

Castleman's Disease in the Neck and Parotid Region: a Report of Two Cases

Boyun ve Parotis Bezi Yerleşimli Castleman Hastalığı: İki Olgu Sunumu

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ABSTRACT

Castleman's disease is also known as angiofollicular lymph node or giant lymph node hyperplasia. It occurs most commonly in adults, equally in men and women. There are two clinical presentations as unicentric and multicentric forms. The two main histological types described are the hyaline vascular and the plasma cellular types. In this study, two cases of unicentric, hyaline vascular Castleman's disease with parotid gland and neck involvement which were surgically excised and histopathologically diagnosed are presented. (*Gazi Med J 2012; 23: 69-72*)

Key Words: Giant lymph node hyperplasia, parotid gland, lymph nodes, hyperplasia

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ÖZET

Castleman hastalığı anjiyofoliküler lenf nodu veya dev lenf nodu hiperplazisi olarak da adlandırılmaktadır. Çoğunlukla yetişkinlerde görülür ve erkek-kadın oranı eşittir. Histolojik olarak hiyalen vasküler tip ve plazma hücreli tip olmak üzere iki ana tipi mevcuttur. Klinik olarak unisentrik veya multisentrik formları bulunmaktadır. Bu çalışmada boyun bölgesinde ve parotis bezinde izole kitle şikayeti ile başvuran, yapılan cerrahi sonrası histopatolojik olarak hiyalen vasküler tip Castleman hastalığı tanısı alan iki olgu sunulmuştur. (*Gazi Med J 2012; 23: 69-72*)

Anahtar Sözcükler: Dev lenf nodu hiperplazisi, parotis bezi, lenf nodları, hiperplazi

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INTRODUCTION

Castleman's disease (CD), also known as angiofollicular lymph node or giant lymph node hyperplasia, is a rare lymphoproliferative disorder that was first described by Benjamin Castleman in 1954 (1). It occurs most commonly in adults, equally in men and women. The two main histological types described for CD are the hyaline vascular type (or angiofollicular) and the

plasma cellular type. In addition, some patients have a mixed variant. There are two clinical presentations as unicentric and multicentric forms.

Unicentric hyaline vascular CD is the most common type which presents with a solitary mass without constitutional symptoms and laboratory abnormalities. On the other hand, multicentric CD, most of which are the plasma cell type, behaves aggressively, presents with multiple lymph nodes, constitutional symptoms

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(e.g. fever, night sweats, fatigue, weight loss) and laboratory abnormalities (anaemia, thrombocytopenia, hypoalbuminuria, polyclonal increase in immunoglobulins). It is also associated with Kaposi's sarcoma, non-Hodgkin's lymphoma and POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes).

In adults, the thorax is the most common site of involvement (60%), followed by the neck (14%), the abdomen (11%) and the axilla (4%) (2). Involvement of the neck and the parotid gland is rare among CD of the head and neck region, and symptoms are non-specific, so diagnosis of the disease is difficult. A review of the literature revealed only a few cases of cervical Castleman's disease presenting with a solitary neck mass. In this article, we present a 36-year-old woman with a solitary mass in the right neck region and a 52-year-old woman with a mass arising from the right parotid gland who were reported as the hyaline vascular type of CD upon histopathological examination.

CASE REPORTS

Case 1

A 36-year-old female presented with a one-week history of swelling in the right neck region. Physical examination revealed a 3x3 cm, mobile, soft and smooth mass located under the sternocleidomastoid muscle (SCM). Neck ultrasonography (USG) was performed and reported a well-encapsulated, hypoechogenic mass along the right common carotid artery, adjacent to the posterior part of SCM with intense vascularity. With USG findings, differential diagnosis of hemangiopericytoma and paraganglioma were suggested primarily. Because of the vascular property of the mass, fine needle aspiration biopsy was avoided. Neck magnetic resonance imaging (MRI) revealed a mass with a size of 17x37.7x52.4 mm located adjacent to the posterior part of the right SCM and lateral to the right internal carotid artery with pathological contrast enhancement (Figure 1). Selective cerebral angiography showed a mass feeding from the branches of the right thyrocervical artery in the right supraclavicular and inferior cervical region. Furthermore, a carotid artery occlusion test performed against possible damage to or interference with the internal carotid artery during the operation revealed pathological changes in electroencephalography without neurological symptoms, so an excisional biopsy was planned.

During the operation, a 30x40 mm mass and four reactive lymph nodes in the right inferior cervical region lateral to the common carotid artery were dissected and removed (Figure 2). There was no direct connection of the mass with adjacent vascular structures. A frozen section biopsy of the specimen was obtained to exclude malignancy which revealed atypical lymphoid hyperplasia. The postoperative period was uneventful. The histopathological diagnosis was consistent with the hyaline vascular subtype of CD.

