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RESEARCH ARTICLE

Imaging Features of Thoracic Manifestations of Behçet's Disease: Beyond Pulmonary Artery Involvement

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Abstract: ***Background*:** Behçet's disease is a chronic multisystemic vasculitis affecting vessels of different sizes in various organs. Thoracic manifestations of the disease show a wide spectrum involving a variety of anatomic structures within the chest. However, pulmonary artery involvement is a typical manifestation of the disease that contributes significantly to mortality in patients. The study aimed to analyze CT features of thoracic manifestations, particularly pulmonary artery involvement, and to quantitatively assess bronchial arteries in Behçet's disease.

***Methods*:** Patients with Behçet's disease who underwent CT scans for suspected thoracic involvement between 2010 and 2018 were included. CT findings of 52 patients were retrospectively analyzed for thoracic manifestations of the disease. Bronchial arteries were assessed regarding diameter in patients with/without pulmonary artery involvement. The pulmonary symptoms were noted.

***Results*:** Of the 52 patients, 67% had thoracic manifestations including pulmonary artery involvement, parenchymal changes, superior vena cava thrombosis, and intracardiac thrombus. Pulmonary artery involvement was observed in 50% of the cohort. Peripheral pulmonary arteries (77%) were the most commonly affected branches, followed by lobar (42%) and central (35%) pulmonary arteries. Other thoracic findings were significantly correlated with pulmonary artery involvement ($p<0.05$). Compared to patients without pulmonary artery involvement, those with pulmonary artery involvement had a higher bronchial artery diameter ($p<0.05$) and occurrence rate of dilated bronchial arteries.

***Conclusion*:** Involvement of peripheral pulmonary arteries is frequently encountered in Behçet's disease and it can resemble pulmonary nodules. Dilated bronchial arteries, which can be observed in cases of pulmonary artery involvement, should be considered in patients with hemoptysis.

ARTICLE HISTORY

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1. INTRODUCTION

Behçet's disease (BD) is a chronic inflammatory disease characterized by recurrent uveitis, oral and genital ulcers, and systemic manifestations such as vascular, thoracic, and central nervous systems. Vasculitis is the main pathological process of the disease that affects both large and small vessels.

Thoracic involvement is a relatively unusual complication of BD [1]. Its characteristic manifestation is pulmonary artery involvement (PAI) that contributes significantly to mortality in patients [2]. Modern imaging methods such as Modern imaging methods such as pulmonary computed tomography (CT) angiography (CTA), magnetic resonance (MR) angiography (MRA) have replaced conventional angiography to diagnose PAI [3, 4]. Some authors believe that CTA is more sensitive than MRA in demonstrating particularly small pulmonary artery aneurysm (PAA) [1, 5]. CT scan can also demonstrate the entire spectrum of thoracic manifestations as well as PAI in BD [3-6]. Moreover, reformatted images and three-dimensional reconstructions are essential to guide endovascular treatment or to prepare surgery.

Radiological findings of thoracic involvement have been reported in BD, but to our knowledge, most descriptions are presented in pictorial essays and clinical studies [7-9]. In addition, to the best of our knowledge, there are no original studies on bronchial artery (BA) diameter measurement in BD. Therefore, we aimed to further analyze the CT features of thoracic manifestations expanding on PAI in BD. We also focused on assessing the diameter of BAs in patients with PAI.

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2. MATERIALS AND METHODS

This retrospective study was approved by our institutional review board (date: June 27, 2018, approval number:2012-KAEK-15/1704).

2.1. Study Population

By using the electronic clinical database at the Atatürk Chest Disease and Chest Surgery Training and Research Hospital,68 consecutively registered patients with BD were identified between January 2010 and April 2018. Of these patients, we included those who had undergone contrast-enhanced CT and/or pulmonary CTA to investigate thoracic involvement and fulfilled the International Study Group criteria for BD diagnosis. We excluded patients with pneumonia, malignancy, and collagen vascular disease.

Electronic medical records of all patients were reviewed for information regarding age, sex, initial symptoms, and medical history.

2.2. CT Protocols

CT examinations were performed with two multi-detector CT scanners (Emotion 6, Siemens, Germany and Alexion 16, Toshiba Medical Systems, Japan). Of 82 CT scans, 53 were obtained according to the pulmonary CTA protocol and 29 according to the conventional chest CT protocol.

