

A rare case of castleman's disease presenting as cervical neck mass

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Abstract

We report the case of a 16-year-old girl who presented with cervical neck mass. Histopathology after total excision confirmed Castleman's disease of the hyaline-vascular type. This unusual condition needs to be considered in the differential diagnosis of masses arising in the neck.

INTRODUCTION

Castleman's disease (CD) is a unusual benign non-neoplastic lymphoproliferative disease. CD can present as a localised mass first described by Castleman in 1954 [1], or in a more aggressive multicentric form primary reported by Gaba [2]. The disease generally involves the mediastinum and abdomen. Peripheral involvement, specially the head and neck localisation are rare. A diagnosis can be suspected clinically but it's achieved only with histologic analysis. Treatment can range from curative surgery for the localized form to the use of chemotherapy for the multicentric one. We describe a case of cervical CD disease, and review the pertinent literature.

CASE REPORT

A 16-year-old girl presented in January 2006 with a painless mass appeared 6 months ago and progressively increasing in size [fig 1]. There were no constitutional symptoms, including fever, night sweating, fatigue, and weight loss and no alteration in the clinical picture was observed. The rest of the history was unremarkable.

On physical examination, a 4 cm nodule on the right side of the neck, at the level III lymph node region was evident. On palpation, the lesion was asymptomatic, hard, movable, and showed well defined and smooth limits. The size of mass did not change with postural position, and there was no pulsation and bruit on auscultation. There was no any other cervical lymph node enlargement.

Ultrasound showed a rounded mixed-echogenic mass well limited in the right posterior chain of the neck, measuring 38

mm, with a good rim perfusion of the mass, depicted by doppler fluxometric analysis. An intradermoreaction was positive at 13 mm. No abnormalities were identified in other routine blood tests. Excision biopsy was performed [fig 2], and histological diagnosis was consistent with the hyaline-vascular type of CD [fig 3]. Up to now, 15 months after diagnosis, the patient is in good health without signs of recidivism or active disease.

Figure 1

Figure 1 : Clinical aspect of our patient with cervical enlargement which was afterwards diagnosed as Castleman's disease.

