Tension Pneumothorax in a Patient with Behçet's Disease: A Rare Condition Managed with Surgery

Behçet Hastalığı Olan Bir Hastada Tansiyon Pnömotoraks: Cerrahi Girişimle Yönetilen Nadir Bir Durum

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Abstract

Behçet's disease is a chronic multisystemic condition characterized by systemic vasculitis in which pulmonary involvement is uncommon, and pneumothorax is a particularly rare complication. We describe here the case of a 53-year-old male with a known history of Behçet's disease who presented with acute chest pain and dyspnea. Chest X-ray revealed a left-sided tension pneumothorax. Thoracic CT revealed large apical bullae, and the patient subsequently underwent video-assisted thoracoscopic surgery (VATS) with wedge resection and partial pleural decortication. Histopathological evaluation was consistent with bullous emphysema and no evidence of vasculitis was observed. This case illustrates that tension pneumothorax can, in rare cases, develop in patients with Behçet's disease, even in the absence of pulmonary vasculitis. VATS may be considered as a safe and effective treatment option in appropriately selected cases.

Keywords: *Behçet's Disease, Pneumothorax, Tension Pneumothorax, VATS, Surgical Management.*

Öz

Behçet hastalığı, sistemik vaskülit ile karakterize ve birçok organ sistemini etkileyebilen kronik bir hastalıktır. Pulmoner tutulum nadir görülür; pnömotoraks ise son derece ender bir komplikasyondur. Burada, ani başlayan göğüs ağrısı ve nefes darlığı ile başvuran, akciğer grafisinde tansiyon pnömotoraks saptanan 53 yaşındaki Behçet hastalığı tanılı bir erkek olguyu sunmaktayız. Hastaya, acil tüp torakostomi uygulanmış, toraks BT'de dev büller saptanması üzerine video yardımlı torakoskopik cerrahi (VATS) ile wedge rezeksiyon ve parsiyel dekortikasyon yapılmıştır. Patoloji sonucu büllöz amfizem ile uyumlu idi ve vaskülit bulgusu saptanmadı. Olgumuz, Behçet hastalarında nadir görülen ancak hayatı tehdit edebilen tansiyon pnömotoraksın, parankimal vaskülit olmadan da gelişebileceğini göstermesi açısından özgündür. VATS, uygun seçilmiş olgularda güvenli ve etkili bir tedavi seçeneği olabilir.

Anahtar Kelimeler: *Behçet Hastalığı, Pnömotoraks, Tansiyon Pnömotoraks, VATS, Cerrahi Tedavi.*

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Although pulmonary involvement in Behçet's disease is rare, it ranks among the most severe and life-threatening complications. The most commonly encountered pulmonary manifestations include pulmonary artery aneurysms (PAAs), pulmonary artery thrombosis, pulmonary infarction, alveolar hemorrhage, pleural effusion, bronchiectasis and, more rarely, pneumothorax. Among these, PAAs can lead to massive hemoptysis and represent the most fatal complication of the disease, often determining the overall prognosis. For diagnosis, thoracic CT angiography and magnetic resonance imaging (MRI) are preferred due to their effectiveness in evaluating aneurysms, thrombosis and parenchymal involvement (1–3). In cases of massive hemoptysis or aneurysms refractory to medical treatment, endovascular embolization (e.g., coil or stent placement) or surgical interventions (lobectomy or segmentectomy) may be required; however, surgery carries high risk and is generally reserved as a last resort (4–6). Pneumothorax is a particularly rare manifestation in Behçet's disease and is typically associated with parenchymal destruction or cavitation. Cases of pneumothorax requiring surgical intervention have been reported in the literature, although most have involved lobectomy or procedures targeting cavitary lesions (4). We present here a case of tension pneumothorax requiring surgical management in a patient with Behçet's disease, and describe the clinical course and treatment approach.