For CTA, the patients were administered 120 ml of non-ionic contrast material at the rate of 3-4 ml/s into an arm vein by using a power injector. A bolus tracking technique triggered at 100 HU on the pulmonary trunk was used with 10 seconds delay. Other scanning parameters were as follows: pitch, 1.43; rotation time, 0.75 s; slice thickness, 1 mm; and reconstruction interval, 1 mm. Tube currents were adjusted using automatic tube current modulation.

For contrast-enhanced chest CT, 75-80 ml of a non-ionic contrast medium was intravenously administered at the flow rate of 2-3 ml/s using an automated injector. Scanning started at 25 s after the initiation of contrast agent administration.

2.3. Radiological Evaluation of the study cohort

CT images were independently reviewed by two experienced radiologists *(with 17 and 19 years of experience, respectively*)who were blinded to the clinical data. The images were analyzed according to thoracic BD findings by using lung and mediastinal window settings predominantly in the axial plane; coronal and sagittal images were used occasionally to evaluate peripheral pulmonary arteries (PAs). Discrepancies between the two readers were resolved by discussion and consensus. PAI was described here as either PAA and/or isolated pulmonary artery thrombosis (PAT). The location of involved PAs for each patient was noted as central, lobar, or peripheral. The pulmonary trunk and main PAs were defined as central, and segmental-subsegmental PAs were defined as peripheral branches. The level and multiplicity of PAI were also identified. The presence of intramural thrombus was also evaluated in patients with PAA. The following lesions were noted in the lung parenchyma: consolidations, ground-glass opacities (GGO), cavities, mosaic attenuation, atelectasis, and nodules. Pleural-pericardial effusion, superior vena cava (SVC) thrombosis, and intracardiac filling defects were also stated.

Follow-up CT images of 18 patients with PAI were also evaluated regarding changes in PAs and parenchyma.

The diameter of BAs was evaluated only in patients with pulmonary CTA. The measurements were conducted on the axial plane by using mediastinal window settings. The maximum diameter was measured for each BA. BAs with a diameter greater than 2 mm were considered to be dilated. The diameters of BAs were compared between patients with and without PAI. In patients with PAI, the mean pulmonary artery (PA) pressure with echocardiographic assessment was also recorded.

2.4. Statistical Analysis

Chi-square test or Fisher's exact test was used for comparison among various qualitative variables. Values were expressed as either mean ± standard deviation or median with interquartile range after testing the normality of variables using the Shapiro-Wilk test. The number and diameter of BAs were compared using the Mann-Whitney U test. A *p*-value of <0.05 was considered significant for all tests. All statistical procedures were conducted using IBM SPSS statistics version 20 for Windows (IBM Corp., Armonk, NY, USA).

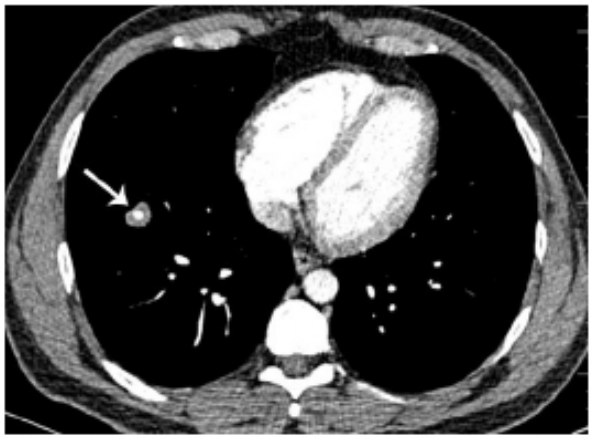

Fig. (1). Axial CT images of a 37-year-old man with hemoptysis demonstrating PA aneurysm in the segmental branches of the middle lobe artery with peripheral thrombosis (arrow). (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

3. RESULTS

The study included 52 patients (median age, 43 years; mean, 44 ±12 years; range, 22-70 years) with a male-to-female ratio of 1. Of these patients, 42 (81%) were presented with respiratory symptoms. The main respiratory symptoms were hemoptysis (42%) and dyspnea (38%). Overall, 35 (67%) patients had thoracic findings on CT scans. PAI,ob-

served as PAA and/or PAT, was detected in 26 (50%) patients(Figs. **1** and **2**). Table **1** presents the details of PAI. Small aneurysms at peripheral PA branches appeared like parenchymal nodules in 4 patients. The continuity of lesions with PA branches was shown with thin sections and reformatted CT images (Figs. **3** and **4**).

