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Pulmonary involvement in Behcet's disease: Definition of Tc99m-MAA lung scintigraphy perfusion patterns according to the affected pulmonary vascular levels

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ABSTRACT

The aim of this study was to define lung perfusion scintigraphy findings in Behcet's disease (BD) patients with different variations of pulmonary involvement. Medical records and imaging findings of 23 patients with pulmonary involvement of BD were retrospectively reviewed. Before scintigraphic evaluation, patients were classified according to the affected pulmonary vascular level on CT angiography (CTA) as follows: Macroscopic pulmonary vascular involvement (Gr-1) [Pulmonary artery aneurysm without thrombosis (Gr-1a), pulmonary artery aneurysm with thrombosis (Gr-1b), pulmonary artery thrombosis without aneurysm (Gr-1c)]; microscopic pulmonary vascular involvement (Gr-2; no macroscopic CTA findings but with clinical diagnosis and/or scintigraphic abnormality). There were 18 patients in Gr-1 and five patients in Gr-2. Segmental/subsegmental perfusion defects were the most common perfusion pattern and no distinctive pattern was observed among all groups. In 12 patients with macroscopic disease and two patients with microscopic disease, perfusion defects were more extensive than involved vessels on CTA and/or also in the contralateral lung. There were 13 patients with scintigraphic follow-up findings. No change was observed in all Gr-1b and Gr-2 patients; there were heterogeneous changes in 6/7 patients in Gr-1c. No typical perfusion pattern could be demonstrated for a given macroscopic vascular category. In patients with microscopic disease, similarly no characteristic pattern could be defined. As almost all patients with microscopic disease showed perfusion anomalies, scintigraphy may be proposed as a first step examination in case of suspected pulmonary involvement in BD. Scintigraphic follow-up may be of value in pure thrombotic pulmonary involvement.

1. Introduction

Behcet's disease (BD) is a triad of recurrent oral and genital ulcers with relapsing uveitis and was first described in 1937 by Hulusi Behcet (Behcet, 1937). Skin, central nervous system, cardiovascular system, gastrointestinal tract and pulmonary system were defined as the main affected areas since its first description (Koç et al., 1992; Akpolat et al., 2002; Düzgün et al., 2002). Pulmonary involvement is a rare condition, usually represented by episodic hemoptysis. Pulmonary vessels may be affected macroscopically (pulmonary artery aneurysms and/or thrombosis) or microscopically (vasculitis of small sized vessels) in BD (Erkan et al., 2001; Uzun et al., 2005; Uzun et al., 2008; Uzun 2011). Differential diagnosis between these pulmonary artery pathologies is important, because clinical signs and symptoms were similar but the treatment and prognosis may be

different. CT angiography (CTA) and Technetium 99m-Macroaggregated Albumin lung perfusion scintigraphy (MAA lung scintigraphy) are used in the diagnosis and follow-up of pulmonary involvement. CTA may represent macroscopic pulmonary involvement, but has limited diagnostic value in small sized vessel vasculitis. MAA lung scintigraphy findings in pulmonary involvement of BD were presented in a limited number of studies (mostly case based) and as yet, no specific radiotracer accumulation pattern has been reported. The aim of this study was to define MAA lung scintigraphy findings in BD patients with different variations of pulmonary involvement.

2. Materials and methods

Medical records of 23 patients from 2006 to 2019 were retrospectively reviewed. Patient's data were reevaluated according to the criteria defined by International Study Group for BD (International Study Group for BD 1990). All patients underwent CTA and MAA lung scintigraphy with a maximum time interval of five days. Both CTA and MAA lung scintigraphy images were reevaluated. For MAA lung scintigraphy, 185 MBq of Tc-99m-MAA was injected intravenously. Standard anterior, posterior, right lateral, left lateral, right posterior oblique and left posterior oblique static images were obtained with either E-CAM (Siemens, USA) or Discovery NM 630 (General Electric, Hayfa, Israel) dual headed gamma camera equipped with high resolution collimators. Radiotracer uptake patterns on MAA lung scintigraphy images were evaluated visually in lung segments. CTA images were obtained either with X press/GX model TSX-002a (Toshiba, Tachigi-Ken) helical CT or Aquilion 16 system (Toshiba Medical Systems Corporation, Japan) multidetector CT scanner. Non-ionic intravenous contrast agent administered at a rate of 4 ml/s with a delay of 15–20 s before scanning. CTA images were obtained during suspended inspiration and were taken from the level of the aortic arch to 2 cm below the level of the diaphragm.