CASE

A 53-year-old male patient presented to the emergency department with sudden-onset chest pain and dyspnea. On physical examination, the trachea was deviated to the right, breath sounds were diminished over the left hemithorax and percussion revealed hyperresonance. Blood pressure was 120/75 mmHg, and peripheral oxygen saturation was measured at 86%. Chest radiograph revealed a left-sided tension pneumothorax, prompting immediate tube thoracostomy (Figure 1 and 2). The patient was hospitalized and admitted to the thoracic surgery unit for further management. His medical history revealed a diagnosis of Behçet's disease, for which he had been taking oral prednisolone and colchicine for the past 10 years. Laboratory tests showed WBC: 12.84 K/uL, Hgb: 13.8 g/dL and Plt: 311 K/uL, while biochemistry and coagulation parameters were within normal limits. Thoracic CT revealed bullous structures measuring up to 5.5 cm in diameter in the left upper lobe (Figure 3).

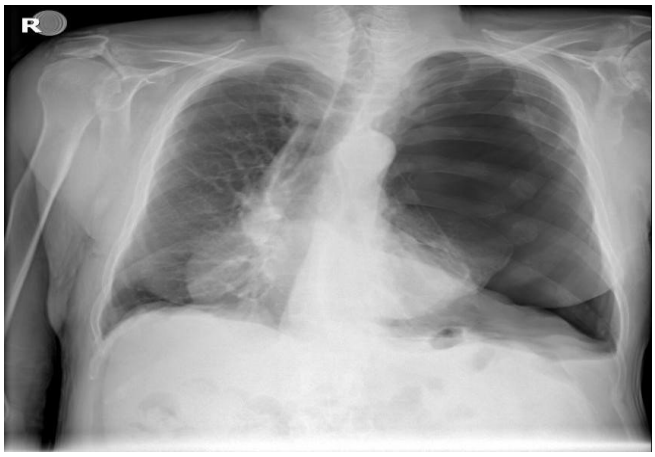

Figure 1: Left-sided tension pneumothorax on posteroanterior chest radiograph, with marked hyperlucency in the left hemithorax, consistent with a large volume of intrapleural air. The left lung is totally collapsed, and the mediastinal structures, including the trachea and cardiac silhouette, are shifted to the right, diagnostic of left-sided tension pneumothorax.

The patient had no history of smoking or occupational exposure that could predispose to bullous lung disease. Thoracic CT scans obtained 15 years earlier showed no evidence of bullous changes or emphysema, indicating the later development of the bullous formations. Following the placement of a chest tube for tension pneumothorax, a persistent and significant air leak continued for 4 consecutive days. Although 4 days may be considered a relatively short observation period, the severity of the air leak, coupled with the presence of giant bullae, prompted the decision for surgical intervention. In addition, the patient had been under chronic immunosuppressive therapy for Behçet's disease, increasing the risk of infection and impaired healing. Early surgical intervention was thus considered the safer and more appropriate approach.

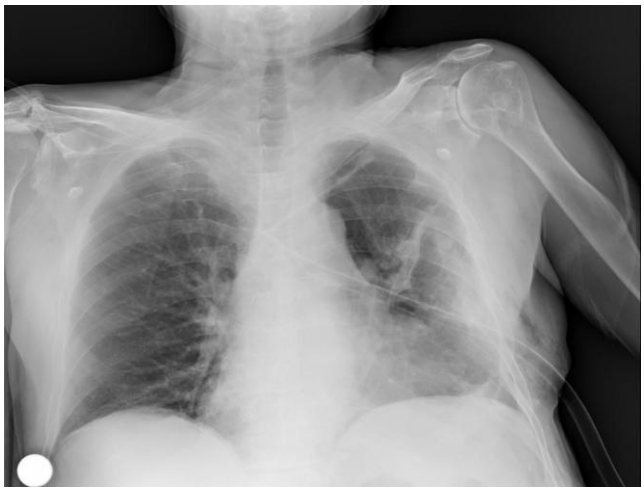

Figure 2: Anteroposterior chest radiograph following emergency tube thoracostomy. The partial re-expansion of the left lung following the insertion of a chest tube for tension pneumothorax can be seen in a comparison with Figure 1. The tube can be seen projecting into the left pleural cavity, as well as the partial return of the mediastinal structures toward the midline

