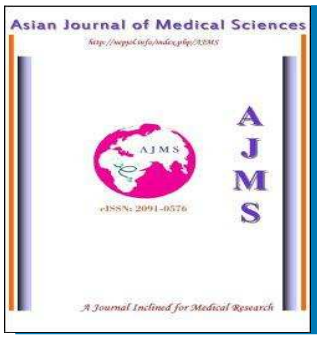

PRIMARY ORBITAL MYXOID LIPOSARCOMA: A CASE REPORT AND REVIEW OF LITERATURE

CASE REPORT AND REVIEW OF LITERATURE, Vol-4 No.4

Asian Journal of Medical Science, Volume-4(2013) http://nepjol.info/index.php/AJMS

[1]Chen Niu, [2]Netra Rana, [3]Zhi Gang Min, [4]Ming Zhang. Department of Radiology , The First Affiliated Hospital of Xi'an Jiaotong University

CORRESPONDENCE:

Prof. Ming Zhang, MD, PhD
Department of Radiology,
The First Affiliated Hospital of Xi'an Jiaotong University
NO. 277, Yanta West Road,
710061, Xi'an, Shaanxi, P.R. China
Tel: +86-13519133887
Fax: +86-2985323428
Email ID:
profzmmri@gmail.com

"Liposarcoma is a malignant tumor of adipose tissue and considered to be the most common soft tissue sarcoma in adults, orbital liposarcoma is extremely rare. Most cases of orbital liposarcoma are primary and rarely metastatic."

ABSTRACT

Liposarcomas are common malignant soft-tissue tumors, which come from primitive mesenchymal cells and differentiate into adipose tissue. These tumors are more commonly found in lower limbs and retroperitoneal region but also reported in pharynx, lung, liver, digestive tract, diaphragm, as well as in the spermatic cord. We reported a case of primary orbital myxoid liposarcoma in a 20-year-old female patient presented with a painless proptosis of the right eye. The mass was pathologically diagnosed as a myxoid liposarcoma. The tumor recurred in 9 months after surgical intervention. The second surgery was performed and followed by postoperative local radiotherapy. No recurrence has been reported after one year of follow-up. We highlighted the role of CT and MRI findings in the tumor diagnosis and the importance of local radiotherapy after surgery.

Keywords: Orbital myxoid liposarcoma; radiotherapy; computed tomography; magnetic resonance imaging

INTRODUCTION

Liposarcoma is a malignant tumor of adipose tissue and considered to be the most common soft tissue sarcoma in adults, though the orbital liposarcoma is extremely rare. It is believed to develop from mesenchymal cells rather than from a preexisting lipoma. Most cases of orbital liposarcoma are primary and rarely metastatic. Literature reports show the commonest site for liposarcomas are lower limbs and retroperitoneal region, but cases in pharynx, lung, liver, digestive tract, diaphragm, as well as spermatic cord also have been reported.[1-3] Head and neck is not a frequent site for this tumor; though, a few case reports have been reported. This rarity makes it more difficult to diagnose and to treat in this clinical setting. We report a case of primary orbital liposarcoma with clinical presentation, radiologic studies (CT and MRI), detailed histopathologic features and subsequent treatment method. A brief review of primary orbital liposarcoma is also presented.

CASE REPORT

A 20-year-old female presented with a three-month history of progressive painless proptosis of the right eye. Imaging performed at the local hospital revealed an irregular isodensity retrobulbar mass displacing the globe. It was initially diagnosed as an inflammatory pseudotumor and was treated with oral steroids and the intravenous hexadecadrol for a month but showed no response. Two months later, a repeated CT scan revealed an irregular isodensity mass with well-defined borders in the right retrobulbar region attached to the lateral wall (Fig. 1A). The fat-density was observed within the mass with a mean CT value of -10 Hounsfield units (HU). The adjacent optic nerve and surrounding tissues were slightly medially displaced. Clinically, the patient was in good physical conditions and had no visual defects. Both eyes movements were normal. No obvious abnormalities and tumor masses were found in physical and imaging examinations. MRI study also revealed a similar findings (Fig. 1B-D). On axial T1 weighted image (T1WI), the mass showed low signal intensity accompanied by small nodular spots of high signal intensity in small areas (Fig. 1B). The signal intensity was much higher on axial T2 weighted image (T2WI) (Fig. 1C). The nodular spots of low signal intensity were observed on the fat-suppressed T2WI (Fig. 1D). The mass showed heterogeneous enhancement on post gadolinium fat-suppressed T1WI (Fig. 1E). The massive mucilage mass with no capsule or clear border was identified during surgery. The optic nerve was found wrapped by the mass and the rectus lateralis muscle was smeared with mucliage. The immunohistopathological study (S-100+, P53 +++) confirmed myxoid liposarcoma (Fig. 1F and 2E). No further therapies were continued postoperatively. Nine months later, the patient again presented with proptosis of the right eye. The imaging and histopathology confirmed recurrence (Fig. 2). The same pathological diagnosis was reported. This time, the patient was managed with surgery and followed by 60Gy of radiotherapy, postoperatively. The patient did not show any sign of recurrence and is free of disease for 12 months follow-up after surgery (Fig. 2F).

