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Case Report

Pulmonary Artery Aneurysm-Related Fatal Hemoptysis in Behcet's Disease: A Case Report with Literature Review

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Abstract

Introduction

Behcet's disease is a multisystemic disorder of unknown etiology. Vascular involvement is identified by the development of aneurysms and occlusion of the vessels. The current report aimed to discuss a case of Behcet disease presented with a bilateral pulmonary aneurysm.

Case presentation

A 41-year-old woman presented with recurrent attacks of hemoptysis for two years. A chest computed tomography scan revealed bilateral pulmonary artery aneurysm. Repeated bronchoscopy showed bleeding from the right lower lobe bronchus. She underwent a right-side thoracotomy with a right lower lobectomy. She was scheduled for left-side intervention as well. Before discharge, she was seen by a rheumatologist and was diagnosed with a case of Behcet disease. She developed attacks of massive hemoptysis (huge and bright blood) after two weeks which led to her passing away.

Conclusion

Although it is rare, Behcet disease could involve major arteries and lead to fatal complications. Surgical intervention remained a viable option for the management.

1. Introduction

Behcet's disease is an inflammatory chronic relapsing disorder with an unknown cause. It is characterized by oral and genital ulcers, as well as uveitis, (the original three symptoms). Behcet's disease is a multisystem disease with vasculitis as the primary pathological finding. It is usually a clinical diagnosis [1]. A pulmonary artery aneurysm is a rare pulmonary vasculature abnormality. They are a very rare finding, with an estimated incidence of 1 in 14000 [2].

Pulmonary artery aneurysms, arterial and venous thrombosis, pulmonary infarctions, recurrent pneumonia, pleurisy, and mediastinal masses are the main features of pulmonary involvement of Behcet's disease, and mostly young men are affected [3]. Venous thrombosis is one of the earliest manifestations of Behcet's disease. Arterial involvement in Behcet's disease is regarded as an unusual finding [4]. It occurs in 1–10% of the patients with Behcet's disease. A pulmonary artery aneurysm may be bilateral and may also rupture causing

sudden death due to massive hemorrhage or hemoptysis from an arteriobronchial fistula [5].

In this report, we described a patient whose right lung was the source of the bleeding which was later determined to be a case of Behcet's disease.

2. Case Presentation

2.1. Patient information and Clinical findings

A 41-year-old woman presented with recurrent attacks of hemoptysis for two years. The amount of hemoptysis had increased in the last two months, and the patient had developed cough and dyspnea. On further questioning, she reported repeated oral and vaginal ulcers.

2.2. Diagnostic assessment

The results of several medical checkups, including a chest computed tomography (CT) scan and bronchoscopy, were normal. Following an increase in hemoptysis, a new CT scan revealed a bilateral pulmonary artery aneurysm (Figure 1 & 2). Repeated bronchoscopy showed bleeding from the right lower lobe bronchus.

2.3. Therapeutic intervention

She underwent a right-side thoracotomy with a right lower lobectomy and was then discharged. She was scheduled for left-side intervention as well. Before discharge, she was seen by a rheumatologist and was diagnosed with a case of Behcet disease.

She was given a prednisolone tablet of 5 mg X2 with antibiotics and analgesia. She developed attacks of massive hemoptysis (huge and bright blood) after two weeks which led to her passing away. The family did not consent for the autopsy examination.

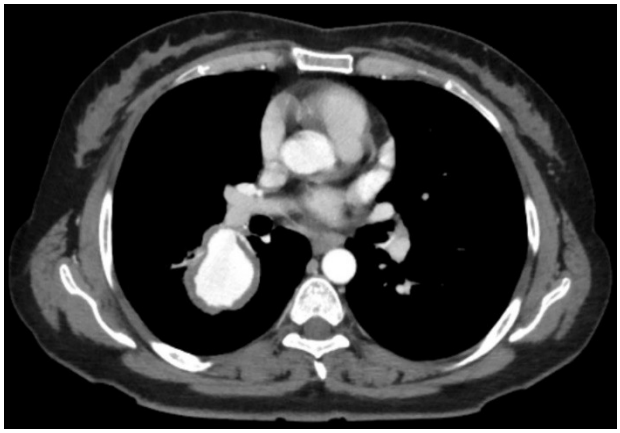

Figure 1: Computed tomography scan of the chest with IV contrast (axial section, mediastinal window) showing aneurysm of the right lower lobe pulmonary artery.