Patients were classified as follows according to the pathologies in the pulmonary arteries according to the CTA findings:

1. Macroscopic pulmonary vascular involvement (Gr-1)
   a. Pulmonary artery aneurysm without thrombosis (Gr-1a)
   b. Pulmonary artery aneurysm with thrombosis (Gr-1b)
   c. Pulmonary artery thrombosis without aneurysm (Gr-1c)

2. Microscopic pulmonary vascular involvement (Gr-2) (without any macroscopic CTA findings but having clinical diagnosis or MAA lung scintigraphy abnormality)

Immunosuppressive therapy was given to all patients and dose was adjusted according to the severity and extent of involved pulmonary arteries and also clinical parameters including massive hemoptysis. Most aggressive treatment with "pulse" corticosteroid (1-3 days) and cyclophosphamide were given to the patients with pulmonary artery aneurysm. Colchicine was used in all patients.

3. Results

MAA lung scintigraphy and CTA findings were reevaluated in 23 patients (17 males, 6 females). The mean age was 33±9 and the age range were 18-56. The most common symptom was oral ulceration (in 22 patients; 95.6%). Cough and hemoptysis were the most common pulmonary symptoms presented in 17 (73.9%) and 15 (65.2%) patients respectively. Pulmonary involvement was diagnosed simultaneously with BD in 17/23 patients (73.9%). During data analysis of this study, 22/23 (95.6%) patients were still alive, one patient was lost to follow-up after 47 months from the diagnosis of pulmonary involvement of BD. Patients were followed-up for a period of 107±46 months. Patient's symptoms and clinical features are presented in Table 1 and 2 respectively.

Table 1. Symptoms of 23 BD patients with pulmonary involvement.

Symptoms	**Number of patients (%)**
Oral ulceration	22 (95.6)
Genital ulceration	15 (65.2)
Ocular lesions	9 (39.1)
Skin lesions	10 (43.5)
Positive pathergy test	4 (17.4)
Pulmonary symptoms	
Cough	17 (73.9)
Hemoptysis	15 (65.2)
Hemoptysis (massive)	10 (39.1)
Dyspnea	11 (47.8)
Chest pain	10 (39.1)
Fever	12 (52.2)

CT angiography findings

Macroscopic pulmonary vascular involvement (Gr-1) was detected in 18 patients according to CTA findings; two patients had pulmonary artery aneurysm without thrombosis (Gr-1a), seven patients had pulmonary artery aneurysm with thrombosis (Gr-1b), nine patients had pulmonary artery thrombosis without aneurysm (Gr-1c). Microscopic pulmonary vascular involvement (Gr-2) was detected in five patients; four patients had normal findings and one patient had thinning of peripheral vascular branches on CTA.

Table 2. Clinical Features of 23 BD patients with pulmonary involvement.

Pt no	Age/ Sex	Presenting pulmonary symptom	BD and pulmonary involvement	Clinical follow-up (months)
1	36/M	Hemoptysis	15 yrs	142
2	33/F	Cough	4 yrs	152
3	21/F	Cough	2 yrs	62
4	49/M	Chest pain	Same time	78
5	56/M	Leg swelling	Same time	123
6	41/M	Hemoptysis*	Same time	141
7	34/M	Hemoptysis*	Same time	141
8	24/F	Hemoptysis*	Same time	52
9	24/M	Hemoptysis	Same time	57
10	36/M	Chest pain	Same time	102
11	40/M	Fever	5 yrs	118
12	32/M	Fever	Same time	130
13	41/M	Dyspnea	2 yrs	133
14	38/M	Fever	Same time	144
15	19/F	Dyspnea	Same time	153
16	42/M	Fever	Same time	150
17	23/M	Hemoptysis	Same time	164
18	29/M	Hemoptysis	Same time	24
19	32/F	Hemoptysis	Same time	47
20	34/M	Dyspnea	1 month	148
21	29/M	Hemoptysis*	Same time	31
22	18/F	Dyspnea	Same time	49
23	40/M	Chest pain	Same time	129

