Primary Follicular Lymphoma of the Conjunctiva in a 12 Year-Old Male

Taghipour Zahir Sh MD 1, Miratashi S A MD2, Nazemian M MD3, Zand S MD3

1. Associate Professor of clinical and surgical pathology, Shahid Sadoughi university of Medical Sciences, Yazd, Iran.
2. Assistant Professor of ophthalmology, Shahid Sadoughi University of Medical Sciences, Yazd, Iran.
3. Medical Student, Shahid Sadoughi University of Medical Sciences, Yazd, Iran.

Received: 15 December 2012
Accepted: 6 March 2013

Abstract

Background
Follicular lymphoma (FL) is the second most common adnexal lymphoma of the eye that almost all of them are reported in elderly patients. Primary FL of the eye has been reported in only two children. Pediatric FL appears to be biologically distinct from typical adult FL. In cases without other organ involvement excision alone with close monitoring is a treatment of choice and the prognosis is excellent.

Case presentation
A 12 year -old male with a nodular lesion involving the inner canthus of the right eye was admitted to ophthalmology clinic. The lesion was painless and the nodule size gradually increased over a period of 6 months. Excisional biopsy demonstrated follicular lymphoma composed of neoplastic lymphoid cells arranged in follicular pattern without germinal centers. Neoplastic cells were positive for Bcl2 and CD20.

Conclusion
Although follicular lymphoma of the conjunctiva is rare in childhood but it could be considered as one of the differential diagnosis in this age group.

Keywords
Lymphoma, Follicular; Conjunctiva,
infiltration of neoplastic cells were not seen. Primary conjunctival follicular lymphoma without other organ involvement was confirmed, and only monthly patient examination was recommended, and till now after nine months no recurrence was detected. Patient is in good health conditions.

**Discussion**

Lymphoma is the most common orbital malignancy (3). Patients usually present with mass like lesion, pain, eye irritation, ptosis, excessive tear production or dropping eyelid (1-3). Most of them are localized at presentation and does not involve other organs. Most ocular lymphoma occurs in old age (5,6), and almost all of them are low grade B-cell lymphoma (2,4). FL is the second most common adnexal lymphoma (2), but almost all of them reported in elderly patients and only a few children with primary FL of the eye have been reported. One of them was a 6 year-old boy with 3mm mass in the conjunctiva which resected with clean surgical margin, and remained untreated. He was monitored closely and remained cancer free for 3 years till now (2). The other one was an 11 year-old girl with 25 mm mass involving the caruncle of the left eye underwent excisional biopsy and following the diagnosis, after staging studies there was no evidence of outside involvement, but based on unusual location of lesion, local radiation was administered over 28 days. She was cancer free till 2006 (7). FL is a member of lymphoma groups originated from germinal center-derived B cells. The main histopathological characteristics are recapitulation of lymph node follicles, lack of tangible body macrophages, crowded follicles of more than usual uniformity and lack of normal germinal centers (1-10). Lymphoid follicles in FL composed of small irregular cells both small cleaved cells or centrocytes admixed with large cells (both large cleaved and non-cleaved cells) (2-8). FL cells are positive for CD19, CD20, CD22, CD23 and CD10 (approximately 30 percent) or of bcl-2 protein expression (approximately 10 percent) and a high rate of apparent cure (9). The differential diagnosis of pediatric-type FL includes follicular hyperplasia and marginal zone lymphoma with prominent follicular colonization. Particularly follicular hyperplasia is much more common in young patients than FL. For differentiation of FL from reactive follicular hyperplasia, bcl2 and bcl6 are helpful markers, and FL cells are positive for these two markers (10). Although treatment of ocular follicular lymphoma has not been studied, but it appears that children with FL may do well following excision alone (without other treatments) and just close monitoring. The prognosis is excellent. We just follow up our patient every month, and until now (9 months after treatment) he remained tumor free.

**Conclusion**

Primary follicular lymphoma of the conjunctiva is a extremely rare tumor in children and should be differentiated with reactive lymphoid hyperplasia. For differentiation of these two lesions from each other, IHC has a significant rule.

**Acknowledgment**

The authors wish to thank IJPHO editor team.

**Conflict of interest**

The authors have no conflict of interest.

**References**

1. Bessel EM, Henk JM, Wright JE, Whitelocke RA. Orbital and conjunctival lymphoma: Treatment and prognosis. Radither Oncol.1988; 13:237-44.
2. Gaffar M, Thebpatiphat N, Przygodzki R, Jacobs S, Miller MM. Primary follicular lymphoma of the conjunctiva in a 6-year-old child. J Aapos. 2010; 14(6):538-40.
3. Freeman C, Berg J, Cutler S. Occurrence and prognosis of extranodal lymphomas. Cancer.1972; 29(1):252-60.
4. Ferry J, Fung C, Zukerberg L, Hasserjian R, Preffer F, Harris N. Lymphoma of the ocular adnexa: A study of 353 cases. Mod Pathol.2006;19:225A.
5. Ohtsuka K, Hashimoto M, Suzuki Y. A review of 244 orbital tumors in Japanese patients during a 21-year period: origins and locations. Jpn J Ophthalmol. 2005; 49(1):49-55.
6. Rosado MF, Byrne GE Jr, Ding F, Fields KA, Ruiz P, Dubovy SR, et al. Ocular adnexal lymphoma: a clinicopathologic study of a large cohort of patients with no evidence for an association with Chlamydiapipettii. Blood.2006; 107(2):467-72.
7. Jenkins C, Rose GE, Bunce C, Wright JE, Cree IA, Plowman N, et al. Histological features of ocular adnexal lymphoma (REAL classification) and their association with patient morbidity and survival. Br J Ophthalmol.2000; 84(8):907-13.
8. Karadeniz C, Bilgiç S, Ruacan S, Sarıaiğlu F, Büyükpamukçu M, Akyüz C, et al. Primary subconjunctival lymphoma: An unusual presentation of childhood non-Hodgkin's lymphoma. Med Pediatr Oncol.1991; 19(3):204-7.
9. Taddesse-Heath L, Pittaluga S, Sorbara L, Bussey M, Raffeld M, Jaffe ES. Marginal zone B-cell lymphoma in children and young adults. Am J Surg Pathol.2003; 27(4):522-31.
10. Swerdlow SH. Pediatric follicular lymphomas, marginal zone lymphomas, and marginal zone hyperplasia. Am J Clin Pathol. 2004; 122 Suppl:S98-109.

Figure 1. Lymphoid cells infiltrated the conjunctival stroma. Conjunctival tissue involved by tumoral lesion composed of neoplastic lymphoid cells arranged in follicular pattern without germinal center X10

Figure 2. Follicular lymphoma composed of small cleaved and centrocytic lymphoid cells admixed with large cells X20

Figure 3. CD20 positive lymphoid cells

Figure 4. Lymphoid cells positive for Bcl2