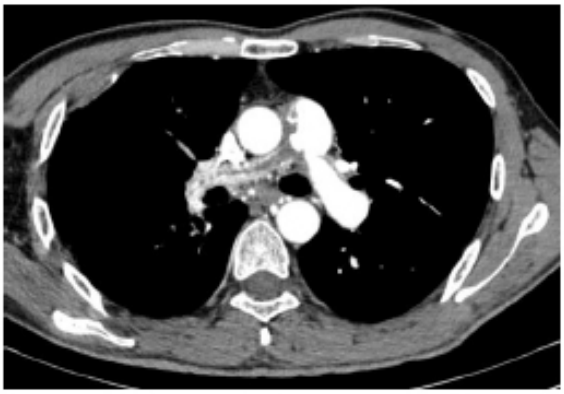

Fig. (2). Axial CT image of a 43-year-old man with dyspnea showing thrombosis of the main and right pulmonary arteries with mediastinal dilated bronchial arteries. (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

Table 1. PA Involvement in 26 patients.

PA Involvement Type	*PA aneurysm*	*Isolated PAT*	*PA Aneurysm+thrombosis*
	20/26 (77%)	**4/26 (15%)**	**2/26 (8%)**
Distribution	***Central***	***Lobar***	***Segmental-subsegmental***
	9 (35%)	11 (42%)	20 (77%)
Location	***Lower Lobes***	***Upper and lower lobes***	***Upper and middle lobes***
	14/26 (54%)	9/26 (35%)	3/26 (11%)

PA: pulmonary artery.

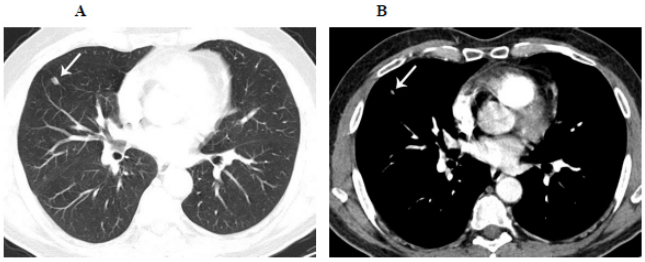

Fig. (3). Axial CT images on parenchymal **(a)** and mediastinal **(b)** window setting of a 53-year-old man with dyspnea showing peripheral PA aneurysm of the right upper lobe suggesting pulmonary nodule (arrow). (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

Abnormal parenchymal findings along with PAI were as follows: peripheral consolidations, nodules, GGO, and cavities. Table **2** shows the clinical and CT characteristics of patients with PAI. Follow-up CT images of 5 patients showed cavities (Fig. **5**).

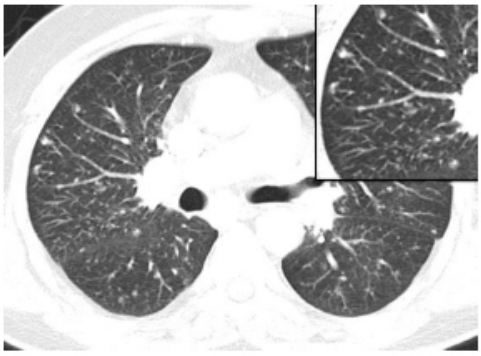

Fig. (4). Axial thin-section CT image of a 51-year-old man with fever indicating multiple small Y- and V-shaped branching opacities representing peripheral pulmonary artery dilatations. (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

Parenchymal findings were subpleural nodules-opacities in patients without PAI. These patients had negative tests or laboratory findings for tuberculosis or other infections.

Two patients had intracardiac thrombus localized in the right ventricles (Fig. **6**). Concentric wall thickening of the left subclavian artery was detected in 1 patient. Three patients showed SVC and other great mediastinal veins thrombosis (Fig. **7**). PAI was significantly correlated with parenchymal and cardiovascular findings ($p<0.05$).

Eighty-one (72%) and 32 (28%) BAs were detected in patients with and without PAI, respectively. Table **3** shows an analysis of the BAs. The diameter of BAs correlated with PAI($p=0.021$). Compared to the PAI subgroup, there were no differences in the correlation of BA diameters between patients with PAA and those with PAT ($p=0.108$). Patients with PAI and dilated BAs more frequently (64%) developed hemoptysis than those with normal BAs. Of the former group, 40% had elevated mean PA pressure as revealed by echocardiography.

