·Case Report·

Mesenchymal chondrosarcoma of the orbit: report of a case and review of the literature

Hong-Bin Lü[1], Yu-Li Yang[2], Qing-Li Luo[2], Wei-Min He[2]

[1]Department of Ophthalmology, the Affiliated Hospital of Luzhou Medical College, Luzhou 646000, Sichuan Province, China
[2]Department of Ophthalmology, West China Hospital of Sichuan University, Chengdu 610041, Sichuan Province, China
Correspondence to: Qing-Li Luo. Department of Ophthalmology, West China Hospital of Sichuan University, Chengdu 610041, Sichuan Province, China. luo-qingli@263.net
Received:2009-01-07 Accepted:2009-05-08

Abstract

• AIM: To report a rare case of mesenchymal chondrosarcoma in the orbit and to explore its clinic manifestations, pathologic characters, management and prognosis.

• METHODS: We report a case of mesenchymal chondrosarcoma of the orbit. The clinical materials, including ophthalmological examination, computed tomography scan of the orbit, histopathology and immunohistochemistry of the biopsy specimen were reported, and its pertinent literatures were reviewed.

• RESULTS: A 36-year-old female was seen with proptosis and decreased vision. Histopathology demonstrated an admixture of undifferentiated mesenchymal cells and islands of mature hyaline cartilage. Immunohistochemical studies revealed positivity for vimentin and S-100, which was consistent with the diagnosis of mesenchymal chondrosarcoma.

• CONCLUSION: Mesenchymal chondrosarcoma in the orbit is extremely rare malignant tumor. Multi-modality treatments (surgery, chemotherapy and radiotherapy) may lead to long-term survival.

• KEYWORDS: mesenchymal chondrosarcoma; histopathology; immunohistochemistry

Lü HB,Yang YL,Luo QL,He WM.Mesenchymal chondrosarcoma of the orbit:report of a case and review of the literature. *Int J Ophthalmol* 2009;2(2):185-188

INTRODUCTION

Mesenchymal chondrosarcoma is a rare malignant tumor derived from primitive mesenchymal tissue with potential chondroblast. It can occur anywhere in the body, both in skeletal and extra-skeletal sites, including the limbs, orbit, vertebra, pelvis, meninges or brain. However, it tends to occur 2 to 3 times more commonly in bones than in extra-skeletal sites. It has a variable clinical course with frequent recurrence and occasional distant osseous and visceral metastatic spread [1]. It was first described in the bone in 1959 by Lichtenstein and Bernstein [2]. Mesenchymal chondrosarcoma in the orbit is extremely rare. Till now there have been only 18 cases overseas [1] and eight cases in China [3-8] reported on mesenchymal chondrosarcoma of the orbit. In these studies, there were no detailed case reports. In the present article, we report a case of orbital mesenchymal chondrosarcoma in detail, and summarize all the cases in Chinese literature, focusing on its clinical course, pathologic characters, management and prognosis in the literature.

CASE REPORT

A 36-year-old woman, without past medical history, complained of proptosis and decreased vision in the right eye from August 2001, and admitted to our hospital in May 2003. Computed tomography (CT) scan of the orbit revealed a firm lesion in right muscle cone, and tumor resection was done in June 2003. The postoperative pathological diagnosis was hemangiopericytoma of the right orbit. After operation, the patient was not given further chemotherapy and/or radiotherapy. She was re-admitted to our hospital with the same symptom in June 2005. On examination, uncorrected vision of the right eye was 1/50. The right eyeball protruded forward with mild limitation of mobility of the eyeball. On palpation, there was an unclear margin, middle-firmed mass, absence of tenderness and fluctuation (Figure 1). Fundus examination showed papilledema that projected 1D. The regional lymph nodes were not enlarged. Visual acuity (VA) of the left eye was normal. The laboratory examination was normal. CT scan of the orbit revealed a right intraorbital 20mm oval-shape, high density, partially calcified and well-defined, local recurrence lesion, without bone destruction (Figure 2). The preoperative diagnosis was hemangio-pericytoma of the right orbit. The patient was operated under general anesthesia through tracheal cannula and via lateral orbit rim approach on June 7, 2005. We discovered a