The patient underwent video-assisted thoracoscopic surgery (VATS) for the excision of the bullous lesions in the left lung via wedge resection and partial parietal pleural decortication. The postoperative course was uneventful, and the patient was discharged on postoperative day 5. Histopathological analysis revealed bullous emphysema, with no evidence of vasculitis in the lung parenchyma. Although Behçet's disease may rarely contribute to parenchymal changes, in our case, no histopathological evidence of vasculitis was found. The bullous changes were thus considered idiopathic rather than directly attributable to Behçet's disease, although the coexistence of Behçet's disease may have contributed to disease complexity. Microbiological cultures obtained from both the pulmonary parenchyma and the parietal pleura revealed no growth of Mycobacterium tuberculosis. The patient remains asymptomatic in the 5th postoperative month, and continues to be followed without complications (Figure 4).

DISCUSSION

Spontaneous pneumothorax is an extremely rare complication in Behçet's disease, with available evidence limited to isolated case reports given the lack of definitive epidemiological data regarding its prevalence. In cases with Behçet's disease, spontaneous pneumothorax typically occurs in the later stages of the disease and is associated with severe pulmonary involvement or vascular complications. It is exceedingly uncommon in patients with milder disease or those with predominantly systemic manifestations (7). In our case, the absence of histopathological evidence of vasculitis in the lung parenchyma suggested that the pneumothorax developed outside the classical setting of pulmonary involvement, and indicated that pneumothorax may occur even in patients without overt pulmonary vasculitis. A review of the literature revealed no direct case reports describing tension pneumothorax specifically related to Behçet's disease. While spontaneous pneumothorax and pneumothorax secondary to large cavitary lesions have been reported in patients with Behçet's disease, no cases of tension pneumothorax have been explicitly documented in the literature (4). Recurrence rates after video-assisted thoracoscopic surgery (VATS) for spontaneous pneumothorax have been reported in the range of 2–25%, depending on the study. Recurrence risk is influenced by additional procedures performed (e.g., pleurodesis, pleurectomy), patient-specific factors and the experience of the surgeon. In a large meta-analysis of 23,531 patients, an average recurrence rate of 10% (range 8–12%) was reported, with increased risk in males, young people, those with low body mass index (BMI), and those with a history of contralateral pneumothorax (8). A large cohort study from Taiwan involving 6,654 patients reported a 1-year recurrence rate of 13.7%, with a total recurrence rate of 24.8% over a mean follow-up period (9).

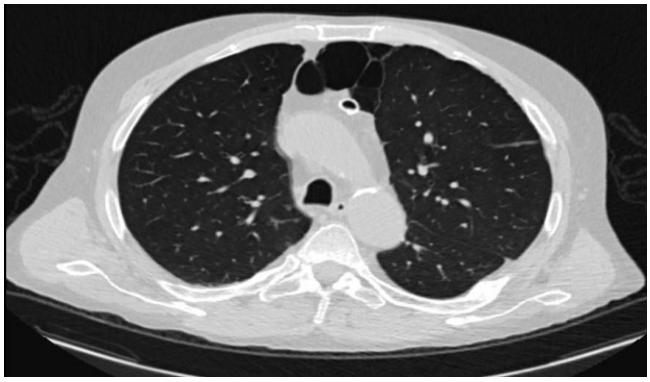

Figure 3: Axial chest CT image demonstrating apical bullous disease. The high-resolution computed tomography (HRCT) of the thorax shows multiple air-filled bullae in the apical segment of the left upper lobe, the largest of which measures 5.5 cm in diameter. The surrounding lung parenchyma appears otherwise preserved, with no radiological evidence of vascular involvement

