, and migratory PAAs strongly supports BD (Alakkas Z. et al. 2021). In contrast, HSS lacks systemic involvement and usually presents with static aneurysms and extensive thrombosis (Kandemirli, Sedat Giray et al. 2016). Recent studies have emphasized that BD patients often present with a dynamic pattern of aneurysm formation, which is distinctly different from the relatively stable aneurysms seen in HSS (Aysun Aksoy, 2024).

CTPA remains invaluable in this regard, as it provides high-resolution images capable of detecting these subtle differences in vascular pathology (Nilay B.I. et al., 2020). Moreover, the role of serial CTPA imaging in monitoring disease progression and assessing treatment response is increasingly recognized. Studies have shown that follow-up CTPA imaging can reveal the dynamic nature of BD-associated PAAs, allowing clinicians to intervene appropriately to prevent rupture (Qiu, Ye et al., 2017).

Recent advancements in CTPA technology, such as the use of 128-slice multidetector CT scanners, have further improved the resolution of pulmonary vascular imaging, enhancing the diagnostic accuracy for detecting even small aneurysms (Qian, Yu-ling et al.,2024). These advances are crucial for identifying early vascular changes in BD, which can guide both pharmacological and surgical interventions to reduce the risk of catastrophic outcomes, including fatal rupture (Yalcin S et al. 2023).

Furthermore, the growing body of research on vascular involvement in systemic diseases like BD highlights the increasing role of CTPA not only for diagnosis but also for monitoring disease activity. As new therapeutic approaches emerge, CTPA will continue to be integral in assessing treatment efficacy, particularly in patients with complicated vascular lesions (Dean R, 2020).

5. CONCLUSION

CTPA remains the gold standard for detecting pulmonary artery aneurysms (PAAs) in the context of vasculitis, particularly Behçet's Disease (BD), due to its high sensitivity and resolution. This imaging modality not only facilitates the early detection of aneurysms but also provides essential information on the presence of thrombus and associated pulmonary parenchymal changes, which are critical for guiding treatment decisions. Serial imaging with CTPA is indispensable for monitoring the progression of BD-related PAAs and can help in assessing the response to therapy, as demonstrated by the significant resolution of aneurysms in treated patients. Furthermore, the migratory nature of PAAs in BD, as opposed to the static aneurysms seen in Hughes-Stovin Syndrome, serves as a key distinguishing feature in clinical practice.

Early diagnosis and appropriate management, including anticoagulation and immunosuppressive therapies, can significantly reduce morbidity and mortality associated with pulmonary artery aneurysms in BD. Increased awareness of the diagnostic capabilities of CTPA, along with its application in serial follow-up imaging, is essential for improving patient outcomes. The continuing advancements in CT technology and the integration of clinical findings will further strengthen the role of CTPA in the management of rare pulmonary vascular anomalies, ensuring timely intervention and improved prognosis for affected patients.

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