Case 2

A 52-year-old woman presented with a painless mass in the right parotid region which had been present for 10 years. There were no constitutional symptoms, including fever, weight loss or fatigue and no enlargement of the mass. Upon physical examination, a soft 5x3 cm mass with smooth margins was located in the right parotid region.

USG examination reported a cystic lesion measuring 55x27 mm, extending to a deep lobe of the right parotid gland and fine needle

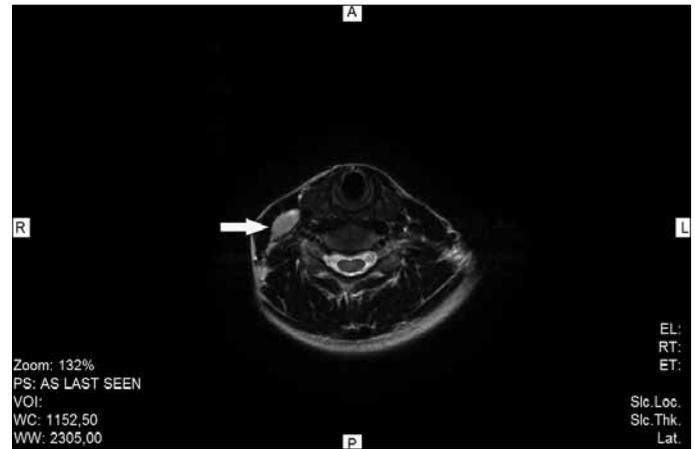


Figure 1. T2 weighted MRI. A solid mass was located in the inferior part of the neck, medial to the sternocleidomastoid muscle and lateral to the right internal carotid artery



Figure 2. Surgical specimen of a 30x 40 mm mass and four reactive lymph nodes dissected from the right inferior cervical region

aspiration was performed under USG guidance. Examination of fine needle aspiration by cytology reported haemorrhagic lymphoid tissue. Subsequent computed tomography imaging detected a 4.5x4x4 cm, lobular, well-circumscribed mass in the right parotid region which extended to the deep lobe of the gland (Figure 3).

As the mass extended to the deep lobe, the mass was removed by superficial parotidectomy and a part of the deep lobe covering the mass. The facial nerve was preserved and no complications were encountered within six months of follow-up. The histopathological examination of the specimen reported CD of the hyaline vascular type (Figure 4).

DISCUSSION

Castleman's disease (CD) is a benign, uncommon lymphoproliferative disorder that was first described by Benjamin Castleman in 1956 (1). Because of its variable clinical presentation, it should be considered in the differential diagnosis of any enlarged lymph nodes or extranodal masses. Head and neck masses account for 14% of CD cases, and 90% of these are located in the neck region. Parotid gland involvement is reported rarely in the literature (3-5).

CD has two clinical manifestations described as unicentric and multicentric disease. In addition to this classification, CD is histo-

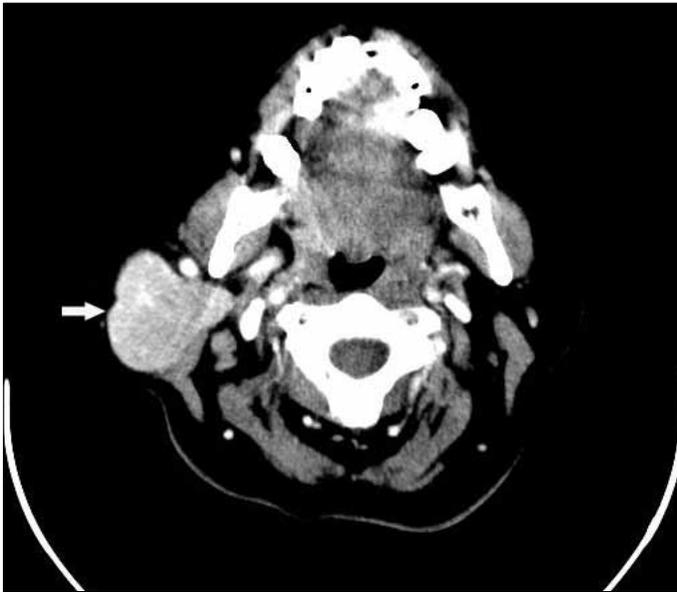


Figure 3. Computed tomography. A lobular, well-circumscribed mass was found in the right parotid region, extending to the deep lobe of the gland