4. Discussion

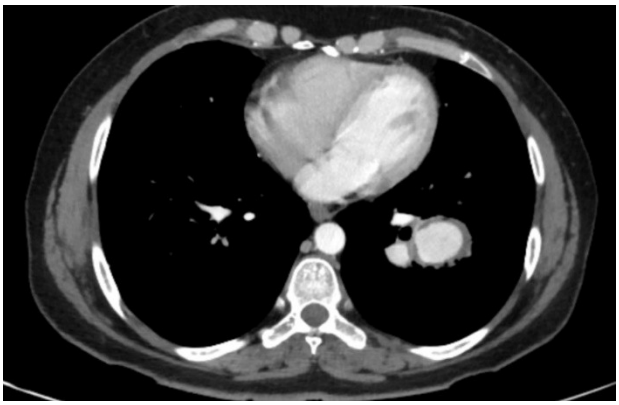

Figure 2: Computed tomography scan of the chest with IV contrast (axial section, mediastinal window) showing aneurysm of the left lower lobe pulmonary artery.

Behcet's disease is a common condition in the Middle East. Behcet's disease was first described in 1937 by Turkish dermatologist Hulusi Behcet [5]. The disease's primary symptoms differ based on the organs affected, including the skin, mucosa, eyes, joints, gastrointestinal tract, vascular system and nervous system. For a very long time, thrombophlebitis had been the known vascular complication of Behcet's disease. However, several reports describing aneurysm and arterial occlusion cases have been published recently [6]. The presence of HLA-B51.3 is closely related to the disease. The definitive diagnosis is based on the identification of several of its most prevalent clinical symptoms and signs as no specific diagnostic test is available [7].

A lung mass attributed to a pulmonary artery aneurysm is one of the most frequent findings of Behcet's disease at chest radiography. The pulmonary artery is the second most common site of arterial involvement, with the aorta being the most common [1]. Pulmonary arterial involvement usually manifests as hemoptysis caused by the rupture of an aneurysm or by thrombosis of pulmonary vessels [7]. The right lower lobar artery and the right and left main pulmonary arteries are where pulmonary aneurysms occur most frequently. Aneurysm diameters vary, and bilateral involvement is not uncommon [1]. Among the vasculitides, Behget's disease is virtually unique as a common cause of fatal pulmonary arterial tree aneurysms [8]. Unfortunately, aneurysms of the pulmonary arteries are a clear sign of poor prognosis and more than half of Behcet's patients with pulmonary artery aneurysms die from pulmonary hemorrhage within three years or that may heal with fibrosis [6]. In the current report, a 41-year-old female patient died after experiencing recurrent hemoptysis for two years.

Early diagnosis of an aneurysm in Behcet's disease is essential due to the potentially fatal complications [7]. The preferred method for documenting pulmonary arterial involvement is intravenous digital subtraction angiography. The presumptive diagnosis of occluded aneurysms is best accomplished with magnetic resonance imaging (MRI) [6]. All patients were routinely followed up at 3-month intervals when Kalko and associates published their experience with 18 arterial aneurysms. Computed tomography scan was used to perform the follow-up evaluation of patients with abdominal aortic and iliac aneurysms. The remaining patients underwent physical

examinations to check for new aneurysm formation and graft patency. If suspicion was present, angiography was used for further investigations [7].

