Vascular involvement in Behçet’s disease: Imaging features

Ödev K*, Varol S and Tunç R

1Konya Chamber of Commerce Karatay University, Faculty of Medicine, Konya, Turkey
2Necmettin Erbakan University, Faculty of Medicine, Konya, Turkey

Abstract

Behçet’s disease is multisystemic and chronic inflammatory disorder with an unknown cause. The clinical triad of oral and genital ulcerations and ocular manifestation was originally described by the Turkish dermatologist Hulusi Behçet in 1937 [1]. Additional clinical manifestation in other locations (skin,joints,gastrointestinal tract,genitourinary tract,central nervous system,cardiovascular system,lung) were described later [2]. The variability of clinical manifestations and the absence of specific histologic or laboratory findings may cause the difficulty in diagnosis. The diagnosis is made on the basis of the criteria published in 1990 by the International Study Group for Behçet’s disease BD [3]. The main underlying pathologic process in Behçet’s disease is vasculitis and perivascular inflammatory infiltrates affecting vessels of different size in various organs. Several types of thoracic involvement associated with Behçet’s disease have been described.

Introduction

Behçet’s disease is multisystemic and chronic inflammatory disorder with an unknown cause. The clinical triad of oral and genital ulcerations and ocular manifestation was originally described by the Turkish dermatologist Hulusi Behçet in 1937 [1]. Additional clinical manifestation in other locations (skin,joints,gastrointestinal tract,genitourinary tract,central nervous system,cardiovascular system,lung) were described later [2]. The variability of clinical manifestations and the absence of specific histologic or laboratory findings may cause the difficulty in diagnosis. The diagnosis is made on the basis of the criteria published in 1990 by the International Study Group for Behçet’s disease BD [3]. The main underlying pathologic process in Behçet’s disease is vasculitis and perivascular inflammatory infiltrates affecting vessels of different size in various organs [4]. Several types of thoracic involvement associated with Behçet’s disease have been described.

This study focuses on the thoracic vascular system involvement of Behçet’s disease. We describe thoracic involvement for one of the most serious aspects of Behçet’s disease: Vascular involvement Superior vena cava syndrome, pulmoner parenchmal and mediastinal involvement in the disease.

Involvement of thoracic vessels

Vascular manifestations of BD, which consist of venous involvement (thrombosis, superficial thrombophlebitis), and arterial involvement (aneurysm, stenosis, occlusion), have been added into the update International Criteria for Behçet’s Disease, since they are one of the major characteristics of BD [5]. The entire arterial tree can be involved in BD, with aneurysm or pseudoaneurysm or thrombosis, pulmonary infarction, pulmonary hemorrhage and pulmonary artery aneurysm. Contrast enhanced helical CT or multislice CT (MSCT) (especially CT angiography) as a noninvasive and sensitive method for detecting aneurysm has been pointed out in many papers [2, 4, 6]. Systemic arterial manifestations of BD are infrequent compared with venous involvement. According to the frequency of arterial vascular involvement is reported in 85% venous, 10% arterial and 5% combined arterial and venous involvement [7]. Pulmonary artery aneurysms (PAAs) are the most common type of pulmonary involvement in BD. Hilar enlargement or the appearance of polylobular and round opacities on the chest radiograph or thorax CT can present pulmonary artery aneurysms (Figure 1) [2, 6]. Thrombosis of the pulmonary arteries in BD is usually in situ thrombosis. Although deep vein thrombosis is common in BD, pulmonary embolism is rare because the thrombi in the inflamed veins of the lower extremities are strongly adherent [8]. The association of hemoptysis with dyspne and pleuritic chest pain is suggestive of pulmonary infarction secondary thromboembolism [2, 8, 9]. Hemoptysis of varying degrees is the most common clinical symptom of pulmonary artery aneurysms. It may be life threatening or fatal [2].

Involvement of the superior vena cava and major mediastinal veins

Superficial thrombophlebitis and deep vein thrombophlebitis are the most frequent venous manifestations. Involvement of the venous system is most frequently seen in the form of thrombophlebitis. Thrombophlebitis can affects veins of the lower extremity, SVC and inferior vena cava [10]. The vascular lesion occurs in the venous system (18-24%) more frequently than the arterial system (7%) [11]. The most serious complications in BD is superior vena cava (SVC) syndrome and Budd Chiari syndrome [12]. Following malignancy, BD is the commonest cause of superior vena cava syndrome in Mediterranean countries [9, 12]. Helical CT or multislice CT with 3D volume rendering can document the obstruction of the SVC or major mediastinal veins and the presence enlarged collateral vessels in the mediastium and chest wall (Figure 2) [6, 13]. Recent reports suggest that 3D gadolinium

*Correspondence to: Kemal Ödev, Konya Chamber of Commerce Karatay University, Faculty of Medicine, Akabe Mah, Karatay/Konya Turkey, E-mail: kemalodev50@yahoo.com

Key words: Behçet’s disease, Lung, CT, MR angiography, MR venography

Received: April 01, 2019; Accepted: April 08, 2019; Published: April 10, 2019
Involvement of the pulmonary parenchyma

Involvement of Behçet disease in the pulmonary parenchyma and pleura is seen in 1-10% of patients during the course of Behçet's disease [13,16]. On the plain chest film or thorax CT visible abnormalities include air-space consolidation, pulmonary infarcts, pulmonary hemorrhage, atelectasis, cryptogenic organizing pneumonia transient opacities and excavated nodul. These radiological abnormalities are nonspecific [2,4].