Perfusion lung scintigraphy findings

Group 1 (18 patients)

In Gr-1a (two patients) multiple segmental and subsegmental perfusion defects were detected in both right and left lungs in two patients. In Gr-1b (seven patients), multiple segmental and subsegmental perfusion defects in both right and left lungs in two patients and only in the lower lobe of right and/or left lung in four patients were determined. In Gr-1c (nine patients), multiple segmental and subsegmental perfusion defects in both right and left lungs in eight patients and only in the lower lobe of right and left lungs in one patient were observed. The extent of defective areas on MAA lung scintigraphy were larger and encompassed more segments than on CTA and/or perfusion defects in the contralateral lung with normal CTA findings were observed in one patient in Gr-1a, three patients in Gr-1b and seven patients in Gr-1c. MAA lung scintigraphy and CTA findings of a patient patient with pulmonary artery aneurysm without thrombosis (Gr-1a) were given in Fig.1 .

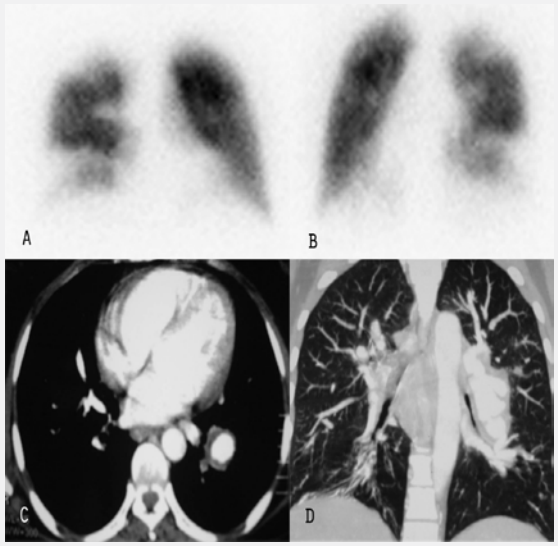

Fig. 1. Segmental and subsegmental perfusion defects in left and right lungs were observed on anterior (A) and posterior (B) perfusion lung scintigraphy images of a 33-year-old female patient. CT angiography of this patient shows non-occlusive huge aneurysmatic pulmonary arteries on axial (C) and coronal (D) images. This patient is an example of "pulmonary artery aneurysm without thrombosis" (Gr-1a). She received intense immunosuppressive treatment without anticoagulation. She is now asymptomatic after 10 years of first presentation with low-dose immunosuppressive after two exacerbation of pulmonary involvement when she was on remission without any treatment.

Group 2 (five patients)

Heterogeneous and decreased radiotracer accumulation in two patients, multiple segmental and subsegmental perfusion defects in two patients were observed. One patient had normal perfusion pattern.

MAA lung scintigraphy follow-up findings

There were 13/23 patients (56.5%) with MAA lung scintigraphy follow-up findings in a range of 1 to 153 months (seven patients in Gr-1c, three patients in Gr-1b and three patients in Gr-2). Perfusion findings were changed in the follow-up of 6/7 patients in Gr-1c. Perfusion changes were heterogeneous; there were newly formed and enlarged defects in some patients as well as patients with narrowed and completely resolved defective areas. A patient with multiple perfusion defects in both left and right lungs in Gr-1c had a completely normal follow-up MAA lung scintigraphy finding. Interestingly changes in follow-up studies were observed only in patients with pure thrombotic vascular involvement (Gr-1c). Fig. 2 represented MAA lung scintigraphy and CTA findings of a patient with newly formed and narrowed defects.

