Secondary Bilateral Orbital Involvement from Primary Non-Hodgkin Lymphoma of the Cheek

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We describe a patient with oculomotor nerve palsy due to secondary orbital infiltration from the primary malignant lymphoma of the cheek. The patient was a 78-year-old female who had non-Hodgkin lymphoma (diffuse large B cell lymphoma [DLBCL]) of the cheek. The patient received chemotherapy and local radiation therapy. The combined treatment brought about complete remission.

About 6 months after the last treatment the patient began to have left blepharoptosis and impaired vision. Findings from ophthalmological and neurosurgical examinations suggested no intraorbital or intracranial lesions. Repeated MRI and CT scans also showed no such lesions. One month later, the patient suddenly had a left oculomotor disturbance, diplopia and exophthalmus, followed by right oculomotor nerve palsy. An MRI revealed bilateral intraorbital tumors. Recurrence at the orbital tissue of malignant lymphoma originated from the left cheek appeared to cause the ophthalmological symptoms. Salvage chemotherapy was performed and her ocular symptoms were recovered. However, the patient died approximately 10 months after recurrent orbital tumor onset.

INTRODUCTION

While primary orbital malignant lymphoma is uncommon, secondary involvement in the orbit from other primary malignant lymphoma sites is particularly rare [4,8]. There are no reports of such cases from the maxillofacial fields. This study describes a patient who developed secondary bilateral orbital involvement after initial treatment for primary non-Hodgkin lymphoma of the cheek.

CASE REPORT

A 78-year female became aware of a bean-sized tumor on the mucous membrane of her left cheek in July 1996, and visited a local hospital where the tumor was completely excised on October 4. Based on the histopathological examination of the resected tumor, the patient was diagnosed as having diffuse large B cell lymphoma (DLBCL) (Fig.1-a, b). During follow-up at the hospital, a tumor recurred at the same site and began to grow. She was referred to the department of Oral and Maxillofacial Surgery on January 20, 1997. The patient had a surgical history for a perforated duodenal ulcer in 1984 and hospitalization to treat a cerebral infarction in 1993. At the first examination, an elastic hard tumor 30 mm by 18 mm with well-defined margins was noted in the left cheek (Fig.2). This lesion was neither painful nor tender. Waldeyer’s ring was intact. No swelling was
detected in cervical, axillary or inguinal lymph nodes. Blood examinations showed a slight decrease in white blood cells (3,290/μl) and platelets (106×10³/ml), but findings were otherwise normal. No other tumors were noted on chest x-ray, chest and abdominal CT scans, gallium scintigraphy or bone marrow examinations.

The first course of CHOP (cyclophosphamide, 750 mg; doxorubicine, 50 mg; vincristine, 1.4 mg; prednisolone, 100 mg×5) therapy began on January 28, 1997. The patient received local radiation therapy (42Gy) between February 13 and March 1. The combined treatment brought about complete remission. The patient began to have left blepharoptosis and impaired vision at the beginning of November. However, ophthalmological examinations did not clarify the cause of these symptoms. Findings from neurosurgical examinations suggested no intraorbital or intracraniatal lesions, and repeated cranial MRI and CT scans also showed no such lesions. About 50 days later, the patient had a left oculomotor disturbance, diplopia, and exophthalmos, followed by right oculomotor nerve palsy (Fig.3). An MRI on December 22 revealed bilateral intraorbital tumors (Fig.4). Scintigraphy showed no accumulation of gallium in other regions, and a bone marrow examination showed no abnormal findings. Recurrence at the orbital tissue of malignant lymphoma originated from the left cheek appeared to cause the ophthalmological symptoms. Although no definitive metastasis to the other regions had been seen in scintigraphy, the possibility of the multiple metastasis could not be denied. Therefore, salvage chemotherapy began in January 1998. A radiological examination on April 30 showed no tumor and the ophthalmological symptoms were relieved, so the patient was discharged. However, malignant lymphoma soon recurred throughout the body. The patient died about 10 months after recurrent orbital tumor onset.
SECONDARY BILATERAL ORBITAL INVOLVEMENT FROM PRIMARY NHL

DISCUSSION

The incidence of relapse according to the primary site in stage I and II extranodal non-Hodgkin lymphoma of head and neck, is very similar, such as Waldeyer’s ring (43.3%), nasal cavity (28.6%) and oral paranasal sinuses (36.4%) [2]. Most patients with relapses developed extranodal distant site extension outside the original treatment fields. The bone, with muscle or skin involvement, was the most frequent site [2,4,5]. Gastrointestinal involvement occurs in a greater number of patients with lymphomas of Waldeyer’s ring [3]. However, to our knowledge, there have been no reports secondary orbital involvement from primary non-Hodgkin lymphomas histologically correlated, as graded by the Working Formulation [6]. Therapy combining radiation and chemotherapy is recommended for higher grade lymphomas [5]. Our patient was histopathologically diagnosed as having diffuse large B cell, intermediate grade lymphoma as classified by the Working Formulation. She was treated with irradiation combined with CHOP therapy. Seven months after the last treatment, a recurrence was discovered in the orbit.

The most common symptoms of orbital malignant lymphomas are subacute development and progressive painless swelling, proposes and diplopia [7]. Our patient’s main symptom was blepharoptosis of the left eye. An ophthalmologist first diagnosed her with oculomotor nerve palsy. However, the cause of paralysis was not detected in the orbit, so a pathological problem in the brain was suspected. Even so, CT and MRI just after the patient reported blepharoptosis did not reveal any mass in the brain or orbit. When she became aware of proptosis about 50 days later, bilateral orbit tumors were recognizable with MRI. MRI accurately determines the extension of lymphoma, but enhanced lesions may be obscured due to the proximity of fat in the orbits [1]. The high intensity of fat in T1-weighted SE sequences has limited the usefulness of Gd-DTPA in orbit imaging. We did not detect tumors in the orbital fat in T1-weighted images using Gd-DTPA in this particular patient during the initial relapse. However, it has been reported that contrast-enhanced fat suppression imaging eliminates these shortcomings [1].

In summary, we reported a patient with secondary bilateral orbital involvement from primary non-Hodgkin lymphoma of the cheek. Even though this type of infiltration of malignant lymphoma is quite rare, clinical physicians should keep in mind the possibility of such infiltration from primary malignant lymphoma in maxillofacial fields.

REFERENCES