Case Report

Mantle Cell Lymphoma in a Lacrimal Gland in a Female and a Review of the Literature

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Abstract

Purpose: To report a rare case of Mantle cell lymphoma in lacrimal gland and review of the literature

Case report: We report a case of a 59-year-old female who presented with an upper eyelid mass in the right eye for 3 months, without pain and irrigation. A computerized tomography (CT) scan showed a mass in the bilateral lacrimal gland region, more significant in right eye. The patient underwent a lacrimal gland mass excision surgery and diagnosis of mantle cell lymphoma by histopathology. Immunochemistry for CD20, CD79a, CD5, and CyclinD1 was positive. She was recommended to the Shantou cancer hospital for chemotherapy.

Conclusion: Mantle cell lymphoma is a rare type of malignant lymphoma, over expressing CD5 and cyclin D1 antigens, which distinguishes it from other B cell lymphomas. (***Eye Science 2014; 29:178-181***)

Keywords: mantle cell lymphoma; Lacrimal gland

Introduction

Primary ocular adnexal lymphomas represent approximately 10% of orbital neoplasms and 8% of extranodal lymphomas. They are localized in the orbit, the conjunctiva, and the eyelids[1,2]. Mantle cell lymphoma (MCL), a rare type of malignant lymphoma with origin in peripheral B cells of the inner mantle zone, accounts for 2 to 7% of lymphomas. MCL of the ocular adnexa is very rare. It may be primary or secondary and affect patients in the sixth or seventh decade of life. The orbit is the most commonly involved site, followed by the lacrimal glands and eyelids[3,4]. In this study, we report a rare case of lacrimal gland mantle cell lymphoma in a 59-year-old female.

Case report

A 59-year-old female patient presented with an upper eyelid mass in the right eye for 3 months, without pain and irrigation. The patient recalled realizing a mass as big as a pigeon egg, gradually growing in size, without swelling the eyelid, without eye redness, without double vision, and without vision loss. Extraocular movements were normal. No proptosis presented.

Clinical examination revealed right supraorbital swelling with round lymphadenopathy in the lacrimal gland regions. On palpation, the tumor was round, firm, smooth, not tender, well-circumscribed, and approximately 20 mm×15 mm in size (Figure 1). A computerized tomography (CT) scan showed a mass in the bilateral lacrimal gland region, more significant in right eye(Figure 2). A B scan of the liver and spleen was normal.

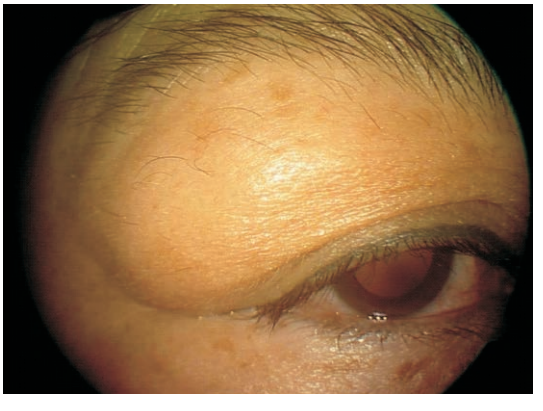

Figure 1 The appearance of the patient. Right supraorbital swelling with round lymphadenopathy in the lacrimal gland regions.

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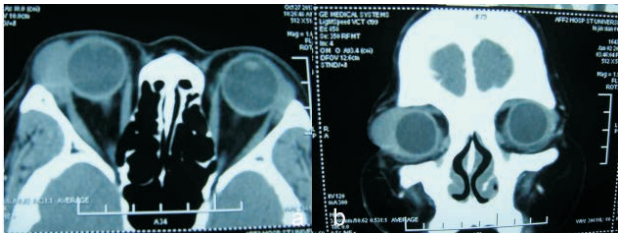

Figure 2 Computerized tomography (CT) scan. Mass in bilateral lacrimal gland region, more significant in right eye (a. horizontal position, b. coronal position)

The patient underwent a lacrimal gland mass excision surgery. During the operation, we performed biopsy of the mass. The mass conformed to the shape of the globe with black-violet color, soft texture, and measured about 24 mm×26 mm×6 mm (Figure 3). Histological examination of the mass showed infiltration by a monomorphic cell population composed of small- to medium-sized lymphocytes with irregular, indented, or cleaved nuclei, condensed chromatin, and scant cytoplasm. The growth pattern was diffuse and partially nodular. Immunohistochemical study revealed that tumor cells were CD20, CD79a, CD5, CyclinD1, Ki-67 30%, and Pax-5 positive and TdT, CD3, CD43, CD68, CD 23, and CD10 negative (Figure 4). The final diagnosis was B-cell non-Hodgkin's lymphoma of the MCL type.

