

Dedifferentiated orbital liposarcoma: a case report

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Abstract

• **AIM:** To report the abnormal type of the orbital liposarcoma-dedifferentiated subtype in a patient.

• **METHODS:** A case report.

• **RESULTS:** A 23 years old Chinese woman with a recurrence of right-sided proptosis was evaluated. Ocular examination revealed proptosis of the right eye with chemosis, hyperemia and limitation of eye movements. Magnetic resonance imaging scanning showed an irregular shaped tumor in the right orbit. The tumor resection was done with a clinical diagnosis of malignant tumor. Histopathological findings revealed the diagnosis of dedifferentiated liposarcoma.

• **CONCLUSION:** The rare occurrence of this tumour should be kept in mind while dealing with orbital tumours.

• **KEYWORDS:** orbit; liposarcoma; tumour

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INTRODUCTION

Primary liposarcomas of the orbit are rare entities^[1]. Only 40 cases have been reported so far^[2-6]. There are no specific clinical symptoms in orbital liposarcoma. The most common presentation is proptosis, with or without pain. Pathology is necessary to confirm the diagnosis, which can also be assisted by a MRI or a CT scan. The most common type is myxoid, but the other 3 subtypes also can occur. Literature search revealed only 1 case of dedifferentiate orbital liposarcoma. Here, we report the second case of dedifferentiated orbital liposarcoma and present the clinical and pathologic characteristics of this disease.

CASE REPORT

A 23-year-old Chinese woman had originally presented to another hospital with a 3-year history of right-sided proptosis. Lipoma was diagnosed at the time, and a tumor

resection was performed. After a recurrence, she was referred for further management.

On ocular examination, there was proptosis of the right eye with chemosis, hyperemia and limitation of eye movements. A palpable mass was found at the medial canthus. The mass was firm and ill-defined. Her visual acuity visual acuity was 20/40 in the right eye and 20/20 in the left eye. The intraocular pressure, and fundi examination were normal. She had no other contributory non-ocular findings, including chest radiograph.

Magnetic resonance imaging (MRI) of orbits showed an irregular shaped tumor in right orbit. T1-weighted showed a medium inhomogeneous high signal in the right orbit (Figure 1A). T2-weighted showed a mixed high signal mass in the nasal orbit involving medial rectus muscle. There were no obvious abnormalities in the surrounding orbital floor. (Figure 1B). The patient underwent an anterior orbitotomy for tumor resection with a clinical diagnosis of malignant tumor. Intraoperatively, the mass was covered by a thin layer of fibrous tissue. The histopathological examination of the whole tumor tissue was done.

Histopathological findings revealed the diagnosis of dedifferentiated liposarcoma. Histologically, the tumor was composed of adipocytes including areas of dedifferentiation. The adipocytes were of varying sizes, with deep staining nuclei. The dedifferentiated components were composed of spindle-shaped cells, resembling a fibrosarcoma. There were often mitotic figures (Figure 2A,B). Immunohistochemically, the tumor cells were positive for CD99, CD34, vimentin, S-100, Bcl-2, CD68, SMA, and mostly positive for ki-67.

Following surgery, the patient received a course of postoperative radiotherapy in other hospital.

She was followed up for 16 months. There was no evidence of recurrence. On the last follow up examination, there was an exotropia in the right eye. There was no proptosis of right eye. Her visual acuity was the same result as pre-operation, intraocular pressure was 16mmHg in the right eye and 18mmHg in the left eye. Both optic discs were normal. There was no evidence of the radiation retinopathy and radiation optic neuropathy. The MRI imaging of both orbits showed that there was no recurrence of tumour mass in the right orbit in the last follow up examination (Figure 3A,B).

