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Idiopathic Fibrosing Mediastinitis with Non-Compressive Symptoms: A Case Report on Its Surgical Treatment

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Abstract

Herein, we report a case of surgically treated idiopathic fibrosing mediastinitis. A 32-year-old man has an unknown fever. Several medications, including prednisolone, failed to improve his symptoms. (18)F-fluorodeoxyglucose positron emission tomography/computed tomography revealed the abnormal accumulation of the mediastinal and left neck lymph nodes. A definite diagnosis could not be obtained despite an incisional biopsy of the left neck lymph node. We performed robot-assisted thoracic surgery resection of the mediastinal lesion. The pathological diagnosis was idiopathic fibrosing mediastinitis. After surgical treatment, significant clinical improvements were observed. Few reports showed the effect of surgery on idiopathic fibrosing mediastinitis patients with non-compressive symptoms.

Keywords

Idiopathic Fibrosing Mediastinitis, Mediastinum, Non-Compression, Symptoms, Robot-Assisted Thoracic Surgery

1. Introduction

Idiopathic fibrosing mediastinitis, also known as sclerosing mediastinitis, is a rare fibroinflammatory disease of the mediastinum [1]-[3]. Its etiology remains unknown in detail. Numerous case reports have described its association with fibroinflammatory and autoimmune disorders involving other structures [1] [2] [4] [5]. Regarding symptoms, idiopathic fibrosing mediastinitis is manifested by the proliferation of fibrous tissue within the mediastinal structures [1]-[5]. However, there are few reports about the non-compressive symptoms of idiopathic fibrosing

mediastinitis.

The optimal therapeutic approach to idiopathic fibrosing mediastinitis remains controversial. The antifungal and anti-inflammatory agents appeared ineffective [1] [2]. In contrast, the role of surgical interventions was reported for patients with symptoms caused by the compression of mediastinal vascular structures [4]-[6]. However, to our knowledge, few previous reports revealed the effectiveness of surgery for symptomatic patients without mediastinal compression syndromes. Herein, we report a surgically treated case of idiopathic fibrosing mediastinitis with non-compressive symptoms.

The formatter will need to create these components, incorporating the applicable criteria that follow.

2. Case Presentation

A 32-year-old man presented to Imakiire General Hospital with a history of intermittent fevers. His laboratory tests indicated elevated white blood counts and inflammatory markers. He was treated with antibiotics with suspicion of upper respiratory tract infection. However, his fever and laboratory tests did not improve. Computed tomography (CT) revealed anterior mediastinal lymph node swelling (**Figure 1**). The fever and laboratory data did not improve despite administering antibiotics and nonsteroidal anti-inflammatories. Two months after the initial presentation, he was referred to the Department of Hematology ruling out the possibility of hematological malignancy. (18)F-fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) demonstrated the increased FDG uptake of the mediastinal and left neck lymph nodes, suggestive of malignant lymphoma or Castleman disease (**Figure 2**). Serological findings, including soluble interleukin-2 receptor and autoimmune etiologies examined, were within normal limits. Four months after the initial presentation, the incisional biopsy of the left neck lymph node was performed. The pathological diagnosis was non-specific lymphadenitis. He was treated with oral prednisolone 50 mg/day, however, there was no effect to improve the clinical manifestations. Six months after the initial presentation, he was therefore referred to the Department of Thoracic Surgery to rule out malignancy.

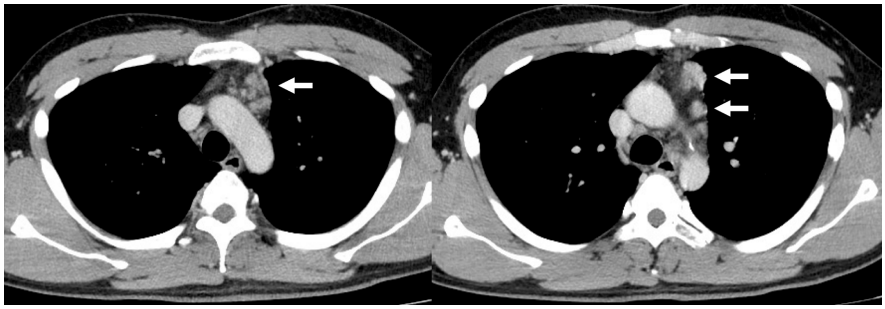

Figure 1. The chest computed tomography (CT) scan showing an anterior mediastinal lesion (arrow).

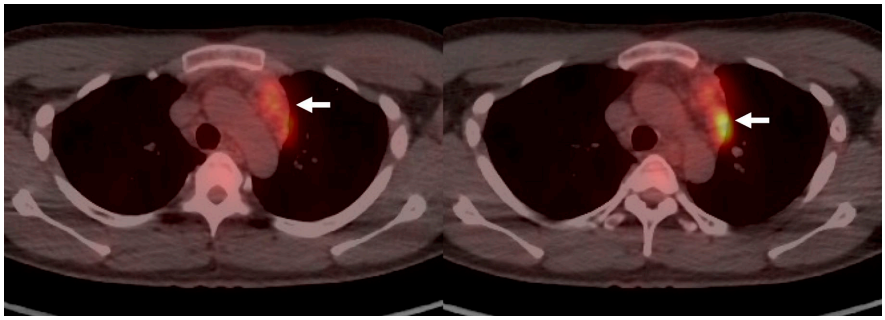

Figure 2. FDG PET/CT demonstrated the increased FDG uptake of anterior mediastinal lesions (arrow).

