Case Report

Castleman's disease: A case report of the unicentric type

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ABSTRACT

Castleman's disease, or angiofollicular lymph node hyperplasia, is a relatively rare disorder characterized by the benign proliferation of lymphoid tissue. Two clinical entities have been described: Unicentric with the disease confined to a single anatomic lymph node and multicentric characterized by generalized lymphadenopathy and more aggressive clinical course. Also, three histopathological subtypes have been described: Hyaline-vascular, plasma cell, and a mixed variant. Preoperative diagnosis of hyaline-vascular Castleman's disease is difficult, and the definitive result is based on postoperative pathological findings. The gold standard therapy is the complete surgical excision.

Key words: Angiofollicular lymph node hyperplasia, unicentric, castleman's disease

INTRODUCTION

Castleman's disease (CD) is an uncommon disorder characterized by a benign proliferation of the lymphoid tissue that may be localized or unicentric Castleman's disease (UCD) and disseminated or multicentric Castleman's disease (MCD).[1] Histologically, CD can be classified as hyaline-vascular (HV) type, plasma cell type, or a mixed type. Patients with localized HV type are usually asymptomatic and are diagnosed during routine imaging studies. The disease usually presents in young adults and is probably slightly more frequent in women. The definitive diagnosis is based on post-operative pathological findings. The aim here was to describe a case of cervical UCD, its diagnostic tools and the peri-operative management.

CASE REPORT

This was case report of a 16-year-old girl who presented with a single, painless mass on the right side of her neck for more than 1½ years. She denied other associated comorbidity or systemic disease. Under physical examination, a firm, mobile, non-tender neck mass about 4 cm × 5 cm was noted in the right supraclavicular fossa. Routine biochemistry and complete blood count data all were within normal limits. Computed tomographic (CT) images of the neck with contrast enhancement showed a 52 mm × 29 mm oval shaped strongly enhancing mass in right supraclavicular region compressing internal jugular vein medially [Figures 1 and 2]. Complete excision of the neck mass was performed under general anesthesia. A histopathological diagnosis of CD, HV type [Figure 3], was made. Since, CD can be UCD or MCD, a thorough physical examination and chest and abdominal CT were carried out to exclude any other lesions. However, no further masses or any comorbidity was found.

DISCUSSION

CD was first described in 1956 by Benjamin Castleman, who identified a group of patients with solitary hyperplasic mediastinal lymph nodes with small germinal center resembling Hassall's corpuscles of the thymus.[2] These lymph nodes had small, prominent, hyalinized follicles associated with a marked inter-follicular vascular proliferation (HV variant of CD). This type of disease is now known as UCD.

MCD is a systemic disease characterized by fever and night sweats associated with generalized peripheral

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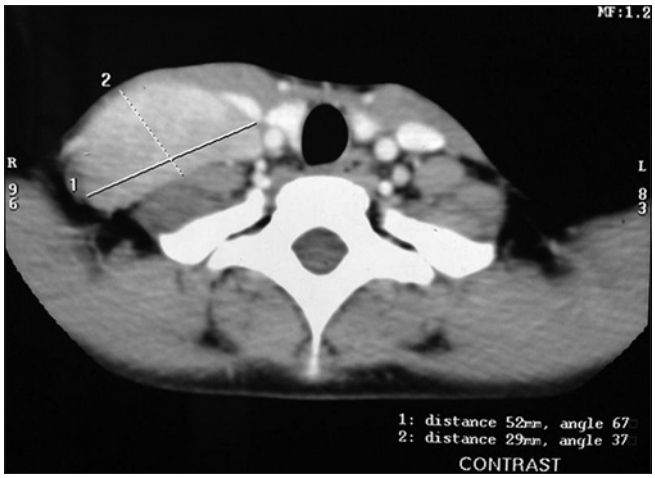