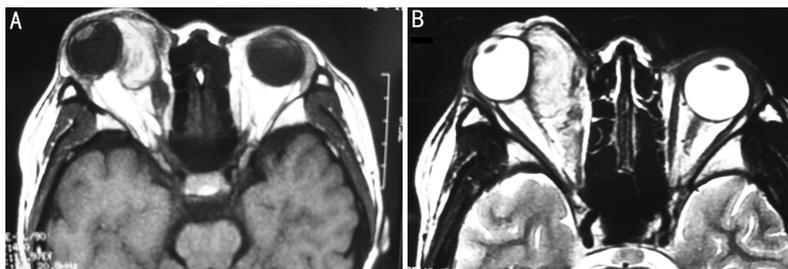


Figure 1 A: T1-weighted (MRI); B: T2-WI showed an inhomogeneous signal, irregular shaped soft-tissue mass in the right orbit involving the medial rectus muscle and the muscle cone. The mass is still normal-defined

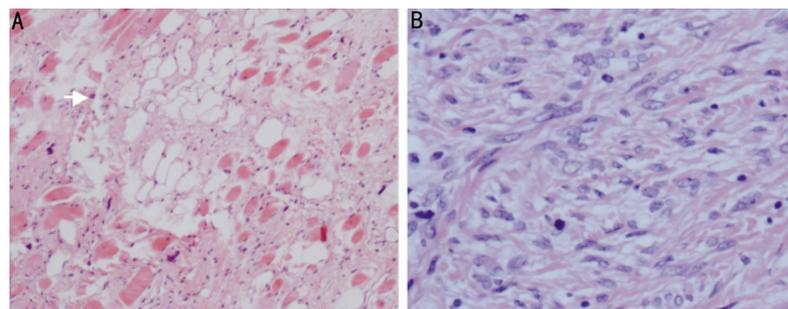


Figure 2 A: Histopathologically, it showed the well differentiated structure, the adipocytes were of varying sizes, with deep staining nuclei (white arrow) (hematoxylin-eosin; original magnification x100); B: The non-liposarcoma components were composed of spindle-shaped cells, presenting structures of the low-grade fibrosarcoma. (hematoxylin-eosin; original magnification x400)

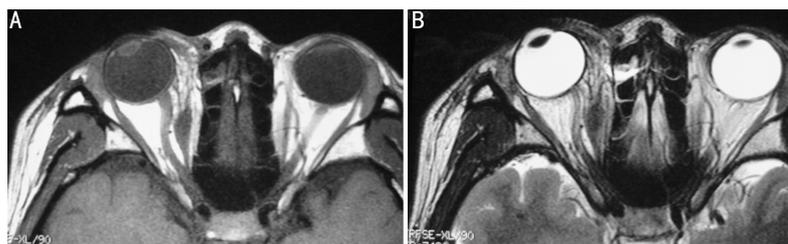


Figure 3 Post operative T1-weighted A; MRI; B: T2-WI showed: a strip-shaped mixed signal shadow can be seen in the right orbit, ill-defined with the medial rectus muscle, accompany with obliteration of the fat. There were no other abnormalities

DISCUSSION

The first description of orbital liposarcoma by Strauss, appeared in 1911, quoted by Lanne *et al*^[3]. To our knowledge, only 40 cases had been reported as orbital liposarcoma within the English language medical literature^[2-6]. The major clinical features (including our case) were proptosis (35/38 cases; 92.1%), diplopia (42.1%), reduction in visual acuity (28.9%) and local hyperaemia (21.1%). Other unusual presentations include pain, limitation of eye movements, and systemic symptoms like weight gain and loss of sense of smell^[2].

Four different subtypes of liposarcoma could be observed in orbit. In the past literature^[2], myxoid (including round cell) was the most common subtype (22 cases, 55%) reported, other subtypes were well differentiated (12 cases, 30%) and pleomorphic (4 cases, 10%), the dedifferentiated subtype was extremely rare (only 1 case, 2.5%), and the histologic type of the remaining 1 case (2.5%) was uncertain^[4]. Here, we presented an additional case of dedifferentiated orbital liposarcoma. The dedifferentiated type frequently produces both poorly and well differentiated deposits. The tumours are composed of mature adipocytes and structures of an

atypical lipomatous tumor, such as an undifferentiated spindle cell sarcoma in our case.

The clinical diagnose of the orbital liposarcoma is difficult because it has no diagnostic symptoms. Orbital CT and MRI are imperative when a tumour is suspected, and will provide evidence of invasion into neighbouring structures. But pathology is the only way for definite diagnose. Complete surgical resection is recommended while a confirmed diagnosis is made^[6]. The role of both radiotherapy and chemotherapy in the management of primary orbital liposarcoma is still unclear.

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