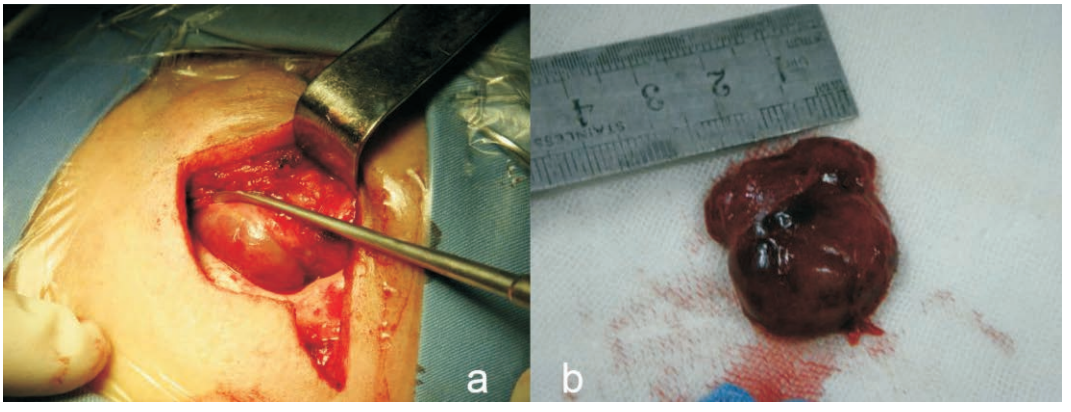

Figure 3 The general appearance of lacrimal gland mass. a. The mass located in the lacrimal gland fossa. b. The mass conformed to the shape of the globe with black–violet color, soft texture, and measured about 24 mm×26 mm×6 mm.

The patient was recommend to Shantou cancer hospital for chemotherapy.

Discussion

Mantle cell lymphoma (MCL) is a rare type of malignant lymphoma with origins in the peripheral B cells of the inner mantle zone. MCL was first described by the German pathologist Lennert et al. in the 1970s as diffuse small neoplastic cells with bizarre and cleaved nuclei and no admixture of blast cells[5]. In 1992, the International Lymphoma Study Group recognized MCL as an independent malignant lymphoma[6]. Until now, much work has been done to characterize this subtype of B-cell non-Hodgkin lymphoma (NHL) both clinically and histopathologically. MCL, which represents 6% to 7% of all NHL, is now evident as a disease of the elderly associated with a male preponderance, a tendency to be seen initially with widespread disease, and an aggressive clinical course. Immunohistochemical and cytogenetic studies have allowed more reliable diagnosis on the basis of over expression of cyclin-D1 and identification of t(11;14)[4,7]. In the periocular region, it makes up 2% to 7% of lymphomas[3,4]. A few cases that have been briefly described in large series of ocular adnexal lymphoma give the impression of advanced disease with poor prognosis; however, the overall clinicopathologic features of MCL in this location have not been well described.

The clinical features of MCL share no particularity. The most common presenting symptoms are ptosis, lid lump, proptosis, and diplopia, whereas the most frequently encountered clinical signs were proptosis, lid mass, ptosis, lacrimal gland mass, and chemosis[8].

