OLGU SUNUMU CASE REPORT

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Castleman's Disease Mimicking a Parotid Gland Tumor

Parotis Bezi Tümörünü Taklit Eden Castleman Hastalığı

Sümeyye EKMEKÇİ,[a]
Gülen GÜL,[a]
Ülkü KÜÇÜK,[a]
Dudu SOLAKOĞLU KAHRAMAN,[a]
Ebru ÇAKIR,[a]
Suat KAPTANER,[b]
Sermin ÖZKAL[c]

Clinics of
[a]Pathology,
[b]Otolaryngology,
Tepecik Training and Research Hospital,
[c]Department of Pathology,
Dokuz Eylul University
Faculty of Medicine,
İzmir

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Correspondence:
Sümeyye EKMEKÇİ
Tepecik Training and Research Hospital,
Clinic of Pathology, İzmir,
TURKEY/TÜRKİYE
ekmekcisumeyye@gmail.com

ABSTRACT Castleman disease (CD) is a rare benign lymphoproliferative disorder of unknown etiology, typically presenting as a solitary mediastinal mass. The head and neck region is the most common site; however, the salivary glands are rarely affected. We present a rare case of a 59-year-old male patient with mixed type CD localized in the parotid gland. The patient underwent a left superficial parotidectomy with a clinical prediagnosis of mucoepidermoid carcinoma. H&E stained sections showed mostly preserved lymph node architecture. Some lymphoid follicles had atrophic germinal centers and mantle zones was expanded. Also there were sclerotic blood vessels radially penetrated the germinal centers and the overall appearance of the follicle was resembling a lollipop. The diagnosis was CD, mixed type. For the accurate diagnosis of this entity, histopathological examination should be performed and physicians should kept in mind this clinical entity so patients can receive adequate and early treatment.

Keywords: Parotid region; giant lymph node hyperplasia

ÖZET Castleman hastalığı (CH), etiyolojisi bilinmeyen, tipik olarak soliter mediastinal kitleler ile kendini gösteren, benign nadir bir lenfoproliferatif bozukluktur. Baş ve boyun bölgesi en yaygın yerleşim yeridir, ancak tükrük bezleri nadiren tutulur. Bu yazıda, 59 yaşında erkek hastada, nadir görülen bir antite olan parotis yerleşimli mikst tip CH olgusu sunulmaktadır. Hastaya mukoepidermoid karsinomu klinik ön tanısı ile sol yüzeysel parotidektomi yapıldı. H&E boyalı kesitlerde çoğu alanda yapısı korunmuş lenf düğümü izlendi. Bazı lenfoid folliküllerin atrofik germinal merkezleri vardı ve mantle zonları genişlemişti. Ayrıca germinal merkezleri radyal olarak penetre eden sklerotik damar yapıları gözlendi ve folikülün genel görünümü bir lolipopa benziyordu. Olguya bu bulgular eşliğinde CH, mikst tip tanısı verildi. Bu antitenin tanısı için histopatolojik inceleme şarttır ve hastaların erken tanı ve tedavisi için hekimlerin bu antiteyi akıllarında tutmaları gerekir.

Anahtar Kelimeler: Parotis bölgesi; dev lenf nodu hiperplazisi

Castleman disease (CD) is a rare benign lymphoproliferative disorder of unknown etiology, typically presenting as a solitary mediastinal mass. The head and neck is the most common site but involvement of the parotid gland is extremely rare.[1] Based on analysis of American databases, the incidence of CD is estimated as 21-25 cases per million person-years.[2] In this article, a patient who has mixed type CD causing a parotid mass is presented.

