Case Report / Olgu Sunumu

A rapidly growing neck mass in childhood: Castleman's disease concomitant with cavernous lymphangioma in the posterior cervical region

Çocukluk çağı hızlı büyüyen boyun kitlesi: Posteriyor servikal bölgede kavernöz lenfanjioma ile birliktelik gösteren Castleman hastalığı

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In this article, we present a 12-year-old girl case with a painless mass in the left posterior region of the neck for two weeks. Two masses were detected during surgery. Histopathologic examination revealed the hyaline vascular type of Castleman's disease for the bigger mass and cavernous lymphangioma for the smaller mass. In the light of literature data, our case was the third childhood cervical posterior triangle Castleman's disease and was the first case reported due to its concomitance with cavernous lymphangioma.

Key Words: Castleman's disease; cavernous lymphangioma; posterior servical region.

Bu yazıda, iki haftadır boynunun sol arka bölgesinde ağrısız kitlesi olan 12 yaşında bir kız olgu sunuldu. Ameliyat esnasında iki ayrı kitleye rastlandı. Histopatolojik incelemede büyük olan kitlenin hiyalen vasküler tipte Castleman hastalığı ve küçük olan kitlenin ise kavernöz lenfanjiyom olduğu gözlendi. Literatür verileri ışığında bu olgu literatürde çocukluk çağı arka servikal üçgen yerleşimli Castleman hastalığı olan üçüncü olgu ve kavernöz lenfanjiyom birlikteliği nedeniyle de bildirilen ilk olgudur.

Anahtar Sözcükler: Castleman hastalığı; kavernöz lenfanjioma; arka servikal bölge.

Castleman's disease (CD) is an uncommon cause of neck masses which can easily be misdiagnosed. It has also been called angiofollicular lymphoid hyperplasia, giant lymph node hyperplasia and angiomatous lymphoid hamartoma in the literature.[1-4] In general it is seen in the mediastinum and abdomen, and the most common extrathoracic site is the neck. The disease was first described by Benjamin Castleman et al.[5] in 1956 as a localized mediastinal lymph node hyperplasia resembling thymoma. After that, the multicentric or systemic form was described by Gaba et al.[6]

Available online at
www.kbbihtisas.org
doi: 10.5606/kbbihtisas.2013.48569
QR (Quick Response) Code

Received / *Geliş tarihi:* June 02, 2012 Accepted / *Kabul tarihi:* November 01, 2012

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Histopathologically, the disease has two basic subtypes; the hyaline vascular and plasma cell (PC), nevertheless a mixed variant has also been described.[7] The localized form is usually with hyaline vascular subtype on histopathological examination and it commonly follows a benign progress. The multicentric form is mostly together with plasma cell subtype and it is characterized by generalized lymphadenopathy with other symptoms such as fever, weight loss, fatigue and excessive sweating. In the localized form, such systemic symptoms do not show up, unless there is a large mass which causes pressure on the adjacent structures. Treatment can be discussed on the basis of the histopathological examination and clinical findings.

We present a case of a 12-year-old girl who had a hyaline vascular Castleman's disease concomitant with cavernous lymphangioma in the posterior cervical region. A PubMed search reveals this to be the third childhood CD located in the posterior cervical region and the first case reported with concomitant cavernous lymphangioma.[8,9] Castleman's disease should be considered as a differential diagnosis in patients with a posterior neck mass though it is not a common disease, and also we must take into consideration that other neck masses could be seen concurrently with CD. In this paper we also aim to review the literature in detail about CD and lymphangioma.

CASE REPORT

A 12-year-old girl with a two-week history of a painless mass in the left side of the neck consulted at our clinic. Her parents noted that the patient had this neck mass in smaller size since her birth, but it increased rapidly for the last two weeks. On physical examination, a 7x6 cm rubbery and tender mass was found in the left posterior cervical region and interestingly it pushed the sternocleidomastoid muscle (SCM) anterolaterally. A complete examination of the ear, nose, throat and also the other systems showed no other pathologic findings. There was no other cervical lymph node enlargement or hepatosplenomegaly. The complete blood count, serological tests and chest radiography were also normal. Neck ultrasonography showed a 4x4x2 cm cystic mass located in the left posterior cervical region. Magnetic resonance imaging (MRI) scans showed that the mass had multicystic loculi separated by multiple septa (Figure 1). Fine-needle aspiration cytology identified numerous lymphocytes and some histiocytes, precluding any specific diagnosis. Subsequently the mass was removed totally by transcervical dissection. During the operation two different masses were observed, which were related to each other and extended medially to the scalenus muscles, anteriorly to the vena jugularis interna and posteriorly to the trapezius muscle. The masses were dissected; also upper and mid jugular nodes were dissected from the surrounding tissues beneath the SCM. Postoperatively, the large and small masses were determined to be 9x6 cm and 3x4 cm, respectively (Figure 2). Histopathologic examination of the specimen revealed that the large and the small masses were of the hyaline vascular type of CD and cavernous lymphangioma, respectively. There were atrophic germinal centers, which were surrounded by concentrically aligned lymphocytes with an "onion skin appearance", in the histopathologic examination of the mass with CD (Figure 3). There were also hyalinized blood vessels penetrating into these germinal centers. In the histopathologic examination of the mass with cavernous lymphangioma, the dilated lymphatic vessels with thick walls had proteinaceous fluid in them (Figure 4). On second year follow-up, the patient remained asymptomatic and there were no remaining or relapsing mass in the neck.