Under general anesthesia, he underwent robot-assisted thoracic surgery resection of the mediastinal lesion. He was placed in the right lateral decubitus position, and CO_2 insufflation was used. Trocars were placed in the 3rd and the 7th intercostal space (ICS) at the anterior axillary line, and the 5rd and the 7th ICSs at the mid-axillary line. During the procedure, the lesion was hard, and dense adhesions were seen fixing the left upper lung lobe to the mediastinal lesion. The anterior mediastinal lesion was completely removed by careful dissection to avoid damaging the lungs and phrenic nerves during the surgery. The postoperative course was uneventful. He was discharged on postoperative day 5.

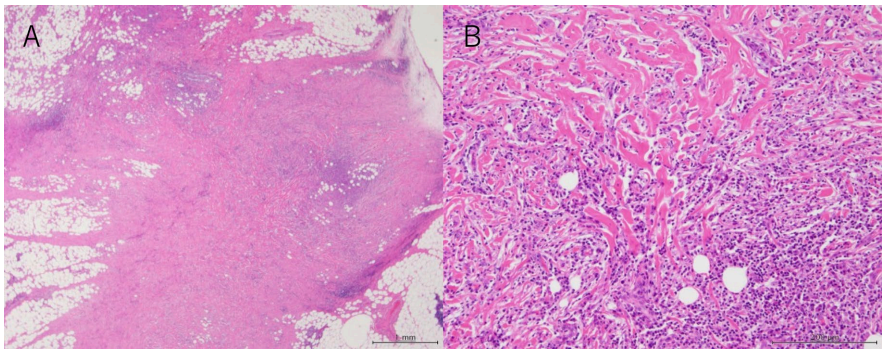

Figure 3. Histological findings of the resected specimen. (A) Low-power magnification view revealed diffuse fibrotic lesions. (B) High-power magnification view showed focal infiltration of lymphocytes. Hematoxylin and Eosin staining.

The pathological findings revealed diffuse fibrotic lesions with hyalinization (**Figure 3(A)**) and the infiltration of lymphocytes (**Figure 3(B)**). No neoplastic lesions were observed. In immunohistochemistry, lymphocytes were identified B lymphocytes as the majority of the mononuclear inflammatory cells along the lesion's periphery. Subsequent workup for infectious, granulomatous, and autoimmune etiologies (IgG4-related disease and Castleman's disease) was negative. A diagnosis of idiopathic fibrosing mediastinitis was made based on pathological findings, highly suggestive radiology, and the exclusion of competing differential diagnoses. His clinical manifestation improved, and he remained asymptomatic 12 months after surgery.

3. Discussion

Idiopathic fibrosing mediastinitis is caused by a proliferation of fibrous tissue in the mediastinum [1]-[3]. The pathogenesis of idiopathic fibrosing mediastinitis remains elusive.

Idiopathic fibrosing mediastinitis has been associated with other idiopathic fibro-inflammatory disorders and autoimmune diseases, including IgG4-related disease. Rossi *et al.* reviewed cases reported in the English language literature throughout 2006-2016 [1]. They showed that 27 of 84 (32%) cases were associated with other idiopathic autoimmune or fibro-inflammatory disorders [1]. Inoue *et al.* suggest that the serum IgG4 level is a good selection indicator for steroid therapy in sclerosing mediastinitis [7]. The present case did not fit the criteria for IgG4-related disease, and it was therefore unclear whether steroid therapy should be used for this case. There are no exact criteria for diagnosing idiopathic fibrosing mediastinitis.

Most clinical symptoms indicate the compression or obstruction of the airways, esophagus, or vascular structures [5]-[9]. Their characteristics differ based on the specific organs that are impacted. However, other clinical symptoms without the compression or obstruction of the mediastinum have been unknown in detail. Although the present case did not have the symptoms of the external compression of mediastinal structures, he had intermittent fever and elevated white blood counts and inflammatory markers. Corticosteroid therapy is the primary treatment for idiopathic fibrosing mediastinitis [1] [2] [5] Although it might appear advantageous, there is currently no standardized protocol [1] [2]. In the present case, the steroid therapy was not effective.

One of the important roles of surgery is open biopsy which could establish a definitive diagnosis and rule out other diseases. However, as a treatment, the role of surgery has shown inconsistent efficacy [4]-[6]. For patients with compression or obstruction of the mediastinum, surgical decompression, and vascular reconstruction might be useful [5]. On the other hand, the surgical treatment for patients without compression or obstruction of the mediastinum does not seem to provide any significant advantage [4]-[6]. However, surgical resection is curative in localized diseases and may ameliorate symptoms. Symptomatic patients with localized disease can also be treated with local therapies. The present case was successfully treated with surgical resection and the symptoms improved. To our knowledge, few reports revealed the effect of surgery on idiopathic fibrosing mediastinitis patients with non-compressive symptoms. We believe that surgery should be considered for patients who do not respond to immunosuppressants for localized idiopathic fibrosing mediastinitis, especially patients with non-compressive symptoms. This is a single case report; therefore, it has several limitations. Further studies are needed to validate these findings and suggest potential future research avenues, such as a retrospective review of surgical outcomes in patients with non-compressive idiopathic fibrosing mediastinitis.

4. Conclusion

We reported a case of idiopathic fibrosing mediastinitis with non-compressive symptoms that were successfully treated with surgical resection and the symptoms resolved. In focal idiopathic fibrosing mediastinitis patients with non-compressive symptoms, it may therefore be worthwhile to try surgical therapy.

Ethical Considerations

The patient agreed to participate in this research and consent was taken to publish the clinical data in the journal.

Conflicts of Interest

The authors declare there are no conflicts of interest regarding the publication of this manuscript.

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