Clinical Case Report

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Anterior mediastinal Hodgkin lymphoma presenting as an extremely hypervascular tumor on computed tomography

A case report

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Abstract

Rationale: In the thorax, Hodgkin lymphoma (HL) most frequently involves the anterior mediastinal and paratracheal regions and tends to spread to contiguous nodal groups. Enlarged lymph nodes typically have homogeneous soft tissue attenuation similar to that of muscle tissue on computed tomography (CT).

Patient concerns: A contrast-enhanced CT examination of a 19-year-old man with right-sided chest pain showed an intense, heterogeneously enhancing mass with organization of serpentine and dilated blood vessels in the right anterior mediastinum that had invaded the upper lobe of the right lung.

Diagnoses: Following a wedge resection, histopathological examination showed Reed-Sternberg cells that were positive for CD-15 and CD-30, which is typical of HL.

Interventions: The patient was started treatment with 6 cycles of doxorubicin, bleomycin, vincristine, and dacarbazine (ABVD) regimen.

Outcomes: After chemotherapy, the patient had shown a partial response to the treatment.

Lessons: This presentation of HL as an extremely hypervascular anterior mediastinal mass on CT imaging has not been previously reported in the literature. This case suggests that HL should be included in the differential diagnosis of a hypervascular anterior mediastinal mass, especially if the patient is a young adult.

Abbreviations: CT = computed tomography, HL = Hodgkin lymphoma, NHL = Non-Hodgkin lymphoma.

Keywords: computed tomography, Hodgkin lymphoma, hypervascular mediastinal mass, thoracic neoplasms

1. Introduction

Hodgkin lymphoma (HL) is a curable malignancy that shows a bimodal age distribution in economically developed countries with peaks in young adulthood and after 50 years of age.[1,2] More than 80% of patients with HL have intrathoracic involvement at the initial presentation.[3,4] In the thorax, HL most frequently involves the anterior mediastinal and paratracheal regions and tends to spread to contiguous nodal groups.[3] Direct extension of HL from the mediastinum to the lung or chest wall is also common with large mediastinal masses.[5] Enlarged lymph nodes typically have homogeneous soft tissue attenuation similar to that of muscle tissue on computed tomography (CT), although they may occasionally be necrotic.[6–9] Rarely, lymphoma may show avid enhancement misleading radiologists in the differential diagnosis of a hypervascular mediastinal tumor.[10]

Herein, we report the case of a 19-year-old man with anterior mediastinal HL that presented as an extremely hypervascular mass with organization of serpentine and dilated blood vessels on the contrast-enhanced CT images. To the best of our knowledge, extreme hypervascularity of HL has not been previously reported in the literature.

2. Case report

2.1. Ethics statement

The patient provided written informed consent for the publication of this report.

Ethics committee approval is not included, as it is commonly accepted that case reports do not require such approval.

2.2. Case presentation

A 19-year-old man presented with right-sided chest pain for 3 weeks. The patient also complained of a productive cough but denied fever or weight loss. The patient had a history of allergic rhinitis but denied smoking. On physical examination, the patient had no clinically detectable lymphadenopathy or organomegaly. Laboratory finding showed an elevated level of C-reactive protein

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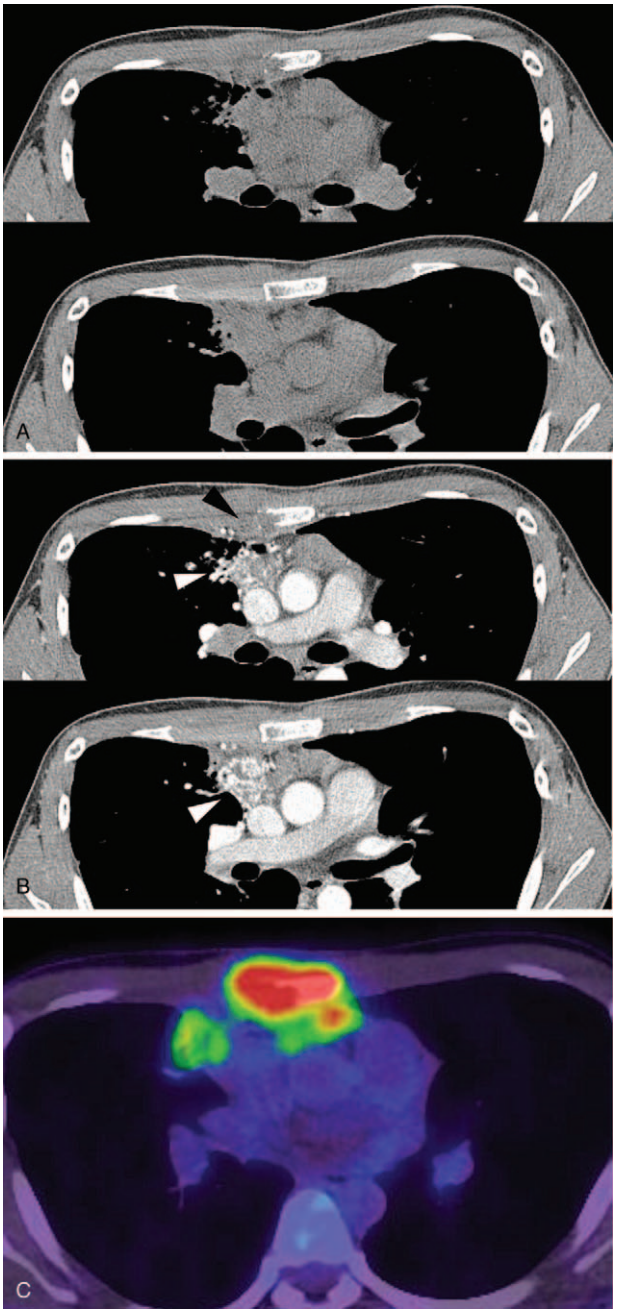