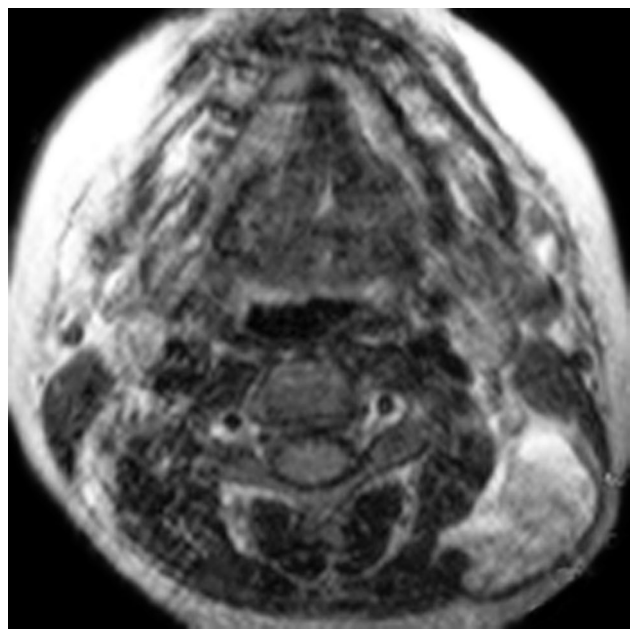

Figure 1. *Axial T_2-weighted image shows the neck mass at the jugulodigastric level.*

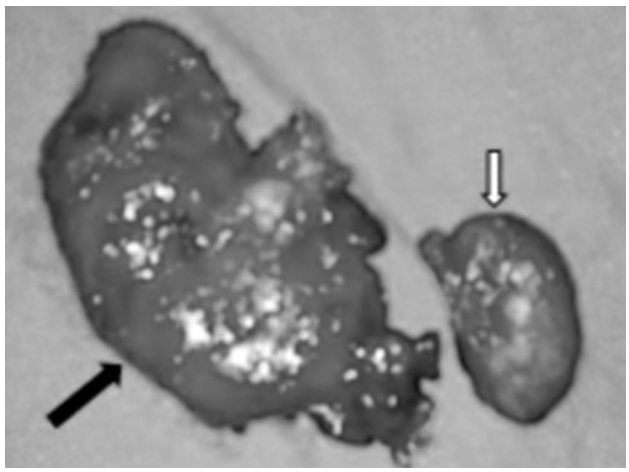

Figure 2. *Black arrow shows the giant lymph node hyperplasia which was diagnosed as Castleman's disease; the white arrow shows cavernous lyphangioma.*

DISCUSSION

Castleman's disease is an uncommon lymphoproliferative disorder which has numerous etiological factors. As a result of chronic antigenic stimulation reactive lymph node hyperplasia can occur. In general, this antigenic stimulation is known to be viral, also there are immunologically mediated and systemic diseases which were associated with CD such as myasthenia gravis, pemphigus vulgaris, temporal arteritis, monoclonal hypergammaglobulinemia, nephrotic syndrome, human immunodeficiency virus (HIV), Kaposi's sarcoma, lymphoma and vascular neoplasms.[2,10-13]

No gender or race predominance has been reported in the literature. Although the disease may occur at any age, it occurs primarily in young adults within the age of 15 to 30.[1,3,4,11,12,14] Frequently, CD has been found in the mediastinum (60-70%), head and neck (14%), retroperitoneum (11%) or axilla (4%).[4] On the other hand, the neck is the most common extrathoracic site.[1-4,11,12,14,15] Pharapharyngeal space placement have even been reported.[16] In the literature, there are six adults and two children with CD reported in the posterior cervical region.[8,10,15,17-19] In this report, we present a 12-year-old girl whose neck mass was associated with cavernous lymphangioma located in the posterior cervical region. When we consider all the reports about this topic, our case is the third childhood CD located in the posterior cervical region and the first case associated with cavernous lymphangioma.

