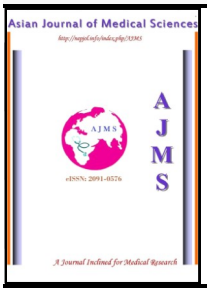
Asian Journal of Medical Sciences 1 (2010) 87-90

ASIAN JOURNAL OF MEDICAL SCIENCES



CT and MRI features of Primary Orbital Lymphoma: review of 14 cases

Jin Chen Wang[1], Rana Netra[1], Wang Yuan[1], Ma ShaoHui[1], Li Min[1], Zhang Ming[1]

[1]First Affiliated Hospital of Xi'an Jiaotong University, NO.277. Yanta West Road, Xi'an 710061, P.R. China

Abstract

Objective: To assess the computed tomography (CT) imaging and magnetic resonance imaging (MRI) features of primary orbital lymphoma and to establish a differential diagnosis.

Material & Methods: Fourteen patients (9 male and 5 female) suffering from primary orbital lymphoma underwent CT scanning with and without contrast. Pre- and post-contrast MRI scans were performed in 10 patients. Their ages ranged from 1.5 to 65 years (average age: 36 years). The course of diseases varied from 2 months to 16 years.

Results: The diagnosis was confirmed pathologically in all cases. Lesions were bilateral in 4 cases and unilateral in 10 cases. Periorbital preseptal tissues were involved mainly in the upper lateral quadrant of the orbit. Focal masses were observed in 5 patients. Lesions that infiltrated the lacrimal gland, adjacent extraocular muscles and intraorbital tissues, along with the extraocular muscles, were found in other patients. The tumors that had advanced into intraorbital tissues and had involved extraocular muscles, displayed either a 'casting sign' or a 'ring sign'. Isodense soft-tissue masses with clear demarcation were found on plain CT film and lesions displayed low to intermediate signal intensity on T1- weighted MRI and relatively low or high signal intensity on T2-weighted MRI. The lesions demonstrated homogeneously marked enhancement on CT and MRI with contrast medium.

Conclusion: CT imaging and MRI - particularly MRI - can demonstrate the location, configuration, inner structure and characteristic manifestations of the orbital lymphomas. This may assist the pre-operative diagnosis of these lesions.

Key Words: Orbital tumor; Lymphoma; Pseudotumor; Computed tomography; Magnetic resonance imaging

1. Introduction

Primary lymphoma of the orbit is an uncommon form of non-Hodgkin's lymphomas. It accounts for 0.01 % of all lymphomas but for 5% to 10% of all orbital tumors.[1,2] The clinical manifestations are non-specific. Radiological diagnoses are only seen in various case reports. This often results in delayed diagnosis and treatment.[3,4] We retrospectively analyzed the computed tomography (CT) and magnetic resonance imaging (MRI) scans of 14 patients.

2. Material and Methods

From the year 1995 to 2008, 14 patients (9 male and 5 female) were treated for primary orbital lymphomas at our hospital. The CT and MRI scans of these patients were reviewed. The median age at diagnosis was 36 years (range: 1.5 to 65). The average duration of symptoms was 3.8 years (range: 2 months to 16 years). The clinical presentations included eyelid swelling and palpable mass (14/14), painless proptosis (11/14), congestion of the bulbar conjunctiva (8/14), tenderness (2/14), dysopia (5/14), limitation of ocular motility (8/14) and upper eye lid prolapse (3/14) . Lymphadenopathy was not detected in other parts of the body following clinical examination.

Radiological examination included plain CT scans in all patients and contrast enhanced CT scans in all but one patient. MRI scans with and without contrast were performed in 10 patients. In the CT scanning process, the orbitomeatal (OM) line was used as the baseline, and each scan layer was 3mm thick. The enhancements of CT imaging were achieved with administration of 50-80 ml

***Correspondence:**
Professor Zhang Ming, Department of Radiology, First Affiliated Hospital of Xi'an Jiaotong University (School of Medicine) NO. 277. Yanta West Road Xi'an 710061, P. R. China Tel: +86-13519133887Fax: +86-2985323428. Email: profzmmri@gmail.com

Omnipaque (Nycomed, USA) at the rate 2.5 ml/sec. MRI examination included routinely transverse and sagittal T1- weighted and T2-weighted scanning. The above scans were repeated after the administration of intravenous (IV) contrast (12-15 ml of Magnevist; Schering, Germany).