Table 1 Characteristics of patients with mesenchymal chondrosarcoma of the orbit

Case No.	Age (years)/sex	Location	Duration	Symptoms	Local Treatment	Recurrence	Metastasis	Follow-up		Author
								Duration	Outcome	
1	32/M	Left	x	x	x	x	x	x	x	Liu [3]
2	28/M	Right	1a	Proptosis, decreased vision	R, E, RT	+	-	4.5a	NED	Yi *et al* [4]
3	30/M	Left	x	Proptosis, decreased vision	R, E, CT	+	-	1a	NED	Yi *et al* [4]
4	20/M	Right	9mo	Proptosis,decreased vision,lacrimation	R	-	+	x	x	Luo *et al* [5]
5	22/F	Right	1mo	Proptosis, decreased Vision	R	+	+	4.5mo	x	Wang *et al* [6]
6	33/F	Right	3a	Proptosis,decreased vision,insight loss	R	-	-	x	x	Li *et al* [7]
7	21/F	Right	3a	Proptosis, insight loss	Multiple R	+	-	11a	NED	Peng *et al* [8]
8	22/M	Left	6mo	Proptosis, insight loss	R, E, RT	-	+	5a	NED	Peng *et al* [8]
9	36/F	Right	3a	Proptosis, decreased vision	R	+	-	x	NED	the present study

CT=chemotherapy; E=exenteration; F=female; M=male; NED=no evidence of disease; R=resection(s); RT=radiotherapy; x=unknown

relatively well encapsulated, and partially calcified lesion in right muscle cone, with mild adhesion to circumference tissue. The lesion deviated from the optic nerve without invading and the bony orbit appeared intact. We performed a macroscopically complete resection. Macroscopically, the size of the tumor was 30mm ×28mm ×20mm, and the cut surface was gray-white in color, rubbery in texture, and smooth in contour, with focally calcified areas. Histopathology showed a malignant tumor with an admixture of undifferentiated mesenchymal oval-shape and fusiform cells and islands of mature hyaline cartilage with some vascular channels (Figure 3). The mesenchymal cell nucleus chromatin was scarce, and had obvious nucleolus, clear atypia and mitotic figures. Islands of mature cartilage with mild atypia of cartilage cells were seen (Figure 4). Immunohistochemistry analysis was negative for desmin but positive for vimentin and S-100 (Figure 5). The pathological diagnosis was mesenchymal chondrosarcoma of the right orbit. The patient was discharged on 14 June 2005. The proptosis had regressed, and the right eye vision was slightly improved(1/25). Postoperative radiation therapy was performed.

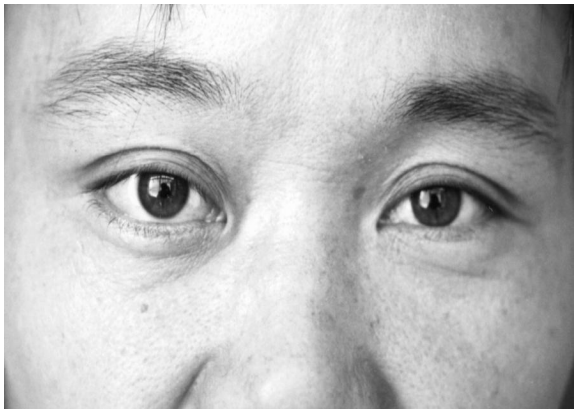

Figure 1 The right eyeball slightly protruded, and fissura palpebra widen

DISCUSSION

Mesenchymal chondrosarcoma in the orbit is extremely rare. On the basis of an Internet search of the literature through Chinese Biomedicine Disc (CBMDISC), Chinese Medical Current Contents (CMCC), Chinese Periodical Full-text Data Base (CJFD) and VIP Information (VIP), etc. from 1978 to 2005, only eight cases have been reported in China (Table 1) [3-8]. Outside China, Tuncer *et al* [1] searched for only 17 cases except his own case. He reported a case of a 5-day-old girl with a lesion in the right lower tarsal conjunctiva present from birth, whose final diagnosis of orbital mesenchymal chondrosarcoma was made by immunocytochemical analysis at the third histologic examination when she was 5 months old. The first pathologic diagnosis was nodular fasciitis at 5 days old and the second pathologic diagnosis was not definitive but suspicious of malignancy at 80 days old.