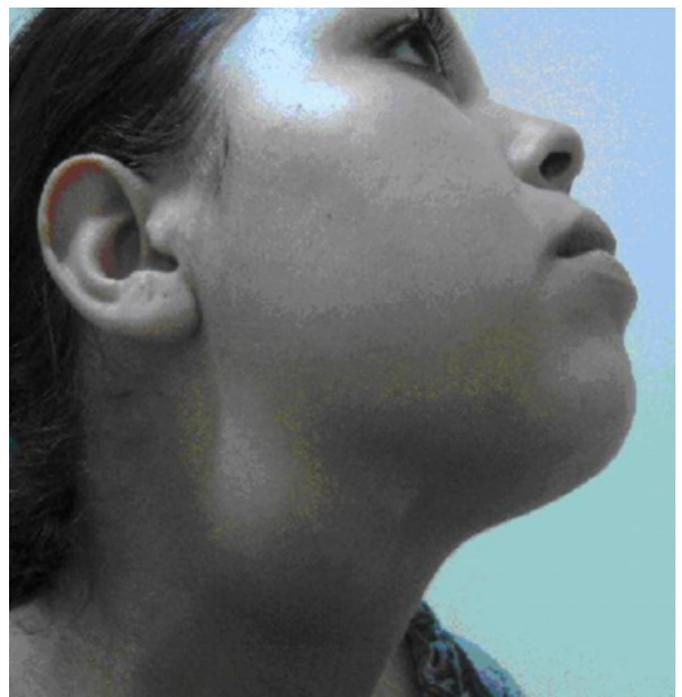


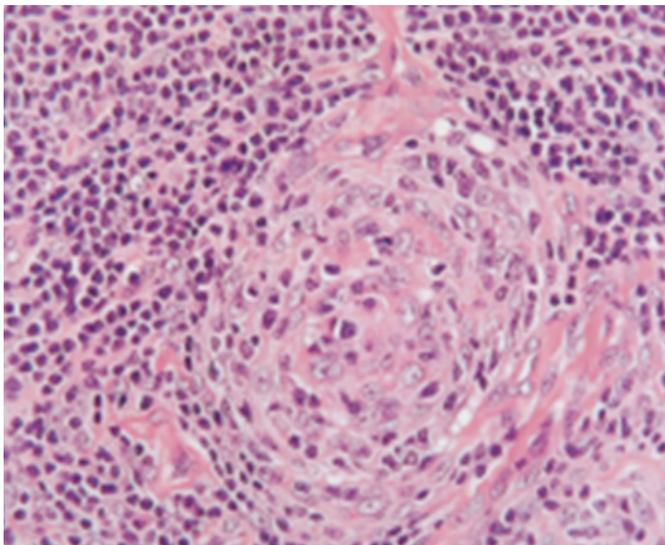
Figure 2

Figure 2: Per-operative photography showing the ovoid encapsulated mass.



Figure 3

Figure 3 : Histopathological aspects of Castleman's disease. A, Typical concentric mantle zone lymphocytes disposed in an onion-skin pattern around the atrophic germinative center (original magnification 100 ×, hematoxylin and eosin).



DISCUSSION

Castleman's disease is a very rare disorder of the lymphoid tissue that is characterized by hypervascular lymphoid hyperplasia. The incidence of the disease in our country is known to be extremely low; however, given the poor access to health institutions in several sectors as well as the diagnosis and treatment of patients in private hospitals, it is quite likely to be underdiagnosed.

CD may occur at any age, but it's more common in adults

than children [3]. The mediastinum is the most common site, accounting for 60% of cases, the neck is involved in only 14% [4]. The majority of cervical cases are lateral rather than midline, occurring in nodes under the sternocleidomastoid muscle [5]. Other reported sites include the parotid gland (21 cases) [6], and there have been individual reports of cases in the floor of the mouth, the submandibular gland, the larynx, the palate and the parapharyngeal space [7,8].

Clinically, the localized form of the disease can be discovered fortuitously at the time of a routine physical examination but it's often represents a diagnostic challenge to the clinician because it tends to mimic other head and neck diseases [9,10]. It lacks specific signs and symptoms, that generally remains asymptomatic unless it begins to compress adjacent structures [11].

In contrast, multicentric disease is clinically more aggressive with systemic manifestations and potentially fatal infective complications.

The differential diagnosis can be done with infectious and inflammatory lesions such as lymphadenitis, tuberculosis, sarcoidosis, toxoplasmosis, cytomegalovirus, mononucleosis, HIV, or some tumors such as neurofibroma, cervical lipoma, Hodgkin's disease, thymoma, non-Hodgkin's disease, and lymph node metastasis.

Final diagnostics of CD depend on histopathology examination which is divided into three subtypes: hyaline-vascular type, plasma cell type, and mixed type [12]. Our case was a hyaline-vascular type, which is characterized by concentrically arranged small lymphocytes around the numerous, small, follicle-like structure with prominent vascular proliferation and hyalinization, it's more common in young adults and older children. This type is usually localized form without constitutional symptoms, and more favorable progress than the other subtypes. The plasma cell type is characterized by sheets of mature plasma cells in the interfollicular spaces and larger hyperplastic follicles with less vascular proliferation [13].

To date, surgical excision is the treatment of choice for Castleman's, especially for the unicentric variant [14]. This treatment allows full recovery in almost all cases. No therapeutic consensus exists at present for multicentric CD. In practice, diverse treatments are used, often in combination, surgery, corticoid therapy, and chemotherapy. Some few rare and more aggressive cases of the two variants of CD are associated with malignancies such as non-

hodgkinlymphoma, Kaposi's sarcoma, follicular dendritic cell sarcoma, or carcinoma [151617].

In conclusion, Castleman's disease should be considered in the differential diagnosis of masses in the neck. Localized CD is almost always of the hyaline-vascular variant and complete surgical excision of the tumor allows full recovery in all cases.

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