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Role of radiotherapy in the management of recurrent castleman's disease

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SUMMARY

Castleman's disease is a benign and rare disease developing in lymph nodes. It is characterized by B-cell hyperproliferation and B-cells produce interleukin-6. This disease has two types; multicentric type often presents with fever, hepatomegaly, splenomegaly or lymphadenopathy, whereas unicentric type is diagnosed occasionally. Multicentric type is treated with combination chemotherapy regimens and unicentric type is treated with surgery and/or radiotherapy.

In this article, a 29-year-old male patient with unicentric type, hyalen vascular Castleman's disease with local recurrence after surgery and treated with radiotherapy is presented with management and related literature review.

Keywords: Castleman's disease, Recurrence, Radiotherapy

ÖZET

Rekürren castleman hastalığının tedavisinde radyoterapinin rolü

Rekürren Castleman hastalığının tedavisinde radyoterapinin rolü

Castleman hastalığı lenf nodlarından gelişen benign ve nadiren görülen bir hastalıktır. B-lenfosit hiperproliferasyonu ile karakterizedir ve B-hücreleri IL-6 üretir. Bu hastalığın iki tipi vardır. Multisentrik tip sıklıkla ateş, hepatomegali, splenomegali ve lenfadenopati ile prezente olur. Fakat unisentrik tip tesadüfen teşhis edilir. Multisentrik tip kombinasyon kemoterapi rejimleri ile ve unisentrik tip ise cerrahi ve/ veya radyoterapiyle tedavi edilir.

Bu makalede 29 yaşında cerrahi sonrası nükseden ve radyoterapi ile tedavi edilen unisentrik, hyalen vasküler Castleman hastalıklı bir olgu tanı ve tedavi aşamaları değerlendirilerek ilgili literatür gözden geçirilmiştir.

Anahtar kelimeler: *Castleman hastalığı, Rekürrens, Radyoterapi*

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Introduction

Castleman's disease (CD) was first described by Benjamin Castleman in 1956 and was classified in the list of rare diseases by ORD (Office of Rare Diseases) of NIH (National Institute of Health) in USA (United States of America). Prevalance of CD is not known but it is estimated to be less than 1/100.000 which means it affects less than 200.000 patients annually in USA (United States of America) (1). CD is separated into unicentric and multicentric types. The most common form is unicentric type which approximately constitutes 80-85 % of all cases. Unicentric type has a more favorable prognosis compared to the multicentric type and often presents incidentally without any symptoms. Multicentric type comprises 15–20 % of all cases with CD and presents with recurrent hyperthermia, hepatomegaly, splenomegaly and lymphadenopathy (2). Additionally, the multicentric type has a potential to progress to malignant lymphoma and has a subtype, namely plasma cell variant, which is associated with HIV infection and HHV–8 infection (3).

CD is classified in noninfectious causes of lympadenopathy in the group of idiopathic and miscellanous causes. The etiology of CD is not clear and the term "giant follicular lymph node hyperplasia" is also used for the disease (4). Male/ female ratio is equal. Unicentric type is usually seen in the 4th decade whereas multicentric type is seen in the 6th decade.

Treatment of unicentric CD is total or partial surgical resection, however, radiotherapy may be delivered in the setting of unresectable disease or as adjuvant therapy. There is no consensus on the management of multicentric CD. A wide range of therapies including corticosteroids, IFN, single or multiple chemoterapeutics, stem-cell transplantation and radiotherapy has been used (5).

The Castleman's disease is rare and accounts one of the rare indications for radiotherapy practice.

Case

A 29-year-old male patient was admitted to the hospital with symptoms of fever and cough. Chest radiogram and subsequent thorax Computed Tomography (CT) revealed a mediastinal mass. The patient was HIV-negative and routine biochemistry was normal. In June 2010, the patient underwent a surgical removal of the mediastinal mass. On microscobic examination, large follicles were scattered in lymphoid tissue. Vascular proliferation along with hyalinization of abnormal germinal centers were shown in follicles. Postoperative pathologic evaluation revealed a hyalen vascular CD (Figure 1). Multiple mediastinal lymphadenopathies in the follow-up CT at three months after surgery revealed local recurrence of the disease (Figure 2). Radiotherapy was planned for the management of recurrent disease. Between September and November 2010, a total radiotherapy dose of 30 Gy was delivered through anterior-posterior fields to the mediastinal region using 18 MV photons in 15 fractions of 200 cGy (Figure 3). Follow-up CT examination of the patient at three months after radiotherapy revealed local control of the disease (Figure 4).

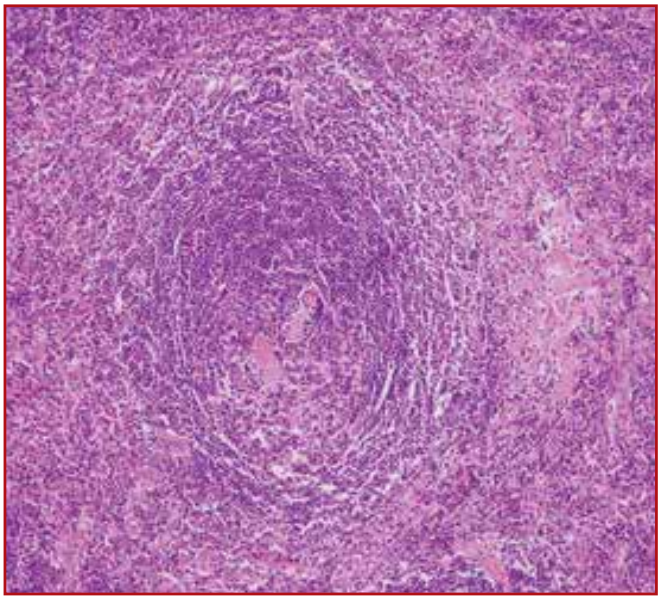


Figure 1. A large follicle with vascular proliferation and hyalinization in germinal center (x200, HE) (arrow showing large follicle)