Lymphangiomas are benign malformations of the lymphatic system and divided into three groups as capillary, cavernous, and cystic (cystic hygroma). These lymphatic malformations can be present at birth and 80-90% of them are diagnosed within the two years of life.[20-23] Trauma, chronic inflammation, obstruction and infections can be the reason of pathogenesis. More than 50% of the lymphangiomas are found in the head and neck region.[20,21] Cystic lymphangiomas are particularly found in the posterior cervical region. However, up to now no cavernous lymphangioma has been described in the posterior cervical region. According to our knowledge this case is the first report of a cavernous lymphangioma associated with CD in the posterior cervical region. Cavernous lymphangiomas are located

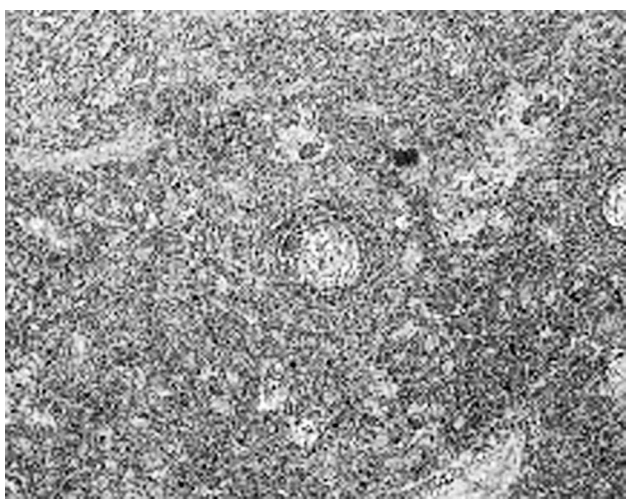

Figure 3. *Hyalinization in the germinal center of the follicle (H-E x 100).*

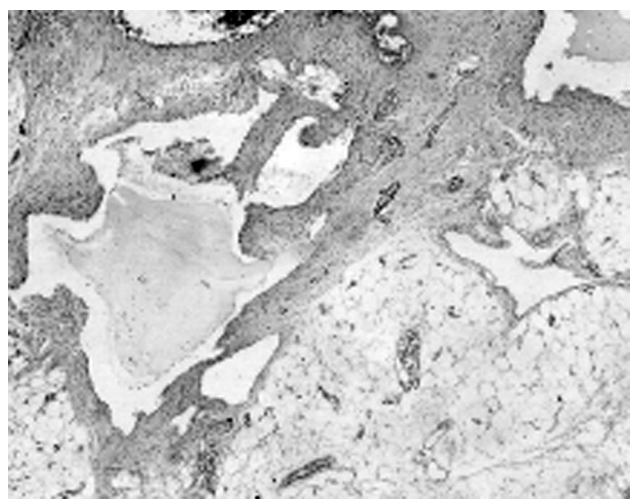

Figure 4. *Dilated vascular channels, with thickened walls, filled with proteinaceous fluid (H-E x 40).*

mostly in the tongue, floor of the mouth and salivary glands[23] and are characterized with capillary dilatation and irregular branching of the lymphatic vessels.[24]

Castleman's disease is classified into two groups: localized (unicentric) and disseminated (multicentric). The localized form appears with a painless, slow growing lymph node enlargement and it exists especially in younger adults. Although rapid growth is an uncommon feature in localized CD,[1,4,11,15] the most prominent complaint was the rapidly growing mass in the neck in our case. We proposed that rapid growth might have occurred due to the cavernous lymphangioma. The localized form usually follows a benign course, though a large mass may cause symptoms due to the pressure on the adjacent structures. The multicentric form of the disease is rare, and it tends to occur in older patients.[1,8,10] In the localized form, regardless of histological subtypes, complete surgical excision allows full recovery and no adjuvant therapy is required.[1,2,4,8,10-13] Recurrence can occur with respect to insufficient resection of the mass. Multicentric disease may require more aggressive therapy. Radiotherapy, chemotherapy (such as cyclophosphamide, vincristine, doxorubicin), steroids and anti interleukin-6 (IL-6) receptor antibody therapies are the choices for the treatment of multicentric diseases.[2,4,8,10-13] In this case we totally excised the neck mass and there were no symptoms or clinical findings postoperatively at the end of the one-year follow-up.

In conclusion, Castleman's disease and lymphangioma must be kept in mind in the differential diagnosis of the posterior neck masses.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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