

Radiotherapy for orbital lymphoma

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Abstract

Lymphoma of the orbit is a rare presentation of non-Hodgkin's lymphoma (NHL). According to the data from the literature, orbital lymphoma represents 8% of all extranodal NHL and about 1% of all NHL [1-3]. Symptoms which are usually caused by orbital lymphoma are proptosis, periorbital swelling, conjunctival (salmonpink) swelling, diplopia, and conjunctival redness and irritation [4]. Radiotherapy is widely used as the primary treatment for orbital lymphoma. Radiation in those cases is often technically challenging, because special effort is made to preserve the integrity of the orbit without compromising local control [1]. The studies have shown that a local control rate is 95-98% for low grade orbital lymphomas [1,5]. The localized primary orbital lymphomas respond well to radiotherapy. The advantages of the radiotherapy treatment are a high-local control rate and durable disease-free status. Carefully planned orbital radiotherapy is well tolerated by patients and obligatory use of a lens shield contributes to greatly reducing the incidence of complications.

Keywords: Conjunctiva, lymphoma, orbital metastasis, radiotherapy

Introduction

Metastasis in the eye are unusual. One of the possible manifestations of metastasis of the eye is an orbital metastasis which occurs as one that occurs within the space between the eyeball and bony orbital walls [4]. Metastases occur in 2% to 3% of patients with cancer [1,4]. Usually orbital metastases are unilateral. Furthermore, lymphoid masses comprise 10-15% of all orbital tumors and up to 55% of malignant orbital masses [6]. Lymphoma of the orbit is a rare presentation of non-Hodgkin's lymphoma (NHL). According to the data from the literature, orbital lymphoma represents 8% of all extra nodal NHL and about 1% of all NHL [1-3]. Orbital lymphoma may be localised in the conjunctiva, lacrimal gland, or in retro-orbital tissues. Hystology is usually MALT or low-grade lymphoma and occasionally and intermediate grade lymphoma [7]. Symptoms which are usually caused by orbital lymphoma are proptosis, periorbital swelling, conjunctival (salmonpink) swelling, diplopia, and conjunctival redness and irritation [4]. Radiotherapy is widely used as the primary treatment for orbital lymphoma. However, radiation therapy gives low local recurrence rates and also ability to preserve ocular function [1,8,9]. Radiation in those cases is often technically challenging, because of the effort to preserve the integrity of the orbit without compromising local control [1].

One of the first cases of orbital metastasis was reported by Horner in 1864 [4]. Generally, radiation treatment is individualized based on the location and extent of orbital involvement, as well as the patient's overall clinical condition [1]. The most important characteristic of radiation treatment lies in the fact it is directed at cure and in same time preserving vision and

the integrity of the orbit. Extensive surgery should therefore be avoided. Treatment with an anterior X ray field or electron beam provides satisfactory therapy for anterior lesion limited to the eyelid or bulbar conjunctiva, with the advantage of sparing orbital structures compared to the use of a megavoltage photon beam. If an anterior ortovoltge field is used, a small lead eye shield can reduce the dose in a lens of less than 5-10% [10,11]. For unilateral retrobulbar tumors, a two field technique using megavoltage photon beams (4-6 MV) is appropriate. A pencil shield placed in both fields will spare the lens. Alternative to the pencil shield protection method is an isocentric technique with two oblique (wedged) fields with a shield inserted in both fields, with the patient looking at the shield for each treatment field [4]. The prescribed radiation dose is 20-30 Gy in 10-20 daily fractions. The studies have shown that a local control rate is 95-98% for low grade orbital lymphomas [1,5]. For patients with larger intermediate and high grade tumors, recommended treatment is based on chemotherapy (e.g., CHOP for III courses) followed by radiotherapy. The role of chemotherapy has not yet been clearly defined, probably due to the wide range of histological types and grades of NHL. The radiotherapy alone should be an option in a low-grade lymphoma stage I and II in the elderly patients. However, distant metastases rating from intermediate to high-grade tumors are 40-67%. This is why patients with advanced disease should be initially treated with chemotherapy followed by localized radiation therapy to achieve temporary disease control [12,13]. The desirable dose should be 30-35 Gy divided in daily fractions with 1.5-1.75 Gy. Furthermore, for more aggressive disease the prescribed radiation doses have to be not less than 40 Gy [14]. Higher

doses are not required and their use results in higher acute and long-term morbidity. The most common complications of radiotherapy in orbital lymphoma are cataract, keratitis and dry eye, especially if the dose is 40 Gy or more. The tolerance dose of the lacrimal gland is 40 Gy in 20 daily fractions. According to the literature the damage of the optic nerve and retina should not occur if the radiation dose is less than 40 Gy.

Therefore, in order to determine the optimal dose, which would ensure local control while minimizing morbidity, the additional data encompassing more patients is needed [1]. We describe a 68-year-old patient with a five year history.

Case report

We present a 68-year-old patient with a five year history of enlarged cervical lymph nodes on both sides of the neck with evidence of hepatomegaly and splenomegaly. After biopsy of the lymph node in the left axilla in the patient was diagnosed with diffuse large B-cell lymphoma. PET/CT did not describe any PET avid mass, except in the orbital mass. During the treatment, patient received chemotherapy with the protocols R-CHOP VIII cycles PR, HTH CVPEVI cycles. Since April 2012 our patient has had the palpable tumor mass in the conjunctiva of the right upper eyelid. The tumor was movable, without inflammations signs and approximately of 0.5 cm in diameter. The patient then received by HT ICE protocol in the fourth series. In July 2012 there was a progression of both eyelids which is resistant in the given corticosteroid therapy. CT endocranium in the end showed only suspected bilaterally infiltration of the lacrimal glands. Ultrasonography of orbits and eyes has shown that at the front upper part of the orbit the change without breaking the rear parts of the orbit. The change on the right side was more prominent. The patient received radiotherapy with the prescribed dose of 18Gy divided in 10 daily fractions. The local control was excellent and there were no complications. Control examinations showed that there is no appearance of the recidive. Patient has been in disease-free status without any complications after radiotherapy, for one year (Figures 1-4 and 5-6).