Involvement of the heart and mediastinum

Intracardiac thrombosis is a rare but serious complication of Behçet's disease [6]. In a case with Behçet's disease pulmonary artery aneurysm accompanied with intracardiac thrombosis and in the other case coronary artery aneurysm associated with PAA have been reported [17]. The other pulmonary complication of BD is mediastinal fibrosis in association with occlusion of SVC. This complicated clinical entity has rarely been described [12,18]. Because radiologic features of mediastinal involvement with SVC syndrome in BD are similar to those of benign and malignant conditions, such as radiation therapy, trauma, tuberculosis, fungal infections, rheumatic fever and neoplasms. However, these clinical entities must be clinically differentiated [18].

References

1. Behçet H (1937) Über rezidivierende, aphthose, durchein Virus verursachte Gaschwure am Mund, am Auge und an den Genitalien. Dermatol Wochenschr 105: 1152-1157.
2. Erkan F, Gül A, Tasali E (2002) Pulmonary manifestations of Behçet's disease. Thorax 23: 493-503.
3. [No authors listed] (1990) Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet 335: 1078-1080. [Crossref]
4. Hiller N, Lieberman S, Chajek-Shaul T, Bar-Ziv J, Shaham D (2004) Thoracic vascular involvements in Behcet's syndrome: a report of 24 cases. J Eur Acad Dermatol Venereol 28: 338-347. [Crossref]
5. Duvatchi F, Assaad-Khalil S, Calamia KT, Crook JE, Sadeghi-Abdollahi, et al. (2014) The International Criteria for Behçet’s Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatol Venereol 28: 338-347. [Crossref]
6. Erkan F (1999) Pulmonary involvement in Behçet disease. Curr Opin Pulm Med 5: 314-318.
7. Kabbaj N, Benjelloun G, Gueddari FZ, Dafri R, Imani F (1993) [Vascular involvements in Behçet disease. Based on 40 patient records]. J Radiol 74: 649-656. [Crossref]
8. Dureux P, Bleyt O, Huchon G, Wechsler B, Chretien J, et al. (1981) Multiple pulmonary arterial aneurysms in Behçet's disease and Hughes-Stovin syndrome. Am J Med 71: 736-741. [Crossref]
9. Hamuryudan V, Yurdakul S, Moral F, Numan F, Tüzün H, et al. (1994) Pulmonary arterial aneurysms in Behçet's syndrome: a report of 24 cases. Br J Rheumatol 33: 48-51. [Crossref]
10. Sağdic K, Ozer ZG, Saba D, Ture M, Cengiz M (1996) Venous lesions in Behçet's disease. Eur J Vasc Endovasc Surg 11: 437-440. [Crossref]
11. El-Ramani KM, Al-Dalaan A, Al-Bala A (1993) Vascular involvement in Behçet's disease.In: Wechsler B, Godeau P eds. Behçet’s disease. Amsterdam: Excerpta Medica 531.
12. Dündar S,Yazici H (1984) Superior Vena Cava Syndrome in Behçet's Disease. Vascular Surg 18:29-30.
13. Ceylan N, Bayraktaroglu S, Erturk SM, Savas R, Alper H (2010) Pulmonary and vascular manifestations of Behçet disease: imaging findings. *AJR Am J Roentgenol* 194: W158-W164. [Crossref]

14. Shinde TS, Lee VS, Rofsky NM, Krinsky GA, Weinreb JC (1999) Three-dimensional gadolinium-enhanced MR venographic evaluation of patency of central veins in the thorax: initial experience. *Radiology* 213: 555-560. [Crossref]

15. Hansen ME, Spritzer CE, Sostman HD (1990) Assessing the patency of mediastinal and thoracic inlet veins: value of MR imaging. *AJR Am J Roentgenol* 155: 1177-1182. [Crossref]

16. Fairley C, Wilson JW, Bartraclough D (1989) Pulmonary involvement in Behçet's syndrome. *Chest* 96: 1428-1429. [Crossref]

17. Yakut ZI, Ödev K (2007) Pulmonary and cardiac involvement in Behçet disease: 3 case reports. *Clin Appl Thromb Hemost* 13: 318-322. [Crossref]

18. Harman M, Sayarlıoğlu M, Arslan H, Ayakta H, Harman E (2003) Fibrosing mediastinitis and thrombosis of superior vena cava associated with Behçet's disease. *Eur J Radiol* 48: 209-212. [Crossref]