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Case Report

CT of Castleman disease in the mediastinum ☆,☆☆,★

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ABSTRACT

Castleman disease is a rare lymphoproliferative disease commonly occurring as a benign localized mass of lymph nodes in the mediastinum. Given that Castleman disease presents as asymptomatic or through non-specific thoracic symptoms, detection is considered complex. Ultimately, surgical resection is the preferred course of action with a greater than 90% relapse-free survival and no malignant transformation reported. In this article, we describe the case of a 34-year-old male with an unclear smoking history who was diagnosed with hyaline-vascular Castleman disease. We focus on optimizing diagnosis and management through the application of radiological imaging modalities, including computed tomography scans.

Introduction

Castleman disease (CD), also known as angiofollicular lymph node hyperplasia or lymph node hyperplasia, was originally described as a benign lymph-node hyperplasia resembling thymoma by Benjamin Castleman in 1954 [1,2]. With the majority of cases located in the chest (70%) along the tracheobronchial tree in the mediastinum or lungs, other common sites of presentation include the neck, pelvis, retroperitoneum, and muscle [3]. Given that patients are usually asymptomatic or present with non-specific symptoms, and there is no significant sex predominance or distinguishable risk factor in its development, CD is considered a challenging preoperative diagnosis and a definite verdict is not established until after tumor resection [2,4]. We present the case of a 34-year-old male with a posterior mediastinal mass that was eventually diagnosed as CD.

Case report

A 34-year-old male with an unclear smoking history presented to his local emergency room with palpitations. An ensuing chest X-ray displayed an incidental finding of a soft tissue mass in the right chest. A follow-up chest computed tomography (CT) scan demonstrated a 6 × 4 cm well-circumscribed,

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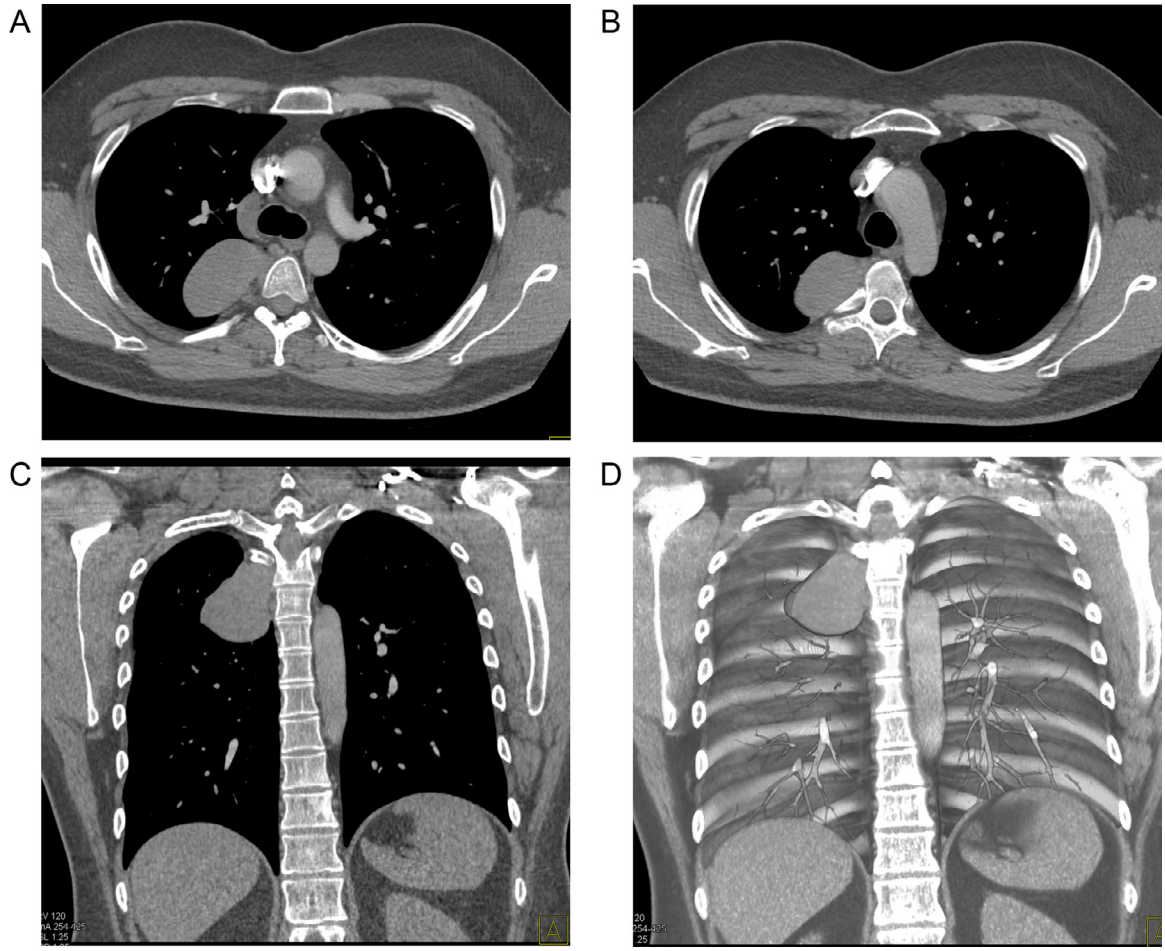