4. DISCUSSION

BD was first described in 1937 by Hulusi Behçet, a Turkish dermatologist, as a symptomatic triad of relapsing aphthous oral ulcers, genital ulcers, and uveitis in 3 Eastern Mediterranean patients [10]. Despite its worldwide occurrence, BD is more prevalent in countries along the ancient Silk Road extending from the Mediterranean to Middle-East Asia to Far East Asia [11, 12]. Besides the classical clinical triad, the involvement of other systems such as the gastrointestinal system, the central nervous system, and the cardiovascular system has been reported in BD [13].

The prevalence of thoracic involvement in BD ranges from 1% to 8% [1]. In our study, 67% of the patients had CT findings suggesting thoracic involvement. A previous study reported pulmonary involvement in 14% of patients with BD [2]. In contrast, Edrees *et al*. showed thoracic involvement in 73% of cases [14], explaining that their study had male predominance and included patients with severe ill-

Table 2. Clinical and radiological characteristics of 26 patients with pulmonary artery involvement.

Case No/Age/Sex	Prominent Pulmonary Symptoms	Pulmonary Artery Involvement	CT Findings
1/24/F	Hemoptysis	+	Multiple PAA, consolidation, pericardial-pleural effusion, mosaic attenuation
2/60/M	Chest pain	+	PAA
3/57/F	Dyspnea	+	PAA, peripheral consolidation
4/29/M	Dyspnea	+	Multiple PAA, peripheral consolidation
5/30/M	Dyspnea	+	PAA, cavity
6/33/M	Chest pain	+	Multiple PAA, peripheral consolidation, mosaic attenuation
7/41/M	Hemoptysis	+	Multiple PAA, GGO, peripheral consolidation, atelectasis
8/51/M	Chest pain, fever	+	PAA, pericardial effusion
9/40/M	Hemoptysis	+	Multiple PAA, ICT, left SCA concentric wall thickening, GGO, peripheral consolidation
10/28/M	Nonspecific	+	PAA
11/37/M	Hemoptysis	+	PAA +PAT, subpleural nodule, atelectasis
12/31/M	Dyspnea	+	PAA
13/26/F	Hemoptysis	+	PAA+PAT, atelectasis, subpleural nodule, peripheral consolidation
14/46/F	Chest pain	+	PAA, mosaic attenuation, subpleural nodule, peripheral consolidation
15/34/F	Dyspnea	+	PAT, cavity, GGO
16/53/M	Dyspnea	+	PAA
17/50/F	Hemoptysis	+	PAA, GGO
18/59/F	Dyspnea	+	PAA, peripheral consolidation, mosaic attenuation, GGO
19/60/M	Dyspnea, chest pain	+	PAA, peripheral consolidation, atelectasis
20/22/M	Dyspnea, fever	+	PAA, SVC and jugular vein thrombosis, atelectasis
21/27/M	Hemoptysis	+	Multiple PAA, GGO, peripheral consolidation, ICT, cavity
22/43/M	Dyspnea	+	PAT, SVC and brachiocephalic vein thrombosis, peripheral consolidation, atelectasis
23/27/M	Hemoptysis	+	Multiple PAA, peripheral consolidations
24/53/F	Hemoptysis	+	PAT, mosaic attenuation, SVC, and bilateral brachiocephalic vein thrombosis
25/38/M	Hemoptysis	+	PAT, GGO, cavity
26/35/F	Hemoptysis	+	Multiple PAA, peripheral consolidation, cavity

GGO: ground-glass opacity, ICT: intracardiac thrombus, PAA: pulmonary artery aneurysm, PAT: pulmonary artery thrombosis, SCA: subclavian artery, SVC: superior vena cava.

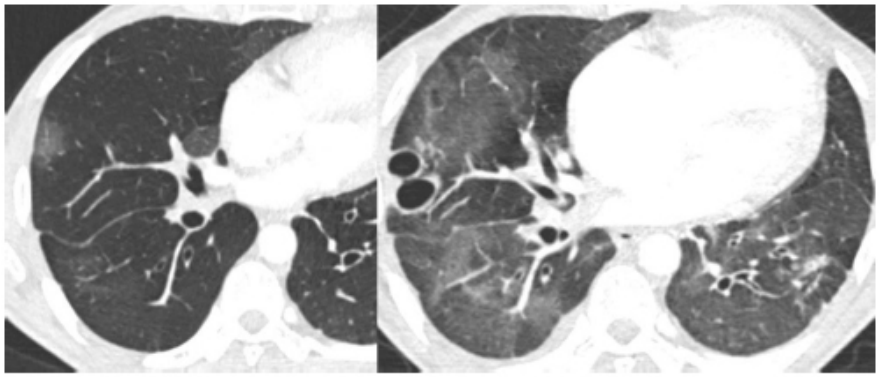

Fig. (5). Axial CT image of a 38-year-old man with hemoptysis revealing peripheral cavitary lesions that evolved from subpleural hemorrhage. (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