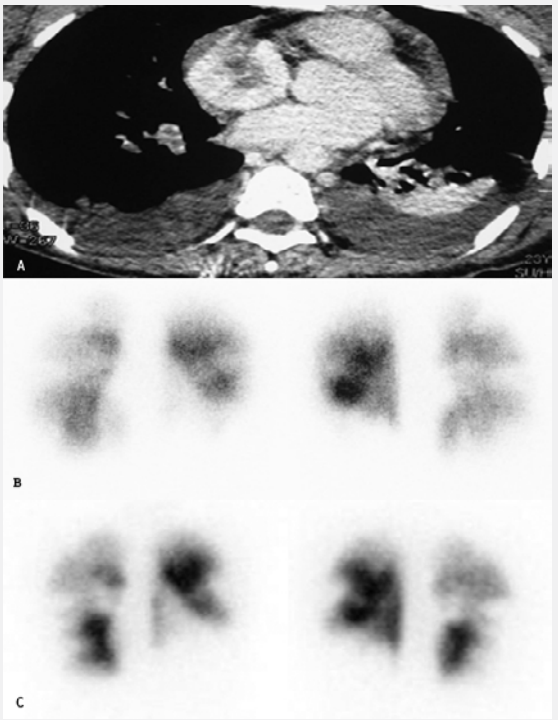

Fig. 2. On CT angiography of the 19 year-old female patient who presented with vena cava superior syndrome with severe dyspnea showed thrombotic occlusion of jugular veins, subclavian veins, vena cava superior, pulmonary arteries as well as right atrium (A). The patient went well without any symptom after 13 years of presentation with intense immunosuppressive treatment and anticoagulation lasting almost 5 years. Anterior and posterior perfusion lung scintigraphy images (B) of patient showed segmental and subsegmental perfusion defects in left and right lungs. After about 13 years of follow-up, newly formed and narrowed defects in the right and left lungs were determined (C). This patient is an example of "thrombotic pulmonary disease" (Gr-1b).

There was no change observed on MAA lung scintigraphy follow-up findings of all Gr-1b and Gr-2 patients. MAA lung scintigraphy follow-up and CTA images of a patient in Gr-2 were given in Fig. 3. Table 3 represented CTA, MAA lung scintigraphy and follow-up findings of patients.

4. Discussion

Technetium 99m-MAA lung scintigraphy has been reported to have high negative predictive value in terms of pulmonary involvement (Caglar et al., 2000). In this study, we classified patients as macroscopic or microscopic pulmonary vascular involvement and investigated the radiotracer accumulation patterns on MAA lung scintigraphy. To our knowledge, this was the first systematic study evaluating this issue.

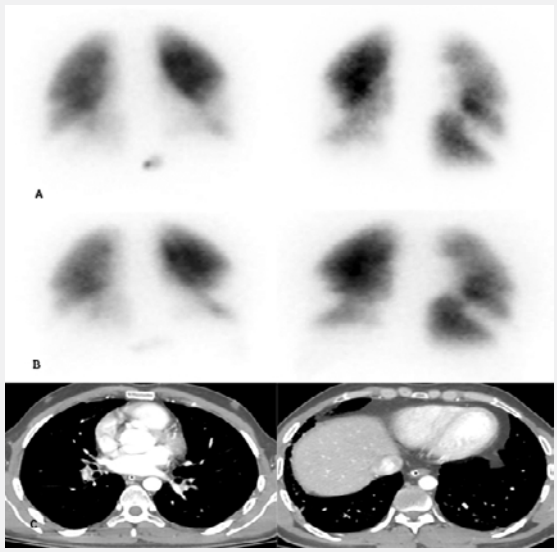

Fig. 3. Anterior and posterior perfusion lung scintigraphy images (A) of a 29-year-old male patient were revealed with multiple segmental and subsegmental perfusion defects in the lower lobe of left and right lungs. No significant change was observed on the perfusion lung scintigraphy images after 17 months of follow-up (B). CTA images displayed thinning in branches of the right and left lower lobe pulmonary artery without any obvious thrombotic lesion (C). This patient is an example of "macroscopic pulmonary vascular involvement" (Gr-2).

Although perfusion lung scintigraphy with 99mTc-MAA is the most frequently performed scintigraphic method in BD patients with pulmonary involvement, some other radionuclide agents were also used to demonstrate pulmonary vascular endothelial damage. Gumuser et al. showed faster clearance half time of 99mTc-DTPA radio-aerosols and decreased 99mTc-HMPAO lung clearance rate in BD patients. They reported the contribution of these findings to predict pulmonary involvement in the early stages of BD (Gumuser et al., 2008; Gumuser et al., 2011). In another study, prolonged lung retention of 123I-meta-iodobenzylguanidine found to be associated with disease severity (Unlu et al., 2001). In a small number of studies with PET tracers, it has been reported that increased accumulation of 18F-fluorodeoxyglucose demonstrates pulmonary artery inflammation (Denecke et al., 2007; Trad et al., 2013).