Fig. 1 – A 34-year-old male with an unclear smoking history presented due to palpitations. A chest computed tomography (CT) scan with IV contrast was performed for further evaluation. (A, B) Pleural-based mass is defined as soft tissue attenuation without erosion of the spine but abutting the pleural surface. The CT attenuation of the lesion was 44 HU. (C, D) Coronal view and volume rendered-view in coronal plane demonstrate the smooth borders of the tumor without bone invasion or enlargement of the neural foramina.

homogeneous, extrapleural soft tissue mass in the posteromedial right upper lobe extending into the adjacent neural foramen (Fig. 1). Consequently, the patient underwent a CT-guided right lung mass biopsy with intraoperative consultation. Results showed cores of lymphoid tissue composed of small lymphocytes negative for malignancy, resembling Castleman disease; however, the findings were inconclusive as the collected material was too scant to render a definite diagnosis. Ultimately, the patient successfully underwent a right robotic resection of the mediastinal mass, which measured 5.5 × 4 × 2.5 cm, with a subsequent surgical pathology indicating the morphology and immunoprofile of the mass was consistent with hyaline-vascular Castleman disease.

Discussion

Castleman disease is a rare nonmalignant lymphoproliferative disorder. Clinically, CD is characterized as either multicentric CD (MCD), meaning that it involves multiple lymphatic regions, or unicentric CD (UCD), involving a single lymph node or one region of lymph nodes [5]. While UCD is clinically more predominant, MCD, which has a less favorable prognosis, is divided into 3 subgroups: human herpes virus-8-associated MCD disease, idiopathic MCD, and polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes-associated MCD. Furthermore, CD is distinguished by 2 pathological subtypes, the plasma cell and hyaline-vascular variants. The plasma cell variant, commonly identified as multicentric, involves hyperplastic germinal centers with the interfollicular region of the node being vascular and containing sheets of plasma cells. Conversely, in hyaline-vascular Castleman disease, which is most frequently unicentric, the implicated lymph node follicles have an expanded mantle zone consisting of concentric rings of small lymphocytes enclosing small atrophic germinal centers [6]. We report a 34-year-old male who was diagnosed with hyaline-vascular Castleman disease following a chest CT revelation of an extrapleural soft tissue mass in the right chest, and a subsequent

CT-guided right lung mass biopsy and surgical pathology succeeding robotic resection. The hyaline-vascular CD subtype is commonly diagnosed incidentally through radiological imaging modalities, with CT considered the optimal imaging tool for a proper diagnosis. UCD most commonly presents as a solitary, enlarged, well-circumscribed lymph node or localized nodal masses with an average size of 5-7 cm [5,7,8]. Lesions located in the thorax and abdomen display homogeneous high contrast enhancement, indicating hypervascularity of the mass. Additionally, the presence of calcification observed in CD is useful in differentiating CD and lymphoma, as calcification is uncommon in untreated lymphomas.

With an estimated annual incidence of 4,900 to 6,000 patients in the United States, unicentric CD accounts for approximately 70% of patients with CD and is typically found in adults between the ages of 30-40, with a faint prevalence among women [6,9]. The typical locations of emergence include the chest (29%), neck (23%), abdomen (21%), and retroperitoneum (17%), as well as the axilla, pelvis, and groin, with rare involvement seen in the lungs, trachea, esophagus, and the spleen [7]. Additionally, most patients present with localized, asymptomatic adenopathy. Surgical resection is the treatment of choice, with radiotherapy considered as an alternative in instances where the mass is unresectable. In the case of inflammation, rituximab, steroids, and embolization are also deemed viable treatment options.

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for participation in research, which includes the permission to use data collected in future research projects including presented case details and images used in this manuscript.

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