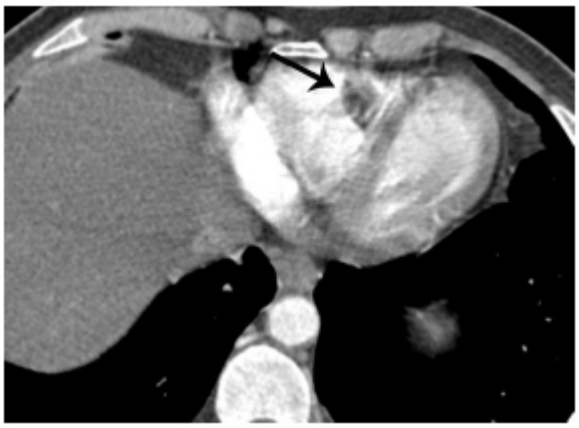

Fig. (6). Axial CT image of 40 -year-old man with hemoptysis showing intracardiac thrombus at the right ventricle confirmed with cardiac MR imaging (arrow). (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

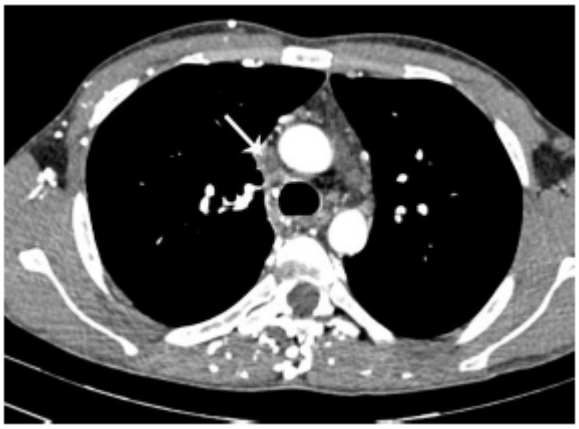

Fig. (7). A 22-year-old man with dyspnea and fever. Axial CTA images demonstrating SVC thrombosis with collateral venous vessels in the mediastinum and the posterior chest wall (arrow). (*A higher resolution / colour version of this figure is available in the electronic copy of the article*).

Table 3. Comparison of BA parameters between the groups with PAI and non-PAI.

-	PAI (*n*=24)	Non-PAI (*n*=11)	*p* value
Age (mean±SD)	39.6±11.5	44.3±14.3	0.31
Number of BAs (median, min--max)	L:2 (1-5)	L:1(1-2)	0.54
	R:1(1-5)	R:1(1-5)	
Diameter of BAs (median,min--max)	1.66 (0.70-7.96)	1.32 (0.70-3.54)	0.02
Frequency of hypertrophied BAs (%)	41.7%	18.2%	0.16

BA: bronchial artery, PA: pulmonary artery, PAI: pulmonary artery involvement

ness. Our results also illustrated a high rate of thoracic findings. This result may be because symptomatic cases are usually referred to the chest disease hospital where the study was conducted. Indeed, 81% of our patients showed pulmonary symptoms higher than that reported in previous studies [14, 15].

Among the various thoracic findings, PAI was the most prevalent in the present study. It is a cause of increased mortality and PAA is a typical manifestation of pulmonary involvement [2]. The underlying pathophysiological process of PAA is inflammation of the vasa vasorum in the tunica media, which destroys the elastic fibers of the media and causes dilatation of the lumen. PAA is not, however, the only form of PAI in BD [15]. PAT can also be detected in up to one-third of patients at presentation, which is *in situ* thrombosis rather than emboli [16]. Tunaci *et al*. claimed that PAT developed during the PAA regression process [17]. However, it remains controversial whether PAA precedes PAT.

PAA may be single or multiple, unilateral or bilateral, fusiform or saccular, and they are located in the main pulmonary arteries, lobar and segmental branches [1]. Tunacı *et al*.reported that PAA occurred mostly in the lobar arteries followed by the main pulmonary arteries [17]. PAI was detected mostly in peripheral PA branches in our series; in 77% of our patients, PAI was observed in the segmental-subsegmental branches. Advances in imaging techniques probably enabled us to identify more cases of involved peripheral PAs. In addition, PAI was predominant in the descending branches of pulmonary arteries, mostly bilateral and multiple; this finding was in accordance with the results of previous studies [15, 18].