The results of our study showed that MAA lung scintigraphy alone cannot specify the affected vascular level in BD patients with pulmonary involvement. This was thought to be the result of vasculitis, vascular remodeling and in situ thrombosis causing nonspecific perfusion defects with the occlusion of pulmonary arteries on perfusion lung scintigraphy. A remarkable finding was that the affected vascular bed areas detected

Table 3. CT angiography, perfusion lung scintigraphy and follow up findings of patients.

Patient No	CT Angiography	Perfusion Lung Scintigraphy	Follow-up Perfusion Lung Scintigraphy	Time Interval (Months)
Group 1 (Macroscopic pulmonary vascular involvement)				
Group 1a: Pulmonary artery aneurysm without thrombosis				
1	Aneurysm in basal posterior branch of left pulmonary artery	*Multiple segmental and subsegmental perfusion defects in left and right lungs	-	-
2	Aneurysm in main pulmonary artery	Multiple segmental and subsegmental perfusion defects in left and right lungs	-	-
Group 1b: Pulmonary artery aneurysm with thrombosis				
3	Arterial aneurysm with thrombosis in right pulmonary artery	*Segmental and subsegmental perfusion defects in the lower lobe of left and right lungs	-	-
4	Arterial aneurysm with thrombosis in lower lobe branch of left pulmonary artery	Segmental perfusion defects in the lower lobe of left lungs	-	-
5	Arterial aneurysm with thrombosis in lower lobe branch of right pulmonary artery	Subsegmental perfusion defects in the lower lobe of right lung	Same findings	46
6	Arterial aneurysm with thrombosis in lower lobe branches of left and right pulmonary artery	*Multiple segmental and subsegmental perfusion defects in left and right lungs	Same findings	20
7	Arterial aneurysm with thrombosis in right pulmonary artery	*Multiple segmental and subsegmental perfusion defects in left and right lungs	Same findings	48
8	Arterial aneurysm with total thrombotic occlusion in left pulmonary artery	Perfusion defect in the lower lobe of left lung	-	-
9	Arterial aneurysm with thrombosis in lower lobe branches of left pulmonary artery	Subsegmental perfusion defects in the lower lobe of left lung	-	-
Group 1c: Pulmonary artery thrombosis without aneurysm				
10	Thrombotic occlusion in lingular branch of left pulmonary artery and laterobazal branch of right pulmonary artery	*Segmental and subsegmental perfusion defects in left and right lungs	-	-
11	Thrombotic occlusion in lower lobe branches of left and right pulmonary artery	*Multiple segmental and subsegmental perfusion defects in left and right lungs	-	-
12	Thrombotic occlusion in lower lobe branch of right pulmonary artery	*Perfusion defects in left and right lungs	Normal findings	126
13	Thrombotic occlusion in segmental branches of left and right pulmonary artery	Multiple segmental and subsegmental perfusion defects in left and right lungs	Defects with narrowing borders	119
14	Thrombosis in right ventricule, nodular density at right and left lower lobes	*Multiple segmental and subsegmental perfusion defects in left and right lungs	Expansion of defect borders in right lung	13
15	Thrombotic occlusion in lower lobe branches of left and right pulmonary artery	*Segmental and subsegmental perfusion defects in left and right lungs	Newly formed and narrowed defects	153
16	Thrombotic occlusion in lower lobe branches of left and right pulmonary artery	*Multiple segmental and subsegmental perfusion defects in left and right lungs	Expansion of defect borders	2
17	Thrombotic occlusion in left and right main pulmonary artery	Multiple segmental and subsegmental perfusion defects in left and right lungs	Newly formed defects	35
18	Thrombotic occlusion in lower lobe branches of left pulmonary artery	*Segmental and subsegmental perfusion defects in the lower lobe of left and right lungs	Same findings	1
Group 2 (Microscopic pulmonary vascular involvement)				
19	Normal	Normal perfusion	Same findings	41
20	Normal	Bilateral heterogenous and decreased perfusion	-	-
21	Thinning in branches of the right and left lower lobe pulmonary artery	Multiple segmental and subsegmental perfusion defects in the lower lobe of left and right lungs	Same findings	17
22	Normal	Bilateral heterogenous and decreased perfusion	-	-
23	Normal	Decreased perfusion in left lung; bilateral segmental and subsegmental perfusion defects	Same findings	31