The diagnosis of MCL is based on morphologic

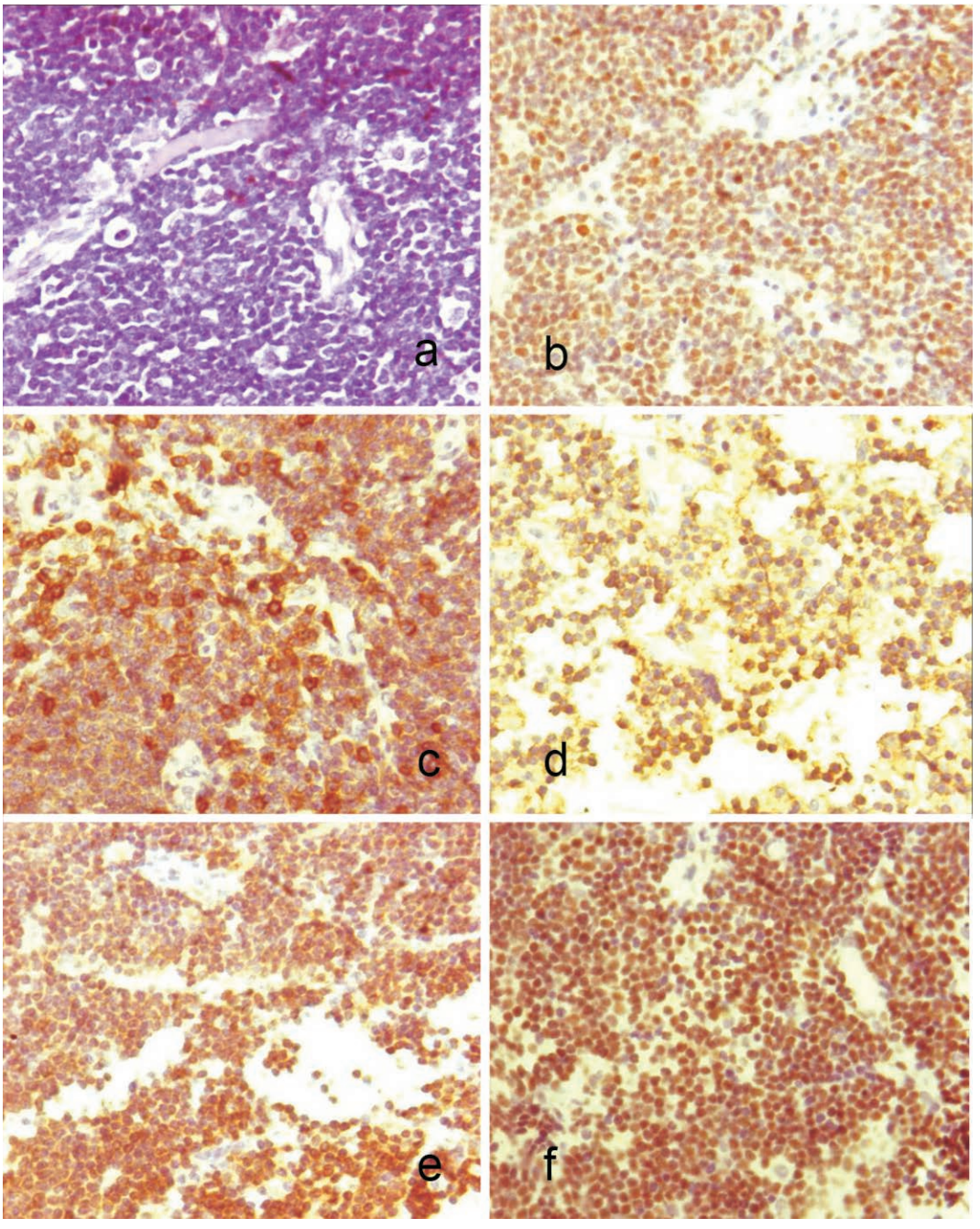

Figure 4 histological examination of the mass. HE stain shows that infiltration by monomorphic cell population composed of small- to medium-sized lymphocytes with irregular, indented, or cleaved nuclei, condensed chromatin, and scant cytoplasm. The growth pattern was diffuse and partially nodular (a,200×). Immunohistochemical study revealed that tumor cells were CyclinD1 (b), CD5(c), CD20(d), CD79a(e), and Pax-5(f) positive(400×).

and immunophenotypical criteria. Histologically, the case we reported showed the main characteristics of MCL: monotony of lymphoid infiltrate and the absence of blast cells. Moreover, the immunophenotype was positive expression of CD5 and cyclin D1 antigens and was negative for CD23 and CD10, which was consistent with MCL. Expression of CD5 and cyclin D1 antigens can distinguish MCL from other small B-cell lymphomas. Cyclin D1 is particularly helpful in the differential diagnosis of MCLs, especially in CD5-negative cases, a form that appears to be frequent in the orbit. This protein is ex-

pressed in 80–90% of MCLs and is absent in all other B-cell lymphomas, with rare exceptions[4,9].

Controversies inevitably arise when treating MCL. The main treatments include chemotherapy and radiotherapy for local control in most patients. Different regimens of chemotherapy medications have been used, such as R-CHOP(rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). Other treat ments such as photodynamic therapy, stem cell transplantation, and anti-VGF were also reported[4,8,10].

The prognosis of ocular MCL is poor, with a median overall survival of 57 months, because 80% of cases are stage Ⅲ/Ⅳ[4].

In conclusion, we reported a case of MCL in the orbital region and reviewed literature about MCL. MCL in the orbital and adnexal region is rare and usually presents in elderly males. The diagnosis of MCL was mainly based on histopathology. Treating MCL is controversial and the disease has a poor prognosis.

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