In our series, peripheral small PAAs looked like pulmonary nodules in three patients. After reviewing the CTA images in more detail with reformatted images, they were recognized as fusiform PAAs. The lesions opacified with the contrast medium and showed continuation with PA branches. Because they were considered pulmonary nodules, these lesions were unnecessarily followed with CT scans and sometimes even with PET-CT for malignancy. Identification and reports of peripheral PAAs have increased recently owing to advances in diagnostic imaging modalities such as CT and MRI, but to our knowledge, this is the first study to report peripheral PAAs for BD [19-21]. Multidetector CT technology yields high-resolution angiographic studies complemented by high-quality reformatted images allowing the identification of the origin and course of the pulmonary arteries. Furthermore, CTA provides information regarding the size and location of the aneurysm as well as thrombus within the aneurysm which is useful in determining the need for intervention and optimum surgical approach [20, 22].

One patient showed multiple small branching opacities such as tree-in-bud, which were confirmed as small aneurysmatic dilatations of subsegmental PA branches on the reformatted images. This finding was previously reported and accepted as pulmonary involvement of the disease [23]. Suspicion of peripheral small PAA was low among radiologists, which was overlooked by the reporting radiologist and detected in retrospective or follow-up studies. We, therefore,

believe that small peripheral PAAs can look like pulmonary nodules or vascular tree-in-bud and require meticulous consideration for PAI. This aspect should be highlighted in nodules' follow-up criteria. Further studies are needed to confirm our results.

In our series, PAI was significantly correlated with parenchymal lesions and cardiovascular findings such as SVC thrombosis and intracardiac thrombus. Seyahi and Takeno *et al*. considered that parenchymal lesions, intracardiac filling defects, and pleural-pericardial effusion were more specific to PAI because they were rarely observed in patients without PAI [24, 25].

Isolated pulmonary parenchymal lesions without obvious PAA and PAT were considered as a microscopic vascular disease [26]. However, the pathological correlation of the parenchymal changes has been reported only in a few cases [27, 28]. Several studies have investigated the role of lung scintigraphy in patients with BD and suggested that microvascular changes could be shown with scintigraphic techniques even in the early stage of BD [29, 30].

We also demonstrated that the PAI group had a larger diameter of BAs and more frequent dilated BAs than the non--PAI group. Elevated PA pressure was noted in some patients with PAI and dilated BAs. Esatoglu *et al*. reported nine patients with PAI who had recurrent hemoptysis during the follow-up period but no relapse of PAI. The authors explained this observation with BA enlargement and showed effective treatment with embolization [31]. Kızıldağ *et al*. showed a close relationship between PAI and hypertrophied BAs [32]; they emphasized that dilated BAs were valuable clues for diagnosing PAI. Our results suggest that in cases of PAI, dilatation of BAs may be secondary to vasculitis of PA rather than being an outcome of vasculitis. We also assume that dilated BAs may indicate increased PA pressure in patients with PAI; hence, we propose investigating pulmonary hypertension in such patients. Furthermore, our results suggest that this finding is important in cases of hemoptysis that are nonresponsive to immunosuppressive treatment.

This study's major limitation was the retrospective study design. CT scanners and examination protocols were not uniform. The number of patients was small in BA investigation, particularly in subgroup analysis. Studies with larger cohorts are required to support our results. Furthermore, we mainly focused on CT findings of PAI. Consequently, we could not correlate cardiac and vascular findings with other imaging modalities such as echocardiography and MR imaging.

CONCLUSION

In conclusion, the present study provided a detailed description of CT characteristics of PAI in patients with BD. The involvement of peripheral PAs is being detected more frequently than previously with advances in CT technology. Small peripheral PAAs can resemble pulmonary nodules. Dilated BAs are observed along with PAI and this condition should be considered in patients with nonresponsive hemoptysis.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

This retrospective study was approved by the institutional review board (Keçiören Training and Research Hospital, Turkey. Date: June 27, 2018, Approval number: 2012-KAEK-15/1704).

HUMAN AND ANIMAL RIGHTS

No animals were used as the basis of this study. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

CONSENT FOR PUBLICATION

The manuscript has no individual' Data such as patient name.

AVAILABILITY OF DATA AND MATERIALS

Not applicable.

FUNDING

None.

CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

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DECLARATION

The study was presented at the ASYOD congress in 2019 as an oral presentation.

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