*Patients with perfusion defects encompassed more segments than pathological vessels on CTA and/or in the contrlateral lung with normal CTA findings.

by scintigraphic evaluation were more extensive than pathological vessels detected by CTA in macroscopic pulmonary vascular disease. In some patients, perfusion defects were observed in contralateral lung with normal vessels on CTA which was probably due to microvascular changes not visible on CTA.

The detection of perfusion abnormality in patients with pulmonary artery aneurysm without thrombosis and in patients with microscopic pulmonary disease was the other important finding. For two decades CTA is widely accepted as a routine imaging tool serving in the amelioration of survival rates of pulmonary

involvement of BD. Hamuryudan et al. showed prominent survival advantage without any therapeutic change in pulmonary BD patients diagnosed by means of CTA after 1992 compared with the patients diagnosed previously (Hamuryudan et al., 2004). In a previous cumulative analysis of pulmonary BD patients, diagnosed and followed up primarily by CTA, 1 and 5-year survival rates of patients with pulmonary artery aneurysm were 57% and 39% respectively (Uzun et al., 2008). In the current study, no patient died during the mean follow-up period of 107±46 months and this finding supported the importance of early diagnosis of pulmonary involvement.

Caglar et al. previously mentioned that, scintigraphic evaluation was ideal for pulmonary involvement of BD patients and normal findings on perfusion lung scintigraphy ruled out pulmonary involvement with a 100% of negative predictive value (Caglar et al., 2000). Our findings mostly supported this detection, because 4/5 patients with microscopic vascular disease had perfusion abnormality on MAA lung scintigraphy. As microscopic involvement might be the earliest form of pulmonary BD, this was thought to be an important result of this study.

In most of the patients in Gr-1c, changes in the perfusion patterns in follow-up MAA lung scintigraphy were remarkable. There was no change observed in Gr-1b and Gr-2 patients on follow-up scintigraphy. We think that, newly formed or improved perfusion defects in the follow-up scintigraphy in Gr-1c patients supported the presence of an unorganized thrombus which represents an early disease and a dynamic process. The ongoing inflammation due to the activity of disease with the failure of compliance on treatment and/or inappropriate treatment caused newly formed defects and appropriate treatment might be the cause of improvement on MAA lung scintigraphy. Although this study was one of the largest case series evaluating MAA lung scintigraphy in pulmonary BD, we have some drawbacks. We have to remark that relatively low number of cases, retrospective evaluation of patients and unblinded nature of evaluation were weak aspects of this study. There was also no histopathological confirmation of pulmonary findings in this case series. This was mostly related to difficulties to perform a biopsy procedure in these severely bleeding cases and ethical issues in the less severe non-aneurysmatic cases who all need urgent immunosuppressive and anticoagulation therapy.

In conclusion, MAA lung scintigraphy findings alone could not differentiate macroscopic and microscopic pulmonary involvement in BD. More extensive vascular involvement was detected on MAA lung scintigraphy in patients with macroscopic pulmonary disease and could be additionally performed to determine the extent of the affected lung area in these patients. Although more patients with pulmonary thrombosis without aneurysm were diagnosed with CTA and longer survival rates had been achieved, we need prospective studies to accurately demonstrate microscopic vascular disease which might be an early form and precursor of thrombotic and aneurysmatic pulmonary vascular disease. In most of the patients with microscopic vascular involvement, multiple perfusion defects or decreased/heterogenous perfusion were observed on MAA lung scintigraphy and this technique can be used as an early diagnostic method in patients with pulmonary symptoms and normal CTA. Follow up studies with MAA scans may be of value in pure thrombotic (Gr-1c) pulmonary involvement, which necessitates further detailed investigation. As CTA provides detailed morphological information about pulmonary vascular pathologies and MAA lung scintigraphy gives additional findings in the evaluation of both macroscopic and microscopic pulmonary disease, these two diagnostic tests can be considered as complementary. However, since almost all patients with microscopic disease and normal CTA showed MAA scan anomalies, perfusion scintigraphy may be proposed as the first step examination in case of suspected pulmonary involvement in BD.

