

Multiple and Huge Axillary and Neck Lymph Nodes

Çoklu ve Büyük Koltuk Altı ve Boyun Lenf Nodları

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ABSTRACT

We wanted to discuss our 3-year-old patient, who first started with small lymph nodes in the axillary region and then appeared on the neck, and these swellings gradually grew and were not accompanied by pain.

Key Words: Neck, axillary, multiple, huge, lymph node

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ÖZET

Önce aksiller bölgede küçük lenf nodları ile başlayıp ve sonrasında boyunda da ortaya çıkan ve bu şişliklerin giderek büyüdüğü ve ağrının eşlik etmediği 3 yaşındaki hastamızı tartışmak istedik.

Anahtar Sözcükler: Boyun, aksiller, çoklu, büyük, lenf nodu

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INTRODUCTION

Our patient was a 3-year-old boy, the second child of a 25-year-old father and 21-year-old mother. There were no problems in his perinatal history or family history. He was born via the normal vaginal route and had a normal vaccination schedule and development. At 30 months of age, a large number of axillary masses measuring 1 cm by 1 cm appeared on both sides, followed by considerably reactive lymph nodes. At 35 months of age, as the axillary masses grew, similar swellings appeared in the patient's neck. So there is no pain, fever and loss of appetite. The patient has not been in contact with anyone ill and he has not travel anywhere that it known to have an outbreak of any disease. He had no night sweating. The patient's temperature was 37 °C, his pulse was 110 beats per minute, and he had 26 respirations per minute. His blood pressure was 100/70 mmHg, he weighed 11 kg (5% percentile) and he measured 86 cm (3% percentile) in length.

On physical examination, we detected a large number of lymph nodes measuring 5 cm by 6 cm, which filled both the supraclavicular and axillary regions. The skin on the masses appeared normal (Figure 1). The liver was palpable four cm from the costal arc but spleen was normal. There was no edema, skin rash, neuropathy and endocrinopathy. Hemoglobin concentration was 6.1 g/dL, white cell count was $13.9 \times 10^9/L$, platelets were $289.000/mm^3$, MCV was 71.6%. Peripheral smear showed microcytic hypochromic anemia and no haemolytic anemia. Serum albumin and globulin levels were 3.92 and 2.30 g/dL, respectively and LDH level was 110 IU/L, with in normal levels). Erythrocyte sedimentation rate was 12 mm/h (normal range=0-15 mm/h) and C reactive protein was 0.8 mg/dl (normal range=0-1 mg/dl) while other tests were normal (HIV test was negative). Ultrasonography showed diffuse homogeneous and hypoechoic lesions of various sizes.

Because of the growing nodes and there was not enough technological equipments (this patient applied before 1990), a biopsy decision was primarily. This means that tests such as serum and urine monoclonal gammopathy, reticulocyte count and HHV8 and PET-CT scan were not performed. We excised the largest lymph node (measuring 5x6 cm) from the right axilla. Microscopic examination revealed hyperplastic lymphoid follicles with atrophied, fibrous and richly vascularized germ centers surrounded by concentric layers of small mature lymphocytes. The inter-follicle tissue was rich in capillary-type vessels.



Figure 1: Clinical photograph of our patient at 3 years of age which showed the all lymph nodes.

What is your diagnosis?

Answer: Because of the patient's chronic history, physical examination and pathological features, we diagnosed multicentric Castleman disease (CD). This patient presented neck CD at an early age rare in the literature.

CD was first diagnosed by Benjamin Castleman in 1954, followed by 13 cases of mediastinal lymph nodes. CD is a rare form of lymphoid hyperplasia and it also known as exogenous angiophilic lymph node hyperplasia, giant lymph node hyperplasia and angiophilic lymphoid hamartoma(1,2). It usually develops in patients between the ages of 2 months and 76 years, most commonly during their teens and twenties.

There are approximately 20 to 25 cases per million each year, and there is no gender difference. It is classified as either multicentric or unicentric, with multicentric CD accounting for 23% of cases. Generally, painless chronic lymph nodes appear on the patient's body. Thirty-three percent of cases involve the thorax, 30% involve the abdomen, and fewer than 15% of cases involve the neck. Pediatric patients with neck involvement account for only 18 articles (29 cases) in the literature(3,4).

The etiology of CD is not clear. Histologically, generally 90% of the masses are of the hyaline vasculature type, 10% are plasma cell and very few are classified as mixed-type CD.

However, human herpesvirus-8 (HHV-8) and interleukin-6 (IL-6) have been shown to play an important role in most cases; they contribute to the stimulation of multicentric CD and plasma cell origin. Therefore, recent care has focused on monoclonal antibody treatment against IL-6 and its receptors(3,4).

CD patients may present with mass compression, fever, fatigue, sweating, weight loss, skin rash and cytopenia. In addition, multicentric CD patients may demonstrate polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes (POEMS syndrome)(3,4).

In the presence of an increasing number of painless chronic lymph nodes, CD should be suspected. Computed tomography and magnetic resonance imaging show homogeneous contrast enhancement, but a complete diagnosis requires more than these tests and an ultrasound. Fine needle aspiration is also generally insufficient for CD diagnosis and typing. Pathological diagnosis is also needed to distinguish CD from lymphoma, which presents similar symptoms. Therefore, an excisional biopsy provides adequate diagnosis and treatment, especially in unicentric CD cases. In multicentric cases, the removal of an appropriate large lymph node is sufficient for diagnosis and typing(3,4,5).

Unicentric CD is treated with total excision. The primary treatment for multicentric CD is multimodal, including chemotherapy and radiotherapy, although multimodal treatment is rare in children. In addition, antiviral therapy and anti-IL-6 monoclonal antibody therapy have recently been used to treat the etiopathogenesis of HHV-8 and IL-6(1,3,5).

CONCLUSION

Castleman disease should be considered when diagnosing the cause of chronic and painless lymph nodes. Definitive diagnosis and treatment requires excision.

Conflict of interest

No conflict of interest was declared by the authors.

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