Figure 1: Computed tomography scan picture of the primary tumor

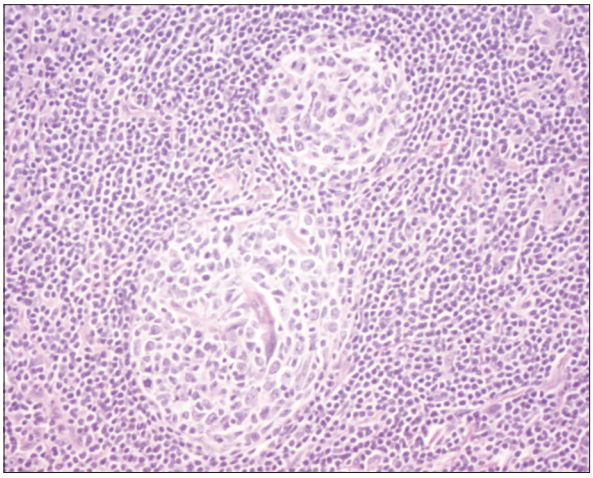

Figure 2: Small lymphocyte encircled follicles with concentric "onion skin" layers and marked inter-follicular vascular proliferation are seen on histology

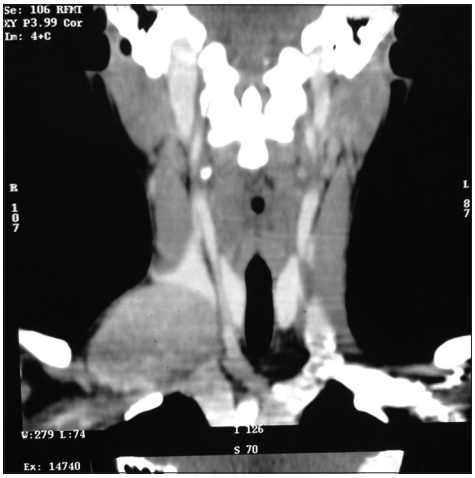

Figure 3: Contrast enhanced computed tomography neck showing a 52 mm × 29 mm oval shaped strongly enhancing mass in right supraclavicular region compressing internal jugular vein medially

lymphadenopathy and hepatosplenomegaly, which is frequently related to the plasma cell variant.[3] Although, the occurrence rate is unknown, it is increasingly relevant nowadays due to its association with human immunodeficiency virus and human herpesvirus-8 (HHV-8).[4] MCD has also been associated with other malignancies, in particular Kaposi's sarcoma and lymphomas. Most patients with MCD die from progression of their disease, disseminated infection, or related malignancies.

A variety of treatments have been used for MCD including surgery, radiation, steroids, antiviral agents, specific antibodies, inhibitors of cytokines activity and chemotherapy.[5] Surgery generally does not have a role in the treatment of MCD, although splenectomy may result in temporary symptomatic improvement.

UCD is the most common type and consists of an isolated benign lymphoproliferative disorder of young adults that is not associated with an HHV-8 infection and usually curable with surgical resection. The vast majority of patients are asymptomatic and their disease is identified incidentally on imaging studies as a soft-tissue mass located in the neck or mediastinum and rarely in the retroperitoneum.

Pre-operative diagnosis of HV CD is difficult. The usual appearance of this entity on a CT or magnetic resonance is that of a non-specific homogeneous mass on non-contrasted studies, with dense enhancement immediately after the infusion of iodinated or gadolinium material and slow washout with the degree of enhancement approaching that of large vessels.[6] The presence of central areas of fibrosis of this tumor is one of the characteristic features of this disease.[7] Fine-needle aspiration cytology is usually inconclusive. The definitive diagnosis is based on post-operative pathological findings. Once CD is diagnosed, MCD must be ruled out. In addition, UCD may be associated with an increased risk of lymphoma (B-cell non-Hodgkin's lymphoma and Hodgkin lymphoma).[8]

The standard therapy for UCD HV form of CD is surgical excision and since these are hypervascular lesions, it is frequently associated with profuse bleeding and pre-operative embolization can help to minimize intra-operative blood loss.[9] Surgery is curative when resection is complete, yielding a 5 year survival rate close to 100%, with recurrences being infrequent.[10] In patients whose lesions cannot be completely resected, outcomes remain favorable. Partially resected masses may remain stable and asymptomatic for many years.[11] Radiotherapy can also achieve clinical response and cure in selected patients. MCD is a more aggressive clinical entity and is most effectively treated with combination chemotherapy, whereas the role of radiotherapy in its treatment remains unclear.[12] Appropriate follow-up should be tailored to the specific CD variant and symptoms. Patients with unicentric disease without systemic involvement should have an additional

radiological assessment 6-12 months after initial therapy, to verify there is not recurrence.

Additional testing or therapy should only be pursued in the event of recurrence or the onset of new symptoms.

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