3. Results

The final diagnosis of each patient was confirmed pathologically as B-cell type, non-Hodgkin's lymphoma. In 4 of the 14 patients there was involvement of bilateral orbits. Tumors were located in the upper quadrant of the orbit (12 cases) and in the lower quadrant (2 cases). The lesions of 5 cases were located in the upper lateral quadrant of the orbit with focal mass. These included 3 cases with clear demarcation, without adjacent tissue involvement and 1 case having indistinct edge and a local bulbar wall that was surrounded by tumor. Enlargement of the lacrimal gland was found in 12 patients. Nine patients showed diffuse, adjacent soft tissue involvement without clear margins. Four of these 9 patients had bilateral orbital tumors (Figure 1 and 2).

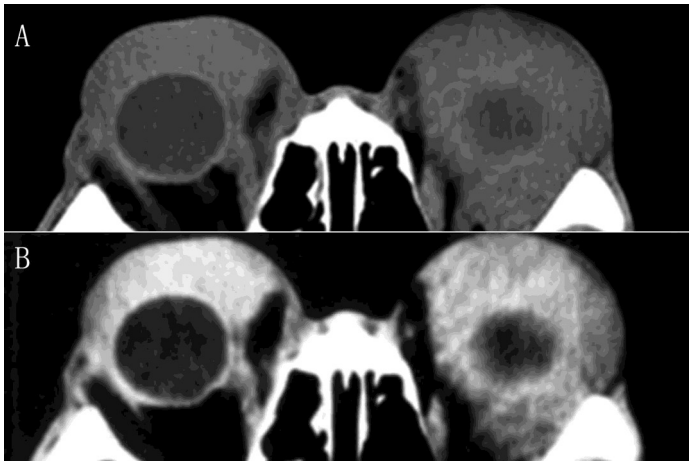


Figure-1(A-B): The CT films of bilateral diffuse lymphomas. The diffuse, slightly hypodense, soft-tissue lesions are shown bilaterally in the upper quadrant of the orbit (A). The lesions surround the eyeball walls "ring sign". The lesions display homogeneously marked enhancement on computed tomography examination with intravenous contrast (B).

The tumor that infiltrated along the extraocular muscles into the orbital tissues, but without involvement of eye-ball structure, was displayed as a "casting sign" (Figure 3). The infiltration of tumor around the eyeball created a "ring sign" in CT and MRI scans (Figures 1and 2).

A CT scan identified the homogeneous mass as isodense, with sharp margins. There was no calcification, hemor-rhage or bony-structure involvement in any of these patients. MRI multi-dimensional scans clearly showed the extent and severity of tumor extension inside the orbit. MRI can display more accurately the location, shape and internal structure of orbital lymphoma. They appeared as relatively low to medium intensity on T1-weighted images (Figure 2).

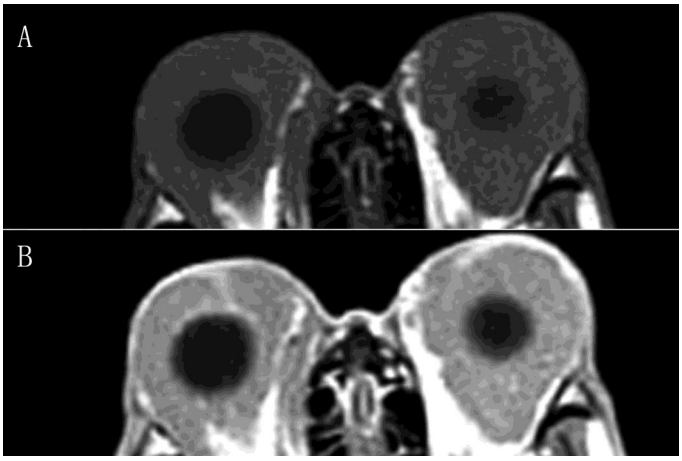


Figure-2(A-B): Transverse T1-weighed MRI (A) shows homogenously intermediate signal intensity and marked enhancement with intravenous contrast (B). The lesions have surrounded the eyeball "ring sign".