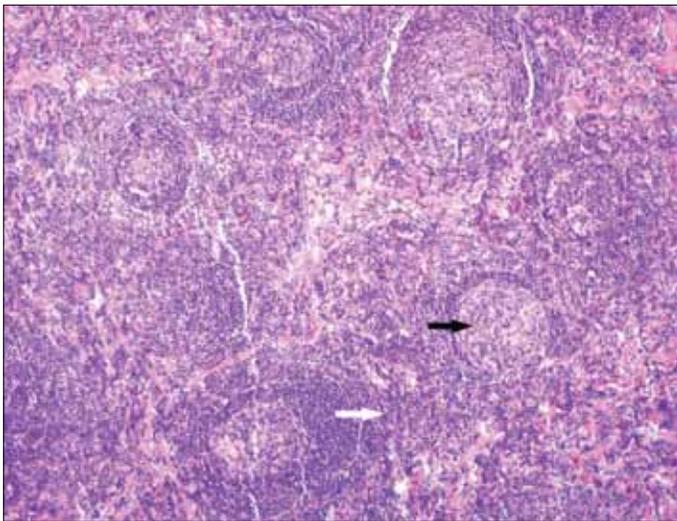


Figure 4. Germinal centre formed by B lymphocytes (black arrow) surrounded by a wide, partly hyalinised mantle zone (white arrow) infiltrated by plasmoblasts

pathologically divided into three subgroups as the hyaline vascular, plasma cell and mixed types. The hyaline vascular type accounts for 90% of CD and is characterised by lymph node follicles with widened mantle zones made of concentric rings of small lymphocytes (in an "onion skin" pattern) that surround small atrophic germinal centres. The germinal centres often have small penetrating hyalinised vessels and prominent follicular dendritic cells (6). The plasma cell type is characterised by increased numbers of plasma cells in the interfollicular and perisinusoidal areas with less vascular proliferation (6, 7). The mixed type is rare and, histologically, is a mixture of the other two types.

Unicentric hyaline vascular CD is the most common type, accounting for approximately 90% of patients with CD; it presents with a solitary mass and a benign course (8). It usually involves the mediastinum (60%), but may also occur in the head and neck region

(14%), the retroperitoneum (11%) and the axilla (4%) (9, 10). Systemic symptoms are unusual (11). In the unicentric plasma cell variant, constitutional symptoms and laboratory abnormalities may be prominent in addition to enlarged lymph nodes much like in multicentric CD. This subtype represents about 10-20% of cases (12).

Multicentric CD is a less common systemic lymphoproliferative disorder which presents with fever, diffuse lymphadenopathy, splenomegaly, severe cytopenia, high serum C-reactive protein (CRP) and other constitutional symptoms. Several studies have shown that human herpesvirus (HHV-8) and interleukin (IL)-6 are associated with CD through the proliferation of B cells (13, 14). Kaposi sarcoma, non-Hodgkin's lymphoma, Hodgkin's lymphoma, and POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes) are conditions associated with Castleman's disease (15, 16).

In this report, we observed different growth rates of the masses, with a rapidly enlarging mass in case 1 in the neck region resembling lymphoma and a slowly enlarging mass over a period of ten years in case 2, similar to other benign pathologies of the parotid gland. These non-specific symptoms impeded the correct diagnosis of CD.

The absence of specific signs, symptoms, definitive radiological findings, a tendency to mimic other head and neck neoplasms and inflammatory lesions complicate the diagnosis of CD. CT imaging and MRI show contrast enhancement, and preoperative angiography shows the high vascularity of the mass. Macroscopically, Castleman's disease tissue usually appears as well-circumscribed, firm, ovoid masses ranging from 3 to 8 cm in maximum diameter (17). Fine needle aspiration (FNA) cytology is an easy and useful technique to assess masses in the neck, but it is not always possible to obtain a definitive diagnosis of CD from FNA specimens. A few cytological case reports of CD have been published in the literature, and no specific diagnostic cytomorphological criteria have been described (18). The most dependable method of establishing a definitive diagnosis is by surgical resection and histopathological examination of the mass.

Surgical removal is almost always successful for unicentric disease, whether the hyaline vascular or plasma cell variant (3). For localised CD, if complete resection is not adequate, irradiation is an effective alternative, with response rates up to 72% (19).

The management of multicentric CD is more difficult and requires systemic therapy. Prednisolone or other glucocorticosteroids, chemotherapy ranging from the use of single agents (vinblastine, oral etoposide, interferon) to multidrug combinations that are used to treat lymphomas are therapeutic options. Also, rituximab, a monoclonal antibody against the CD20 antigen found on lymphoid cells in HHV-8-related CD and a humanised monoclonal antibody against the IL-6 receptor (tocilizumab) have been studied and objective improvement can be achieved with these drugs (20).

CONCLUSION

Castleman's disease should be kept mind in the differential diagnosis of a mass encountered in the neck and parotid region in patients with or without other systemic conditions. For unicentric disease, the most accurate method for a definitive diagnosis and correct management is surgical excision and histological examination.

Conflict of Interest

No conflict of interest was declared by the authors.

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