Conflict of interest

There is no conflict of interest to declare.

Ethical approval

The study was approved by the Ethics Committee of Ondokuz Mayıs University hospital (date 27.06.2019, No. 2019/504). The study was conducted in accordance with the principles of the Declaration of Helsinki.

REFERENCES

Akpolat, T., Akkoyunlu, M., Akpolat, I., Dilek, M., Odabas, A.R., Ozen, S., 2002. Renal Behcet's disease: A cumulative analysis. Semin. Arthritis Rheum. 31, 317-337.

Behcet, H., 1937. Uber rezidivierende aphthose, durch ein virus verursachte Geschwure am Mund, am Auge und an den Genitalien. Dermatol. Woschenschr. 105, 1152-1157.

Caglar, M., Ergun, E., Emri, S., 2000. 99Tcm-MAA lung scintigraphy in patients with Behçet's disease: Its value and correlation with clinical course and other diagnostic modalities. Nucl. Med. Commun. 21, 171-179.

Denecke, T., Staeck, O., Amthauer, H., Hänninen, E.L., 2007. PET/CT visualises inflammatory activity of pulmonary artery aneurysms in Behçet disease. Eur. J. Nucl. Med. Mol. Imaging. 34, 970.

Duzgun, N., Anil, C., Ozer, F., Acican, T., 2002. The disappearance of pulmonary artery aneurysms and intracardiac thrombus with immunosuppressive treatment in a patient with Behcet's disease. Clin. Exp. Rheumatol. 20, 56-57.

Erkan, F., Gül, A., Tasali, E., 2001. Pulmonary manifestations of Behçet's disease. Thorax. 56, 572-578.

Gumuser, F.G., Pirildar, T., Batok, D., Sakar, A., Ruksen, E., Sayit, E., 2008. Assessment of alveolar epithelial permeability in Behçet's disease with 99mTc-DTPA aerosol scintigraphy. Ann. Nucl. Med. 22, 349-355.

Gumuser, G., Pırıldar, T., Tarhan, S., Batok, D., Ruksen, E., Sakar, A., Sayit, E., 2011. Technetium-99m-hexamethylpropylene amine oxime lung scintigraphy findings in patients with Behçet's disease. Nucl. Med. Commun. 32, 363-368.

Hamuryudan, V., Er, T., Seyahi, S., Akman, C., Tüzün, H., Fresko, I., Yurdakul, S., Numan, F., Yazici, H., 2004. Pulmonary artery aneurysm in Behçet syndrome. AMJ. 117, 867-870.

International Study Group for Behçet's disease,1990. Criteria for diagnosis of Behçet's disease. Lancet. 335, 1078-1080.

Koç, Y., Güllü, I., Akpek,G., Akpolat, T., Kansu, E., Kiraz, S., Batman, F., Kansu, T., Balkanci, F., Akkaya, S., 1992. Vascular involvement in Behcet's disease. J. Rheumatol. 19, 402-410.

Trad, S., Bensimhon, L., Hajjam, M.E., Chinet, T., Wechsler, B., Saadoun, D., 2013. 18F-fluorodeoxyglucose-positron emission tomography scanning is a useful tool for therapy evaluation of arterial aneurysm in Behçet's disease. Joint Bone Spine. 80, 420-423.

Unlu, M., Akıcıoglu, C., Yamac, K., Onder, M., 2001. Pulmonary involvement in Behçet's disease: Evaluation of 123 I-MIBG retention. Nucl. Med. Commun. 22, 1083-1088.

Uzun, O., Akpolat, T., Erkan, L., 2005. Pulmonary vasculitis in Behcet disease: A cumulative analysis. Chest. 127, 2243-2253.

Uzun, O., Erkan, L., Akpolat, I., Findik, S., Atıcı, A.G., Akpolat, T., 2008. Pulmonary involvement in Behçet's disease. Respiration. 75, 310-321.

Uzun, O., 2011. Pulmonary involvement in Behçet's disease and Takayasu's arteritis. Eur. Respir. Mon. 54, 32-45.