According to the foreign literature [1,9-13], the mean age at diagnosis of orbital mesenchymal chondrosarcoma is approximately 23 years (ranged from 5 months to 84 years), with a female predominance (72%). The youngest patient (a 5-month-old girl) with orbital mesenchymal chondrosarcoma was reported by Tuncer *et al* [1]. While in the Chinese literature [3-8], the mean age is 27.75 years (ranged from 20 to 36 years), with the female predominance of 37.5%. The reason for our lower occurrence rate in female may be that the cases are less (Table 1).

Orbital mesenchymal chondrosarcoma is a malignant tumor with proptosis and decreased VA as the primary symptoms, accompanied by various degrees of limitation of eye ball mobility and optic nerve compression, no evident delimitation with surrounding soft tissue. Some patients complained of

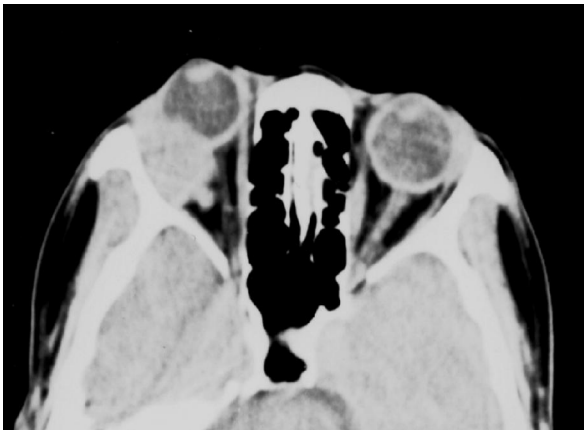

Figure 2 CT scan showed an oval-shape, high density, partially calcified and well-defined local recurrence lesion without bone destruction

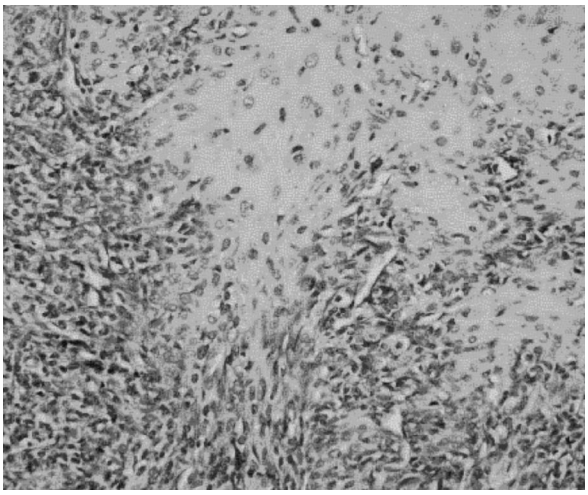

Figure 3 An admixture of undifferentiated mesenchymal oval-shape and fusiform cells and islands of mature hyaline cartilage with some vascular channels The mesenchymal cell nucleus chromatin was scarce, and had obvious nucleolus, clear atypia and mitotic figures (HE×248.16)

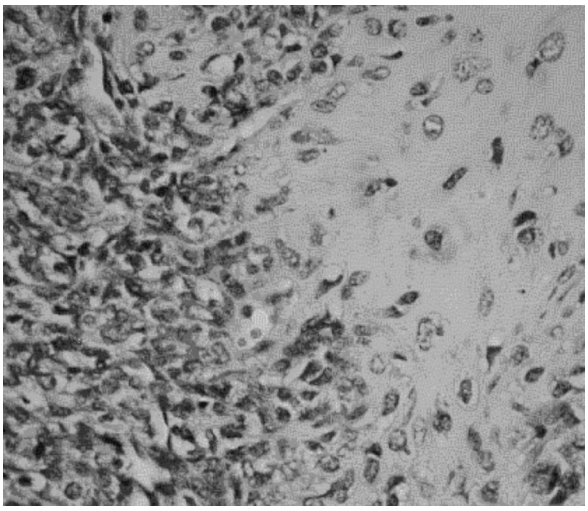

Figure 4 Islands of mature cartilage with mild atypia of cartilage cells were seen (HE×454.08)