Figure 1. CT and PET/CT images of a 19-year-old man with Hodgkin lymphoma. (A) A nonenhanced chest CT scan demonstrates an ill-defined and homogeneous mass in the right anterior mediastinum with no calcification or fat. (B) A contrast-enhanced chest CT scan demonstrates an intense, heterogeneously enhancing mass with organization of serpentine and dilated blood vessels (white arrowheads) that has invaded the upper lobe of the right lung. Additionally, a contiguous retrosternal and right parasternal soft tissue lesion with destruction of the right sternal body (black arrowhead) and multiple enlarged prevascular lymph nodes can be seen. (C) A PET/CT scan demonstrates heterogeneous hypermetabolic lesions in the right anterior mediastinum, right upper lobe, and sternum as well as enlarged lymph nodes. CT = computed tomography, PET = positron emission tomography.

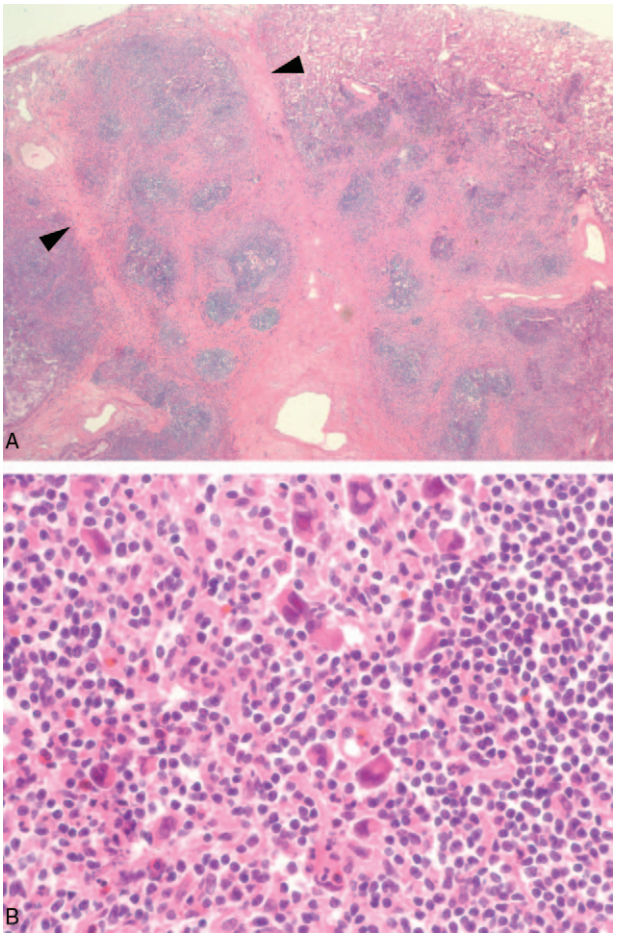

Figure 2. Histopathological findings in a 19-year-old man with Hodgkin lymphoma. (A) Characteristic broad collagen bands (black arrowheads) surrounding nodules composed of a highly variable number of Reed–Sternberg cells, lymphocytes and other inflammatory cells are seen in the lesion (H and E staining, ×12). (B) The typical morphology of Reed–Sternberg cells are seen in the lesion (H and E staining, ×400).

and eosinophilia. The remainder of the physical and laboratory examination was unremarkable. Upon admission, chest radiography revealed a widening of the aortopulmonary stripe. A nonenhanced chest CT scan (Sensation 16; Simens Medical Solutions, Forchheim, Germany) showed an ill-defined and homogeneous mass in the right anterior mediastinum (Fig. 1A) that did not contain calcification or fat. A contrast-enhanced chest CT scan showed an intense, heterogeneously enhancing mass with organization of serpentine and dilated blood vessels that had invaded the upper lobe of the right lung (Fig. 1B). Additionally, a contiguous retrosternal and right parasternal soft tissue lesion with mild enhancement destroyed the right side of the upper sternal body, and multiple enlarged lymph nodes were seen in the supraclavicular, prevascular, aortopulmonary window, right paratracheal, and right hilar regions (Fig. 1B). ^{18}F-fluoro-2-deoxyglucose positron emission tomography (FDG-PET)/CT showed heterogeneous hypermetabolic lesions in the anterior mediastinum, right upper lobe, and sternum in addition to the enlarged lymph nodes already seen by standard CT imaging (Fig. 1C). The organization of serpentine and dilated blood vessels within the mass made the differential diagnosis especially difficult. The initial differential diagnosis included a malignant vascular tumor, vascular malformation, and chronic infectious lesion such as from tuberculosis or a fungus. However, a malignant germ cell tumor or lymphoma could not be excluded. A CT-guided percutaneous transthoracic needle biopsy was not deemed appropriate due to the high risk of bleeding. Instead, an ultrasound-guided needle biopsy of the sternal lesion and