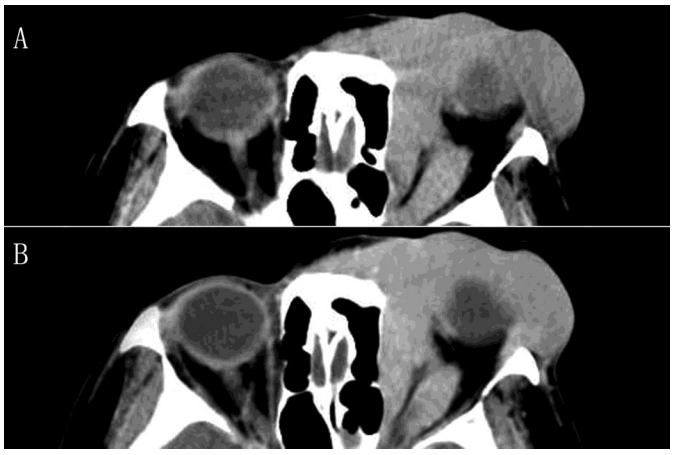


Figure-3(A-B): A diffuse lymphoma of the left orbit on computed tomography films. A diffuse, isodense soft-tissue mass is shown in the upper quadrant of the left orbit (A). The lesion progresses along the orbital wall "casting sign". The left eyeball proptosis and superior rectus muscle thickening can be observed simultaneously. The mass displays homogeneously marked enhancement on computed tomography examination with IV contrast (B).

Table-1: CT and MRI features of primary orbital lymphoma

Imaging features	No of cases	%
Presentation		
Bilateral	4	29
Unilateral	10	71
Growth Patterns		
Focal	5	38
Diffuse	9	62
Imaging Signs		
Casting sign	5	38
Ring sign	3	21
Hemorrhage	0	0
Calcification	0	0
Bone destruction	0	0
Homogeneous enhancement	14	100

Mass was viewed as slightly hypointense or isointense on T2-weighted images in 4 cases and relatively hyperintense

in 1 patient. The tumors displayed homogeneously intermediate to marked enhancement with the administration of contrast on both CT and MRI scans (Figures 1, 2 and 3). The findings of total cases are shown in table 1.

4. Discussion

Orbital lymphoma refers to a lymphoma occurring in the conjunctiva, lacrimal gland, eyelid and ocular musculature. The extraconal area has previously been shown to be the most frequent location of lymphomas in the orbital area.[5] Orbital lymphomas account for 5%-10 % of all orbital tumors. It is, however, the most common orbital malignancy.[1,2] They may arise spontaneously in the periorbital and/or orbital region (primary lymphoma) or arise in this region associated with systemic spread (secondary lymphoma). Several studies indicate that the most common lymphoma arising from tissues surrounding the eye is the low-grade B-cell lymphoma: so-called extranodal marginal zone lymphoma (MALT lymphoma).[6-8] Sjö LD et al found that more than 50% of orbital and ocular adnexal lymphomas were of the MALT lymphoma subtype, whereas diffuse large B-cell lymphoma (DLBCL) predominated intraocularly.[7] Orbital lymphomas can occur at any age, but majority of orbital lymphomas occur around 60 years of age or older, with female predominance.[9-11] Clinical manifestations of the orbital lymphomas include swelling and prolapses of the eyelid, painless but palpable masses, and exophthalmia and oculomotor defects.[12] These clinical findings are neither specific nor early signs of the development of primary orbital lymphomas. Early diagnosis can only be made with a needle biopsy.[3]