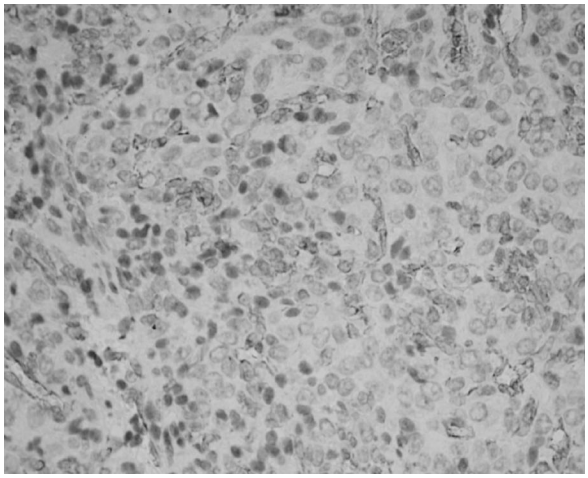

Figure 5 Immunohistochemistry showed vimentin (++) (× 503.36)

pain in eyes and headache. With progression of disease, it can invade the orbit soft tissue and local bone. Only one case had extended to the intracranial region reported by Khouja *et al* [13]. Distant metastasis is rare.

Orbital mesenchymal chondrosarcoma is most common in the retrobulbar soft tissues and characteristically appears on CT as a heavily calcified well-defined mass, usually located in the muscle cone space. The calcifications may be so heavy as to be seen on plain film radiography. Not all lesions, however, demonstrate calcification. In the settings, it presents as a well-marginated muscle cone mass with attenuation similar to that of the extraocular muscles.On dynamic contrast enhanced CT studies, mesenchymal chondrosarcoma demonstrates less rapid enhancement than nearby vascular structures [9].

The differential diagnosis of mesenchymal chondrosarcoma includes hemangiopericytoma, rhabdomyosarcoma, myxochodrosacoma, osteochondroma as primary orbital tumors and lymphoma, neuroblastoma, extraocular retinoblastoma, chloroma, synovial sarcoma and chondrosarcoma as tumors affecting the orbit secondarily (direct invasion or metastatic). Most of these tumors are round and oval shape histopathologically. Immunohistochemistry helps to differentiate among these tumors, such as desmin positivity in rhabdomyosarcoma, leucocyte common antigen positivity in lymphoma, neuron-specific enolase positivity in neuroblastoma, and vimentin and S-100 positivity in Mesenchymal tumors [1,9,12,14]. Moreover, although the histologic appearance of the tumor is distinctive, its rarity, uncommon locations in the orbit, and high vascularity may lead to the erroneous diagnosis of a vascular tumor such as hemangiopericytoma[1]. In fact, our case was misdiagnosed as hemangiopericytoma

at the first pathological diagnosis after operation and the diagnosis of mesenchymal chondrosarcoma was made by histological examination and immunohistochemistry vimentin and S-100 positivity after the second operation.

Most mesenchymal chondrosarcomas are resistant to chemotherapy and radiotherapy. The surgical resection is the mainstay of therapy. Complete removal of the tumors either by orbital resection or by exenteration may be adequate in some cases. However, if the tumor cannot be resected or seems histologically aggressive, chemotherapy and radiation therapy should be considered [1]. If thorough resection is difficult by preoperative estimation, radiotherapy and/or chemotherapy may be given for thorough resection. Tuncer *et al*[1] reported a case of 5-month-old girl who received one course of vincristine, actinomycin-D, and cyclophosamide and then two courses of ifosfamide, epirubicin and cisplatin (IEC), and then radiation therapy. After three courses of chemotherapy and radiotherapy, exenteration was carried out because the margin between the tumor and the globe remained indistinct. Postoperatively, three courses of IEC chemotherapy were administered. The patient was cured at last. In a review, 10 cases of extra-skeletal mesenchymal chondrosarcoma have been reported by Jacobs *et al* [12]. In this study, the orbit was exenterated in four cases; and in other six cases, the tumor was resected. Only adjuvant radiation therapy was used in two cases, whereas both radiation and chemotherapy were used in the other two cases. Of these four patients, two patients died 5 years and 6 months after treatment respectively.