CASE REPORT

A 59 years old man admitted to our otorhinolaryngology clinic with a complaint of tinnitus. On physical examination, a painless swelling on his neck

was observed. There were no signs of inflammation and palpable lymph nodes, and no evidence of facial nerve involvement. The patient underwent a left superficial parotidectomy with a clinical suspect of mucoepidermoid carcinoma. Hematoxylin and eosin (H&E) stained sections of the formalin-fixed and paraffin-embedded tissues showed mostly preserved lymph node architecture. Some lymphoid follicles had atrophic germinal centers and mantle zones was expanded with the cells some what aligned concentrically (Figure 1). Also there were sclerotic blood vessels radially penetrated the germinal centers and the overall appearance of the follicle was resembling a lollipop (Figure 2). Immunohistochemically germinal centers showed positivity for CD23. There were vascular structures in the germinal centres of the follicles and CD34 positive blood vessels were extending out from the germinal centers in some follicles (Figure 3).

Small lymphocytes, immunoblasts and sheets of CD138 positive mature plasma cells were seen at the interfollicular regions. Plasma cells showed polytypic light chain expression. Immunhistochemical analysis of HHV-8 was negative. The diagnosis was CD, mixed type. The same histologic findings were found in other lymph nodes in the neck dissection material. With the help of the clinical findings, the case was accepted as a multicentric CD. The patient was treated with cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP) chemotherapy. Clinically and radiologically no recurrence was noted during a follow up period of 20 months.

DISCUSSION

Castleman's disease, also known as giant lymph node hyperplasia or angiofollicular lymph node hyperplasia, was first defined in 1954 by Castleman and his colleagues.[3] It usually appears as a mediastinal, cervical, mesenteric, or retroperitoneal mass.[4]

Commonly the disease is located in the mediastinum (60%), and head and neck area accounts for 14% of cases.[1] CD of the parotid gland is an extremely rare entity, with only 26 cases reported in the English literature.[5,6]

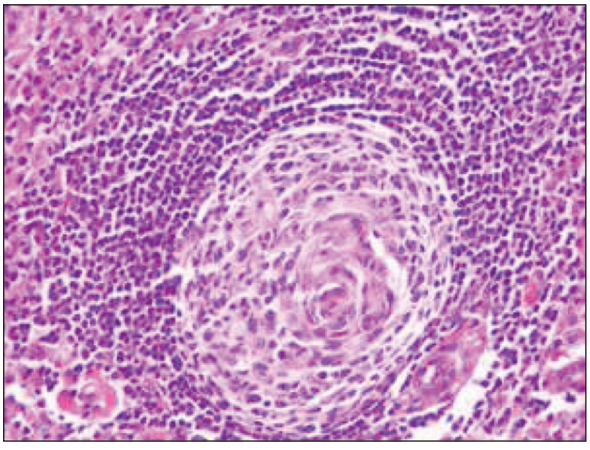

FIGURE 1: Atrophic germinal centers and mantle zone consisting of a concentric layering of lymphocytes- onion skin appearance (HE, x400).

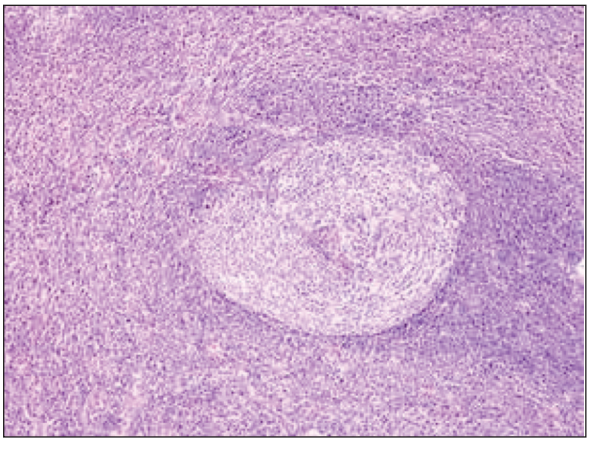

FIGURE 2: Sclerotic blood vessels radially penetrated the germinal centers-lollipop follicles (HE, x100).