supraclavicular lymph node was performed, but this did not yield a specific diagnosis. Next, anterior mediastinal and chest wall excisional biopsies with wedge resection of the anterior segment of the right upper lobe were performed by video-assisted thoracoscopic surgery (VATS). Intraoperatively, the mass was found to be a hypervascular mediastinal tumor that had invaded the anterior segment of the right upper lobe. The invaded lung tissue showed a fibrotic mass with multiple areas of necrosis. Hematoxylin and eosin staining of the tumor tissue showed characteristic broad collagen bands surrounding nodules composed of a highly variable number of Reed–Sternberg cells, lymphocytes, and other inflammatory cells, which is a typical pattern for nodular sclerosis HL (Fig. 2). Immunohistochemistry of these atypical cells was positive for CD-15 and CD-30 and negative for CD-3, CD-20, and ALK. The in-situ hybridization test for Epstein–Barr virus was negative, and a bone marrow biopsy was positive for lymphoma.

The patient was staged as having IIB HL and was started treatment with 6 cycles of doxorubicin, bleomycin, vincristine, and dacarbazine (ABVD) regimen. A sequential therapy including 2 cycles of cisplatin, cytarabine, and dexamethasone (DHAP) regimen and 3 cycles of carboplatin, ifosfamide, and etoposide (ICE) regimen was planned. After chemotherapy, the patient had shown a partial response to the treatment.

3. Discussion

Lymphomas are divided into HL and non-Hodgkin lymphoma (NHL). The 2008 World Health Organization classification schema recognizes 2 histological types of HL: nodular lymphocyte predominant and classic HL. Classic HL is further divided into 4 subtypes: nodular sclerosis, mixed cellularity, lymphocyte depletion, and lymphocyte rich.[1] The nodular sclerosis subtype, which constitutes 60% to 80% of HL, is frequently observed in adolescents and young adults and usually involves the mediastinum.[5,11]

Thoracic involvement is more common with HL than with NHL.[3] Pulmonary involvement at the time of initial diagnosis is also more common with HL than with NHL.[6] In the thorax, HL most frequently involves the anterior mediastinal and paratracheal regions and tends to spread to contiguous nodal groups.[3]

On CT imaging, HL typically presents with enlarged lymph nodes that can be seen as multiple, rounded soft tissue masses or as bulky soft tissue masses caused by nodal aggregation. In the majority of cases, homogeneous soft tissue attenuation similar to that of muscle tissue is seen on CT imaging. Occasionally, large HL masses may present as heterogeneous with areas of low attenuation representing necrosis, hemorrhage, or cyst formation.[6–9] In a case series by Hopper and colleagues, necrotic and cystic-appearing mediastinal lymph nodes were found during the initial presentation in 21% of HL cases, most commonly in the nodular sclerosis type of HL; however, this finding was not associated with patients' overall survival or length of remission.[7] Necrotic mediastinal lymph nodes are most likely due to ischemia and necrosis from a rapidly growing lymphomatous process. In rare cases, calcification can also be seen in the lymph nodes on pretreatment scans,[12] although calcification is more commonly present after treatment. Rarely, lymphoma may show avid enhancement, which can cause the radiologist to exclude Hodgkin lymphoma from the differential diagnosis.[10] Hypervascular mediastinal masses are characterized by intense enhancement after contrast administration, often with recruitment of adjacent abnormally enlarged feeding vessels.[10] The differential diagnosis of a hypervascular anterior mediastinal mass includes Castleman disease, paraganglioma, carcinoid, ectopic parathyroid adenoma, vascular malformation, and hypervascular nodal metastasis.[10,13] To the best of our knowledge, intense nodal enhancement with extreme hypervascularity in HL has not been previously reported in the literature. In the present case, an ill-defined anterior mediastinal mass demonstrated heterogeneously intense enhancement with organization of serpentine and dilated blood vessels.

In addition, the mass in this case involved the sternum, which is also a rare manifestation of HL. Extranodal involvement is less common with HL than with NHL.[1,14,15] Moreover, the most commonly involved extranodal sites are the gastrointestinal tract, respiratory system, thyroid, skin, genitourinary tract, and central nervous system.[15] Bone constitutes an unusual extranodal site in HL, and sternal involvement of HL is especially uncommon.

In conclusion, this case of HL presented with unusual imaging features, including an extremely hypervascular anterior mediastinal mass. To the best of our knowledge, there have been no reports describing HL tumors with organization of serpentine and dilated blood vessels. This case suggests that HL should be included in the differential diagnosis of a hypervascular anterior mediastinal mass, especially if the patient is a young adult.

Author contributions

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Writing – review & editing: Soo Jeong Lee, Ji Young Rho.

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