Given that pneumothorax in Behçet's disease is often associated with significant pulmonary and vascular involvement, close and multidisciplinary follow-up is essential after surgery. During the first year in particular, regular clinical assessments, chest imaging, and symptom monitoring are recommended for the early detection of recurrence and complications. Follow-up imaging is also recommended for the evaluation of underlying conditions such as pulmonary artery aneurysms, cavitary lesions or pneumatoceles. Clinicians should remain vigilant for the potential development of recurrent pneumothorax, infection or bronchopleural fistula, and promptly re-evaluate the patient should such symptoms arise. In a case reported by Gülyüz et al. (10), a young male patient with Behçet's disease developed bilateral spontaneous pneumothorax that was managed conservatively. This case suggests that pneumothorax may represent a potential pulmonary manifestation of Behçet's disease, although most previously reported cases have involved either bilateral or non-tension forms. In contrast, our case involved a rare presentation of tension pneumothorax requiring surgical intervention. The absence of significant parenchymal lesions in the case reported by Gülyüz et al. (10) and the subsequent development of bilateral pneumothorax suggests that such complications may arise not only in advanced stages of the disease, but also in the absence of a clear vascular pathology. Similarly, no histopathological evidence of vasculitis was found in our case, indicating that spontaneous pneumothorax in Behçet's disease may also occur in its less active or atypical forms. While most reported cases in the literature were managed

conservatively, our patient required surgical intervention due to persistent air leakage and the presence of large bullae. The present case can thus be considered unique, and is presented to the literature due to both the clinical decision-making and therapeutic strategy employed. It demonstrates that spontaneous tension pneumothorax, although rare, may occur in patients with Behçet's disease even in the absence of histopathological vasculitis, and that VATS-based surgical intervention can be considered a safe and effective treatment option in appropriately selected cases.

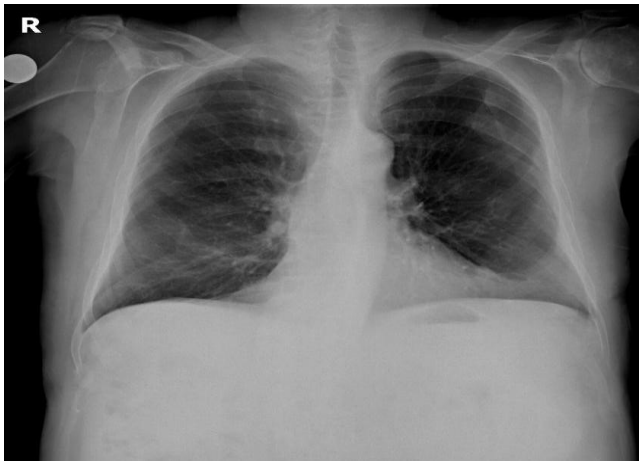

Figure 4: Postoperative posteroanterior chest radiograph. The complete re-expansion of the left lung following video-assisted thoracoscopic surgery (VATS) and wedge resection can be seen. Mediastinal structures are in normal alignment, with no evidence of residual pneumothorax or subcutaneous emphysema

CONCLUSION

Spontaneous pneumothorax is an exceptionally rare and serious complication of Behçet's disease, although only a limited number of cases have been reported in the literature to date. Most previously reported cases involved bilateral or non-tension forms, with surgical intervention typically indicated in the presence of parenchymal destruction or cavitary lesions. The present case contributes uniquely to the existing literature by documenting a spontaneous tension pneumothorax in a patient with Behçet's disease that was managed surgically via video-assisted thoracoscopic surgery (VATS), despite the absence of histopathological evidence of vasculitis. The broader clinical spectrum of pulmonary involvement in Behçet's disease is thus highlighted, supporting the role of surgery as a safe and effective therapeutic option in well-selected patients. In cases with life-threatening presentations such as tension pneumothorax, prompt diagnosis and appropriate management are critical for the reduction of morbidity and mortality. The presented case also underlines the need to consider surgical management in Behçet's patients who present with persistent air leak and bullous disease, even in the absence of classic vasculitic findings.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - A.C., H.Y., P.K.; Planning and Design - A.C., H.Y., P.K.; Supervision - A.C., H.Y., P.K.; Funding - A.C., H.Y., P.K.; Materials - A.C., H.Y., P.K.; Data Collection and/or Processing - A.C., H.Y., P.K.; Analysis and/or Interpretation - A.C., H.Y., P.K.; Literature Review - A.C., H.Y., P.K.; Writing - A.C., H.Y., P.K.; Critical Review - A.C., H.Y., P.K.

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