Previous reports found that most of these tumors are in the unilateral eye and occur without the infiltration of adjacent tissue.[13] Our findings showed that most of these tumors involved the eye on one side and 9 out of 14 cases displayed evidence of infiltration. This may be related to the long disease course found in most of our cases. The advances that have been made in CT and MRI techniques have made them better tool for the morphological diagnosis of primary orbital lymphomas.[5,11,14,15] In accordance with other reports, we found that the tumors were located mainly in the periorbital preseptal area. Localized masses with clear demarcations have previously been described as being the features of lymphomas.[5] The involved lacrimal glands become enlarged and displace the eyeball inwards and forwards. The eyeball wall is surrounded by diffuse, infiltrated tumors that do not involve the structures inside the eyeball. These can be described as the 'casting sign' and the 'ring sign' that are seen in CT and MRI scans. The optic nerve and bony structures are rarely involved. In CT scanning, the edges of tumors are irregular and the extraocular muscles are dislocated by the infiltration of tumor. Calcification, hemorrhage and loss of rapid flow through vessels were not observed in our patients. The soft-tissue mass is homogeneous and isodense when seen on CT scans. It shows a low to intermediate signal intensity on T1-weighted MRI and slight hypointensity or slight hyperintensity on T2-weighted scans. The tumors display homogeneously intermediate to marked enhancement with the administration of contrast agent. These imaging features were evident in most of our cases.

Previous studies have reported that orbital lymphomas have several characteristics, although they are not specific, and can be seen in a variety of orbital lesions. Dynamic contrast-enhanced MRI scans may show more specific characteristics, which may distinguish them from other diseases.[16,17] The contrast index (CI) curves of orbital lymphomas display relatively rapid increases, reach a maximum CI at 45~120 sec, and decrease rapidly in most cases. The majority of maximum CIs are less than 2.0. Therefore, CI curves of orbital lymphomas show comparative diagnostic features and the pattern of the lesions can be differentiated from other lesions. Although dynamic contrast-enhanced scanning was not performed in our patients, the lesions displayed remarkable enhancement at the early stage.

The differential diagnosis includes orbital inflammatory pseudotumors, diffuse lymphangiomas, lacrimal adenomas, metastases and other lymphoproliferative diseases. Orbital inflammatory pseudotumors are usually retrobulbar lesions, while orbital lymphomas locate in the anterior superior orbit.[14] On contrasted CT or MRI, diffuse lymphangiomas show no/or heterogeneous enhancement. Lacrimal adenomas typically reveal hyperintense T2 signal that is higher than that of orbital lymphomas, however, show lower contrast degree after either contrast CT or MR scan. Orbital metastases are often associated with bone destruction fossa orbitalis, and show heterogeneous density or intensity. It is often impossible to distinguish orbital lymphomas from other orbital lymphoproliferative diseases, i.e. reactive lymphoid hyperplasia or pseudolymphoma, just based on image findings. In these cases, parameters including imaging features (e.g., characteristics of margin, associated bony changes, enhancement patterns), pathophysiologic basis, age of

presentation, chronicity of the disease and response to the drug/hormone therapy may help to establish the diagnosis.[18]

5. Conclusion

In conclusion, examination of CT scan and MRI, particularly MRI demonstrates the location, configuration, internal structure and characteristic manifestations of orbital lymphomas. Differential diagnoses can therefore be made radiologically.