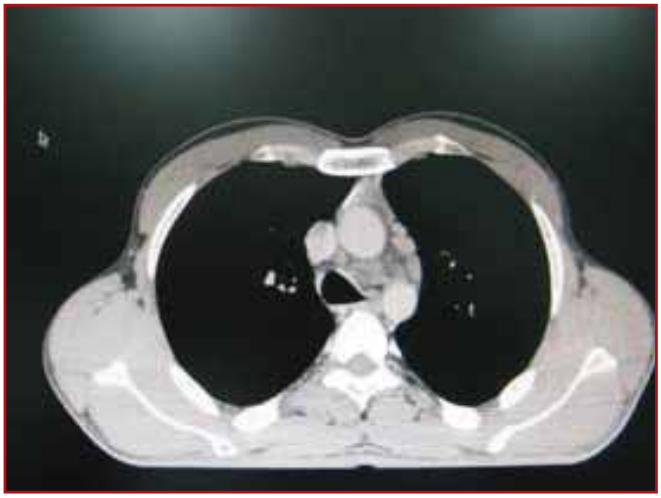
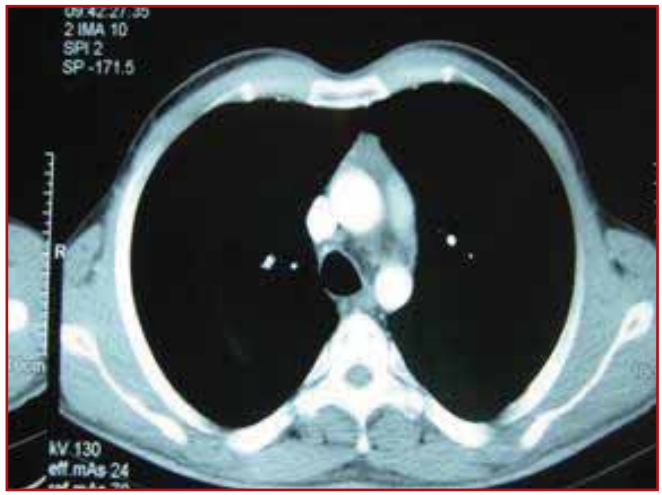


Figure 2. Local recurrence after surgery (arrows showing local recurrence)



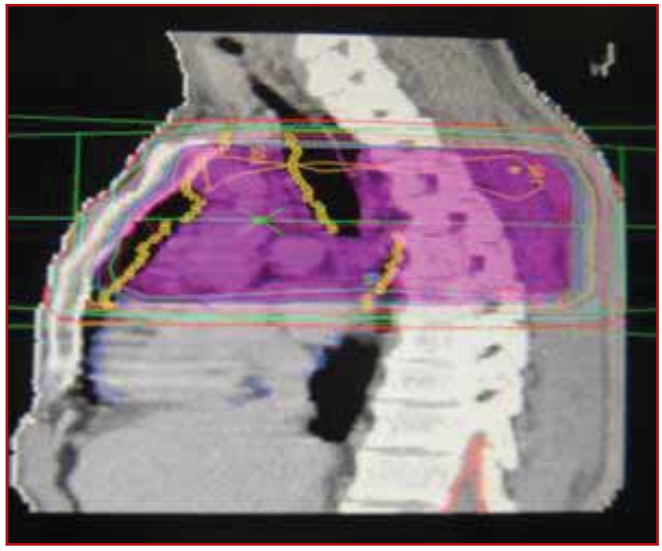


Figure 4. Follow-up CT examination in postradiotherapy 6 months (arrow showing local control)

Discusion

Castleman's Disease is a slowly progressing and rare disease (6). Surgery is the standard treatment in the management of unicentric CD. Radiotherapy may be delivered alone with curative intent as a viable alternative to surgery. Multicentric CD is treated primarily with chemotherapy and the effect of radiotherapy in multicentric CD has yet to be proven (7).

In the review by Rodriguez et. al. including 5 cases, 2 cases were treated with surgery, 1 case was treated with radiotherapy alone, 1 case received postoperative radiotherapy and 1 case was treated with corticosteroids only (8).

For the unicentric CD, the most common site of involvement is mediastinal lymph nodes although it may occur in any lymph node region throughout the body (9). In our patient with CD, the site of local recurrence was the mediastinal lymph nodes detected by the postoperative follow-up CT examination.

Late complications of radiotherapy include pemphigus vulgaris, esophageal and tracheal stenosis (10). However, our patient with CD had no acute or late complications associated with radiation therapy.

In the literature, doses in the range of 30 Gy - 45 Gy have been used for treating unicentric CD (11). The prescribed dose was 30 Gy in 15 fractions in our patient which is consistent with the literature. However, even doses as high as 60 Gy were delivered to treat unicentric CD (12). In the study by Vries et. al., neoadjuvant radiotherapy of 40 Gy was delivered to facilitate surgical resection and downsizing of the disease was achieved at 6 weeks after radiotherapy which allowed complete resection (13). Farruggia et al. reported a case of pediatric CD treated with steroids, chemotherapy, surgery and radiotherapy of 45 Gy (14).

Surgery is the standard treatment for unicentric CD. Radiotherapy is a viable therapeutic option in the treatment of recurrent CD. Given the rarity of CD, multicenter studies including more patients are warranted to further refine the optimal management for this entity.

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