In the current case, a final chest CT scan revealed bilateral pulmonary artery aneurysms. The bleeding was discovered to be coming from the right lower lobe of the chest during bronchoscopy. Gebitekin et al., also reported cases of bilateral aneurysms in their 1997 study. Alternatively, during pulmonary angiography, a CT of the thorax confirmed a bilateral pulmonary artery aneurysm and significant bleeding was found during bronchoscopy from the left lower lobe bronchus [5].

Various therapies with different results have been described. The severity of the hemoptysis and the number of aneurysms are the key factors in selecting a therapy. Immunosuppressive drugs alone or in combination with steroids are most beneficial when given in the early stages before irreversible damage to the arterial wall develops [6]. Due to technical difficulties and the development of a new postoperative false aneurysm, treating Behçet aneurysms surgically is difficult for vascular surgeons. Since medical treatment for aneurysmal lesions is ineffective, surgical intervention is required. Some authors suggested performing the normal arterial segments during the operation or preferred extra-anatomic bypass procedures to avoid anastomotic complications [7]. In cases with bilateral and multiple aneurysms, transcatheter embolization of a pulmonary artery segment should be tried if hemoptysis continues. Surgery must be the first course of action if the embolization procedure is unsuccessful. In the presence of this potentially fatal symptom, the source of the bleeding must be identified [6].

5. Conclusion

Although it is rare, Behcet disease could involve major arteries and lead to fatal complications. Surgical intervention remained a viable option for the management after embolization.

Declarations

Conflicts of interest: The author(s) have no conflicts of interest to disclose.

Ethical approval: Not applicable.

Patient consent (participation and publication): Consent has been taken from the patients and the family of the patients and the patient gave consent for the publication of the report.

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Authors' contributions: SAM participated in data collection. FHK designed the study. DMH, RKA participated in preparing the manuscript. DHB and BHA critically revised the manuscript. RQS, FHK confirmed the authenticity of the data. SHT and RJR The radiologists performing the assessment case. All authors approved the final version of the manuscript.

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References

1. Chae EJ, Do K-H, Seo JB, Park SH, Kang J-W, Jang YM, et al. Radiologic and clinical findings of Behçet disease: comprehensive review of multisystemic involvement. Radiographics. 2008;28(5):e31. doi:10.1148/rg.e31
2. Gupta M, Agrawal A, Iakovou A, Cohen S, Shah R, Talwar A. Pulmonary artery aneurysm: a review. Pulmonary Circulation. 2020;10(1):2045894020908780. doi:10.1177/2045894020908780
3. Cohle SD, Colby T. Fatal hemoptysis from Behcet's disease in a child. Cardiovascular pathology. 2002;11(5):296-9. doi:10.1016/S1054-8807(02)00117-5
4. Gouëffic Y, Pistorius M-A, Heymann M-F, Chaillou P, Patra P. Association of aneurysmal and occlusive lesions in Behçet's disease. Annals of vascular surgery. 2005;19(2):276-9. doi:10.1007/s10016-004-0179-2
5. Gebitekin C, Yılmaz M, Şenkaya I, Saba D, Sağdıç K, Özer G. Fatal haemoptysis due to pulmonary artery aneurysm in Behçet's disease. 1997. doi:10.1016/S1078-5884(97)80027-5
6. Tüzün H, Hamuryudan V, Yildirim S, Beşirli K, Yörük Y, Yurdakul S, et al. Surgical therapy of pulmonary arterial aneurysms in Behçet's syndrome. The Annals of thoracic surgery. 1996;61(2):733-5. doi:N/A
7. Kalko Y, Basaran M, Aydin U, Kafa U, Basaranoglu G, Yasar T. The surgical treatment of arterial aneurysms in Behçet disease: a report of 16 patients. Journal of Vascular Surgery. 2005;42(4):673-7. doi:10.1016/j.jvs.2005.05.057
8. Secrets t. Behçet's disease and cogan's syndrome. Rheumatology Secrets E-Book. 2014:248. doi:N/A