The prognosis for patients with orbital mesenchymal chondrosarcoma is acceptable. In the Chinese literature [3-8], the shortest follow-up is 1 year and the longest is 11 years with no evidence of disease [8]. In the 18 cases reviewed and reported by Tuncer *et al* [1], seven patients was not clearly followed up. In the other 11 patients, three patients died at 6 months, 2 years and 5 years after treatment because of widespread metastasis, local recurrence, and treatment-related toxicity respectively, and eight patients were known to have survived for 5 to 18.5 years after treatment. Our patient has survived 2 years after the first operation and now is followed up.

In conclusion, orbital mesenchymal chondrosarcoma is extremely rare, with only eight published cases in the Internet-based Chinese literature, including one case of orbital mesenchymal chondrosarcoma misdiagnosed as hemangiopericytoma at first. Immunohistochemistry vimentin and S-100 positivity help to differentiate it. The prognosis is related to the quality of surgical resection and the additional chemotherapy and/or radiotherapy in case of incomplete resection.

Acknowledgement: We gratefully acknowledge the help of Department of Pathology of West China Hospital, Sichuan University.

REFERENCES

1 Tuncer S, Kebudi R, Peksayar G, Demiryont M, Cizmeci O, Ayan I, Gorgun O, Darendeliler E. Congenital mesenchymal chondrosarcoma of the orbit. Case report and review of the literature. *Ophthalmology* 2004;111(5):1016-1022

2 Lichtenstein L, Berstein D. Unusual benign and malignant chondroid tumors of bone: a survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors including a few multicentric ones and chondromyxoid fibromas. *Cancer* 1959;12:1142-1157

3 Liu ZJ. The pathological results of 13 cases of mesenchymal chondrosarcoma. *Tianjin Medi Drug* 1980;13(2):175-176

4 Yi YZ, Luo YR. Two cases of mesenchymal chondrosarcoma in the orbit. *Chin Ophthalmol* 1983;19(3):172-174

5 Luo HY, Wang Y, He CY. A case of mesenchymal chondrosarcoma in the orbit. *Acta Academiae Medi Militaris Tertiae* 1998;20(6):491

6 Wang TG, Qin W, Xie HP. One case of mesenchymal chondrosarcoma in the orbit. *Acta Academiae Medi Militaris Tertiae* 2006;28(20):2083

7 Li JJ, Wang ZX. A case of mesenchymal chondrosarcoma in the orbit. *Clin J Radio* 1999;18(3):153

8 Peng YG, Wang DQ. The treatment of mesenchymal chondrosarcoma in the orbit. *Chin J Pract Ophthalmol* 2000;18(1):57-58

9 Koeller KK. Mesenchymal chondrosarcoma and simulating lesions of the orbit. *Radiol Clin North Am* 1999;37(1):203-207

10 Shinaver CN, Mafee MF, Choi KH. MRI of mesenchymal chondrosarcoma of the orbit: case report and review of the literature. *Neuroradiology* 1997;39(2):296-301

11 Kashyap S, Sen S, Betharia SM, Dada VK. Mesenchymal chondrosarcoma of the orbit: a clinicopathological study. *Orbit* 2001;20(1):63-67

12 Jacobs JL, Merriam JC, Chadburn A, Garvin J, Housepian E, Hilal SK. Mesenchymal chondrosarcoma of the orbit. Report of three new cases and review of the literature. *Cancer* 1994;73(2):399-405

13 Khouja N, Ben Amor S, Jemel H, Kchir N, Boussen H, Khaldi M. Mesenchymal extraskeletal chondrosarcoma of the orbit. Report of a case and review of the literature. *Surg Neurol* 1999;52(1):50-53

14 Cai L, Gao ZF, Huang XY. Clinicopathology analysis of mesenchymal chondrosarcoma in soft tissue. *Journal of Peking University (Health Sciences)* 2006;38(5):501-505