Figure 1. Before treatment.

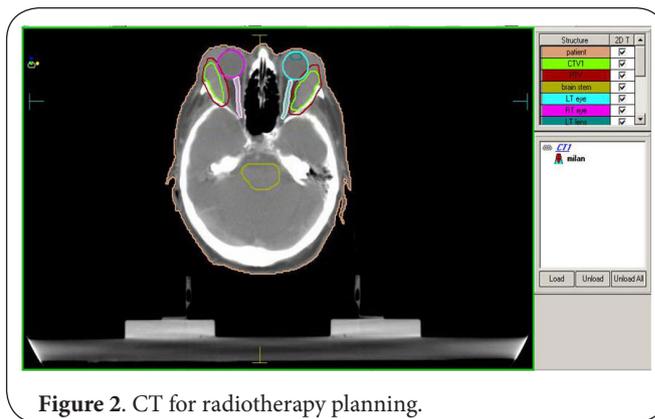


Figure 2. CT for radiotherapy planning.

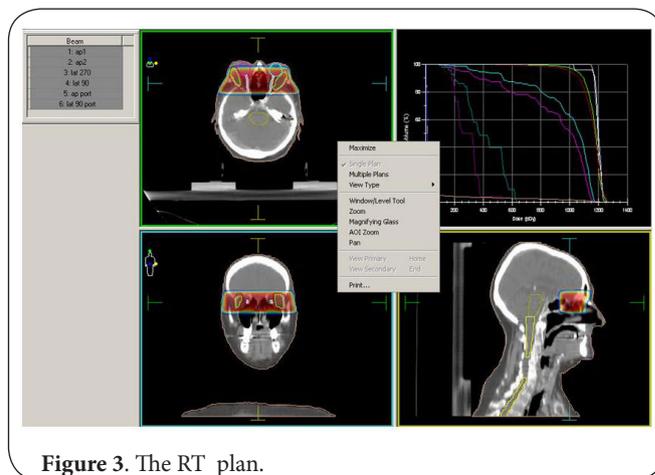


Figure 3. The RT plan.

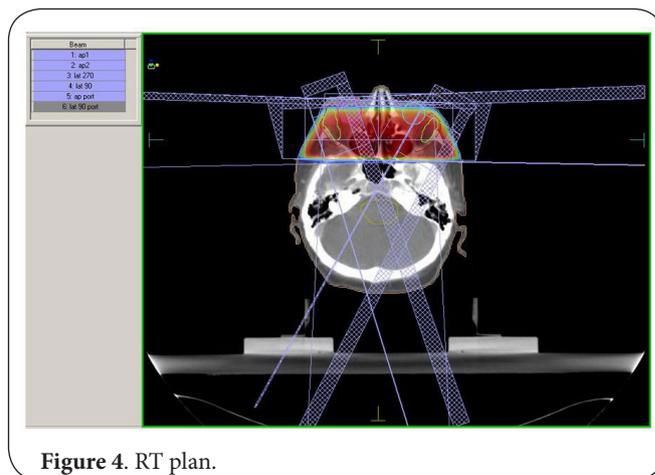


Figure 4. RT plan.

Conclusion

In conclusion, the localized primary orbital lymphomas respond well to radiotherapy, with a high-local control rate and durable disease-free status. In the application of the radiotherapy treatment, expected complications are dry eyes, keratitis, cataract, and retinopathy. However, carefully

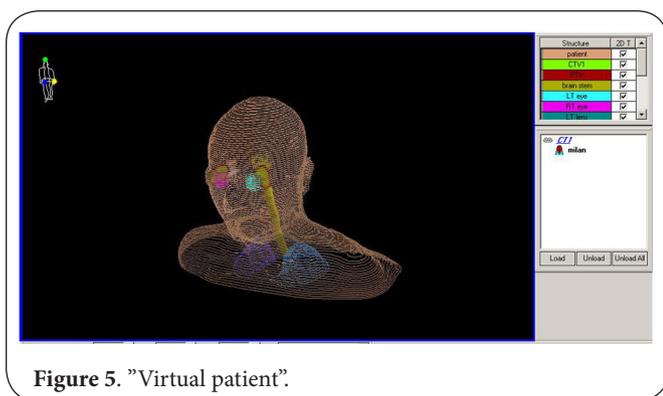


Figure 5. "Virtual patient".



Figure 6. After treatment.

planned orbital radiotherapy is well tolerated by patients and obligatory use of a lens shield contributes to greatly reducing the incidence of cataract. The lowest effective dose for orbital lymphoma which should be used may be between 15 to 30Gy. The recommended daily dose is 1.8Gy and inevitably, the newer technology in radiotherapy may offer better sparing of the eye.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Authors' contributions	ME	MM
Research concept and design	✓	✓
Collection and/or assembly of data	✓	✓
Data analysis and interpretation	✓	✓
Writing the article	✓	✓
Critical revision of the article	✓	✓
Final approval of article	✓	✓
Statistical analysis	✓	✓

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