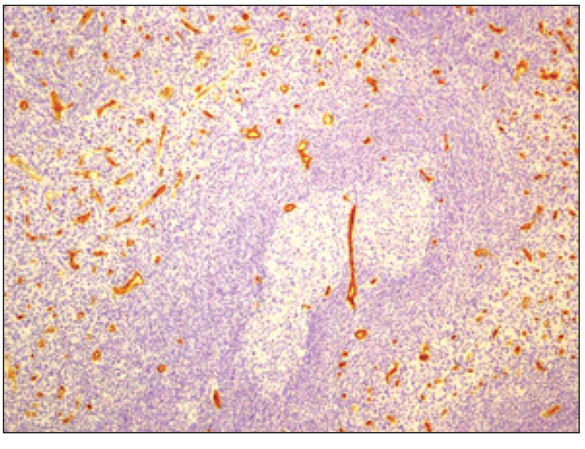

FIGURE 2: CD34 positive blood vessels (CD34, x100).

CD has 2 histologic types; hyaline vascular and plasma cell. Cases with mixed histological features have also been reported.[7,8] There are 2 clinical types: localized and multicentric.[9]

Hyaline vascular CD is mostly observed in young adults with a mean age of 30-40 years and constitutes 90% of all CD cases.[1] Plasma cell variant as the less common form is a diagnosis of exclusion and is not well-defined histologically. The plasma cell variant is frequently associated with constitutional symptoms, cytopenias, hypergammaglobulinemia, splenomegaly, and increased IL-6 levels, and tend to be multicentric.

The microscopic examination of the lymph nodes reveal mostly preserved architecture, follicular hyperplasia in variable rate, and prominent paracortical plasmacytosis.[2]

The wide spectrum of clinical manifestations cause challenge in the diagnosis. The disease has no specific clinical, radiological, or cytological features. It is a diagnosis of exclusion with the aid of histopathological examination. Therefore, it is important for the physicians to be aware of this clinical entity so patients can receive adequate and early treatment. Surgery is the treatment of choice for the solitary form[1], whereas chemotherapy, radiotherapy and steroids are proposed for the multicentric type and inoperable cases.

Radiation therapy (RT) is an appropriate option for masses that cannot be resected by surgery. RT can be a definitive treatment modality for unicentric CD with a succesfull control rate and few complications.[8]

The differential diagnosis includes Warthin tumor, pleomorphic adenoma, schwannoma, hemangioma, lymphangioma, teratoma, sarcoma and metastatic nodes.[6,10] The prognosis of CD is variable and depends predominantly on disease subtype.[2] Because of its unusual location and tendency to mimic other neoplasms, parotid gland CD presents considerable diagnostic difficulties. Radiological findings are beneficial, but not diagnostic.[5] CD is a heterogeneous disorder that can represent a continuous spectrum of disease or several different diseases all together; clinically it might be asymptomatic or could be lethal, The final diagnosis could be achived by histopathological evaluation, The treatment has to be surgery which appears to be curative in local lesions; in inoperable or systemic variants the treatment is radiation therapy and chemotherapy.[10]

It is important to remember this entity for enlarged lymph nodes even in extreme localizations.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Sümeyye Ekmekci, Sermin Özkal;* ***Design:*** *Sümeyye Ekmekci, Gülen Gül, Ebru Çakır;* ***Control/Supervision:*** *Ebru Çakır, Sermin Özkal;* ***Data Collection and/or Processing:*** *Sümeyye Ekmekci, Ülkü Küçük, Suat Kaptaner;* ***Analysis and/or Interpretation:*** *Sümeyye Ekmekci, Ülkü Küçük, Dudu Solakoğlu Kahraman;* ***Literature Review:*** *Sümeyye Ekmekci, Gülen Gül, Ülkü Küçük, Ebru Çakır, Dudu Solakoğlu Kahraman;* ***Writing the Article:*** *Sümeyye Ekmekci, Ebru Çakır, Gülen Gül;* ***Critical Review:*** *Ebru Çakır, Sermin Özkal;* ***References and Fundings:*** *Sümeyye Ekmekci, Gülen Gül;* ***Materials:*** *Sümeyye Ekmekci, Ülkü Küçük, Suat Kaptaner.*

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