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Short Communication

The Place of Radiotherapy in Castleman's Disease

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Castleman's Disease (CH) is angiofollicular lymph node hyperplasia seen as a result of abnormal proliferation of plasma cells and B-lymphocytes in lymphoid tissues. Etiology is also accused of viruses and interleukin-6. There are clinically unicentric and multicentric, histopathologically, hyaline-vascular, plasma cell and mixed types. The most common unisentric hyaline-vascular type is seen. This type of abdominal, mediastinal, cervical masses are detected. Systemic findings of the plasma cell type and organomegaly are common. Most patients present with nonspecific properties such as symptoms of an inflammatory disease. Fever is an almost universal symptom and most patients present with night sweats, weight loss, malaise or fatigue and lymphadenopathy. During the physical examination or when imaging studies are performed, they grow clinically with the enlarged lymph node. Diagnosis is made by histopathological examination. Although CH is not a malignant disease, it may be associated with malignant diseases or it may be transformed into malignant diseases over time. Therefore, long-term follow-up of patients is important [1-5].

Unisentric disease is typically localized, symptoms are minimal. It is treated with surgery alone. Rarely, it may not be resectable due to mass size or localization. In such patients, radiotherapy RT is a good treatment option because of high response rates (complete response in about 50% of cases). Systemic treatments for multicentric CH are considered in symptomatic local disease that cannot be treated by surgery or radiotherapy [1-4]. Talat et al. have performed a systematic review and meta-analysis of 404 patients with CH between 1954 and 2009. In 278 unicentric Castleman patients, 249 patients underwent surgical treatment, 13 patients underwent combined therapy (surgery and chemotherapy) and 16 patients were treated with immunosuppressive therapy alone. Ten years follow-up revealed deaths from 13 patients. The 3-year disease-free survival rate was 89,7% and the 5-year disease-free survival rate was 81,2%. They reported that surgical excision in patients with unicentric CH is safe and should be accepted as the gold standard for treatment [6]. Five retrospective studies and one case series reported the use of RT in the treatment of unicentric CH bir when the literature was reviewed. Three of these studies were the studies that included patients who had undergone radiotherapy because of not being resected. As a result of all these studies, a significant reduction in tumor size was reported with RT. However, possible complications and high recurrence rates have been reported [2,3,5-7]. In a retrospective study by Keller and colleagues, they concluded that radiotherapy in patients with unresectable, unicentric CH lesion would reduce the tumor and result in regression of symptoms [8]. Uysal et al. looked at the efficacy of 11 RT with inop and recurrent CH. 54.5% of the patients underwent total excision and 45,5% of them underwent incisional biopsy. Approximately 63,6% of patients with CH were treated with RT and 27,3% with chemotherapy alone, and one patient (9,09%) received both RT and chemotherapy. Three-year survival was 83%, and 3-year disease-free survival was 91%. Acute toxicity was observed in two patients receiving 45 Gy. No late toxicity due to RT [2].

There is no standard treatment in multicentric CH. The choice of treatment is based on the patient's HIV/HHV-8 positivity status and then the clinical aggressiveness of the disease. In multicentric CH, monoclonal antibodies against single/combined chemotherapeutics, interferon, thalidomide, interleukin-6 and CD-20, and antiviral drugs are used. The choice between these agents depends on the applicability of the primer and the experience of the clinician [1-3]. Tomita et al. 29-year-old female patient with cervical multicentric, plasma cell subtype CH planned after treatment with prednisone. With Intensity Modulated Radiotherapy (IMRT), 44 Gy has been treated with 22 fractions. Recurrence of cervical lymph nodes was not observed four years and 3 months after IMRT. Parotid function was observed dramatically in quantitative saliva scintigraphy 3 and 12 months after IMRT. Radiotherapy has been reported to be an option for multicentric CH and is an effective way to minimize xerostomia in head and neck lesions of IMRT [3]. In another case report, tocilizumab, an IL-6 inhibitor, was started in a negative multisentric Castleman patient for HIV and HHV-8 who had failed surgery and had no response to radiotherapy and chemotherapy. They reported no response despite 2 months of use [4].

In conclusion, CH is a rare but life-threatening disease. One of the treatment options of CH, RT information is based on several retrospective studies in the literature, small case series and expert opinions. Therefore, a standard treatment is not available and clinical practice is changing. There is a need for studies to evaluate the indication, dose, toxicity and efficacy of RT with multicenter prospective studies.

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