6. References

1. 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;49:49-55. doi:10.1007/s10384-004-0147-y PMid:15692775
2. Lutz SC, Anderson SF, Wu CY, Townsend JC. Non-Hodgkin's orbital lymphoma. Optom Vis Sci 2001;78:639-45. doi:10.1097/00006324-200109000-00008 PMid:11587197
3. Zhang C, Wei R, Cai J. Ocular adnexal mucosa-associated lymphoid tissue lymphoma: a pathological and imaging study. Yan Ke Xue Bao 2004;20:80-3. [Article in Chinese] PMid:15301101
4. Alford MA, Nerad JA, Conlan RM, Comito M, Giller RH. Precursor B-cell lymphoblastic lymphoma presenting as an orbital mass. Orbit 1999;18:17-24. doi:10.1076/orbi.18.1.17.2723 PMid:12048694
5. Lemke AJ, Hosten N, Neumann K, Grote A, Felix R. Space-occupying lesions of the lacrimal gland in CT and MRI exemplified by 4 cases. Aktuelle Radiol 1995;5:363-6. PMid:8580134
6. Suh CO, Shim SJ, Lee SW, Yang WI, Lee SY, Hahn JS. Orbital marginal zone B-cell lymphoma of MALT: radiotherapy results and clinical behavior. Int J Radiat Oncol Biol Phys. 2006;65:228-33. doi:10.1016/j.ijrobp.2005.11.035 PMid:16503386
7. Sjö LD. Ophthalmic lymphoma: epidemiology and pathogenesis. Acta Ophthalmol. 2009;87 Thesis 1:1-20
8. Yi WS, Xu XL, Xiang Q, Jiang HB. Characteristics of ocular adnexal non-Hodgkin lymphoma. Zhong Nan Da Xue Xue Bao Yi Xue Ban. 2008;33:826-30.[Article in Chinese] PMid:18812662
9. Ahmed S, Shahid RK, Sison CP, Fuchs A, Mehrotra B. Orbital lymphomas: a clinicopathologic study of a rare disease. Am J Med Sci 2006,331:79-83 doi:10.1097/00000441-200602000-00013 PMid:16479179
10. Charlotte F, Doghmi K, Cassoux N, Ye H, Du MQ, Kujas M, et al. Ocular adnexal marginal zone B-Cell lymphoma: A clinical and pathologic study of 23 cases. Virchows Arch. 2006,448:506-16 doi:10.1007/s00428-005-0122-0 PMid:16323006
11. Ferry JA, Fung CY, Zukerberg L, Lucarelli MJ, Hasserjian RP, Preffer FI et al. Lymphoma of the ocular adnexa: A study of 353 cases. Am J Surg Pathol. 2007,31:170-84 doi:10.1097/01.pas.0000213350.49767.46 PMid:17255761
12. Sullivan TJ, Whitehead K, Williamson R, Grimes D, Schlect D, Brown I, et al. Lymphoproliferative disease of the ocular adnexa: a clinical and pathologic study with statistical analysis of 69 patients. Opthal Plast Reconstr Surg 2005,21:177-88 doi:10.1097/01.IOP.0000159173.42243.AD
13. Akansel G, Hendrix L, Erickson BA, Demirci A, Papke A, Arslan A, et al. MRI patterns in orbital malignant lymphoma and atypical lymphocytic infiltrates. Eur J Radiol 2005;53:175-81. doi:10.1016/j.ejrad.2004.04.005 PMid:15664279
14. Hosten N, Schörner W, Zwicker C, Lietz A, Serke S, Huhn D. Lymphocytic infiltrations of the orbit in MRI and CT. Lymphoma, pseudolymphoma and inflammatory pseudotumor. Rofo1991;155:445-51.(In German) PMid:1954365
15. Lemke AJ, Kazi I, Landeck LM, Zaspel U, Hosten N, Felix R. Differential diagnosis of intraconal orbital masses using high-resolution MRI with surface coils in78 patients. Rofo 2004; 176: 1436-46. (In German) PMid:15383975
16. Cytryn AS, Putterman AM, Schneck GL, Beckman E, Valvassori GE. Predictability of magnetic resonance imaging in differentiation of orbital lymphoma from orbital inflammatory syndrome. Ophthal Plast Reconstr Surg. 1997;13:129-34. doi:10.1097/00002341-199706000-00007 PMid:9185195
17. Asaumi J, Yanagi Y, Hisatomi M, et al. The value of dynamic contrastenhanced MRI in diagnosis of malignant lymphoma of the head and neck. Eur J Radiol 2003;48:183-7. doi:10.1016/S0720-048X(02)00347-9
18. Colin S. Poon, Michael Abrahams, James Abrahams. Imaging of the Head and Neck: Orbit. In: John R. Haaga. CT and MR imaging of the whole body, 5th edition. Philadelphia, MOSBY/ELSEVIER, 2009;478-491.