DISCUSSION

According to WHO (2002), liposarcomas are classified as atypical liposarcoma, (i.e.,well-differentiated liposarcoma), myxoid liposarcoma, pleomorphic liposarcoma, round cell liposarcoma, as well as dedifferentiated liposarcoma. The well-differentiated liposarcomas include lipoma-like liposarcoma and sclerosing liposarcoma. Myxoid liposarcoma account for most of the reported orbital liposarcomas.[4,5] Drevelegas et al.,[6] found that pathologically myxoid liposarcoma were composed of lipoblasts in varying stages of differentiation, plexiform vascular network and myxoid matrix. According to the literature review, both genders are equally affected where female

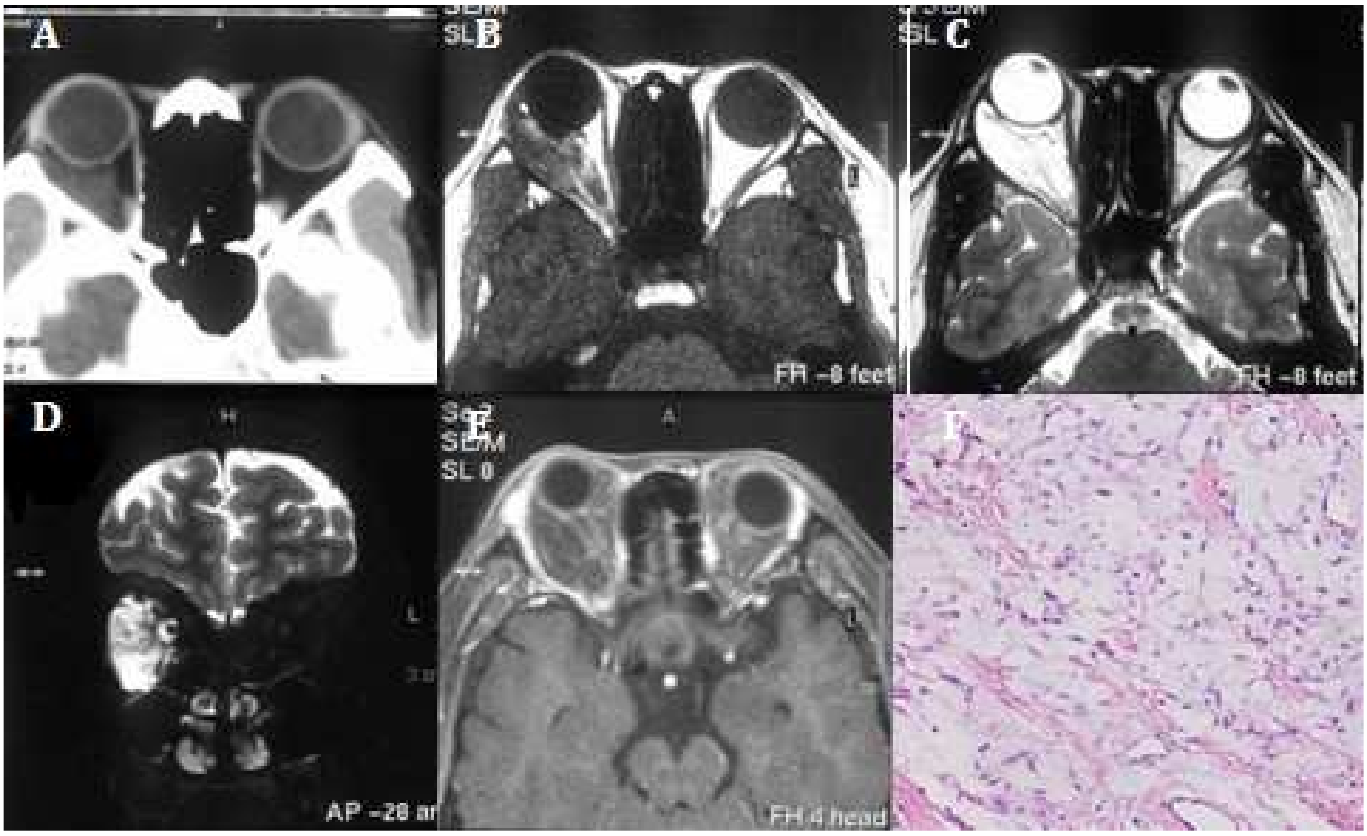

Figure 1: (A) CT scan shows an irregular well-defined isodensity mass in the right orbit with fat-density (mean CT value of -10 HU). (B) MRI scan shows a heterogeneous signal intensity mass on T1WI. (C) The mass shows much higher signal intensity on T2WI. (D) On fat-suppressed T2WI, nodular septas of low signal intensity are present. (E) The mass is heterogeneously enhanced on gadolinium-enhancement and fat-suppressed image. (F) Lower magnification (H and E, ×10) of the tumor tissue showing lipoblasts, vasculatures and myxoid background, the characteristic of the myxoid liposarcoma.

predominance is slightly higher, and the age ranges from 20 to 60 years old.[4,5,7] There are no characteristic clinical findings or diagnostic tests that are specific for liposarcoma. Patients may present with a progressive mass, proptosis, diplopia, visual impairment, and possible pain.[7] Symptoms are normally caused by the compression of orbital structures and may develop over several weeks to years before receiving the medical treatments. No tumors could be detected in other body parts/ locations in case of primary liposarcoma.[8] In our case, the patient presented a painless isolated proptosis of the right eye for a few months without any evidence of mass in other parts of the body, which was fitted with those described in literatures.[4,8] It is notable that orbital inflammatory pseudotumors also present the similar symptoms, they are very sensitive to hormonotherapy and may help to make a possible differential diagnosis.

Different forms of lipoblasts and myxoid matrix in myxoid liposarcoma have distinct characteristics in CT and MRI scans.[5] In our case, CT scan showed the isodensity mass with the low density in some small

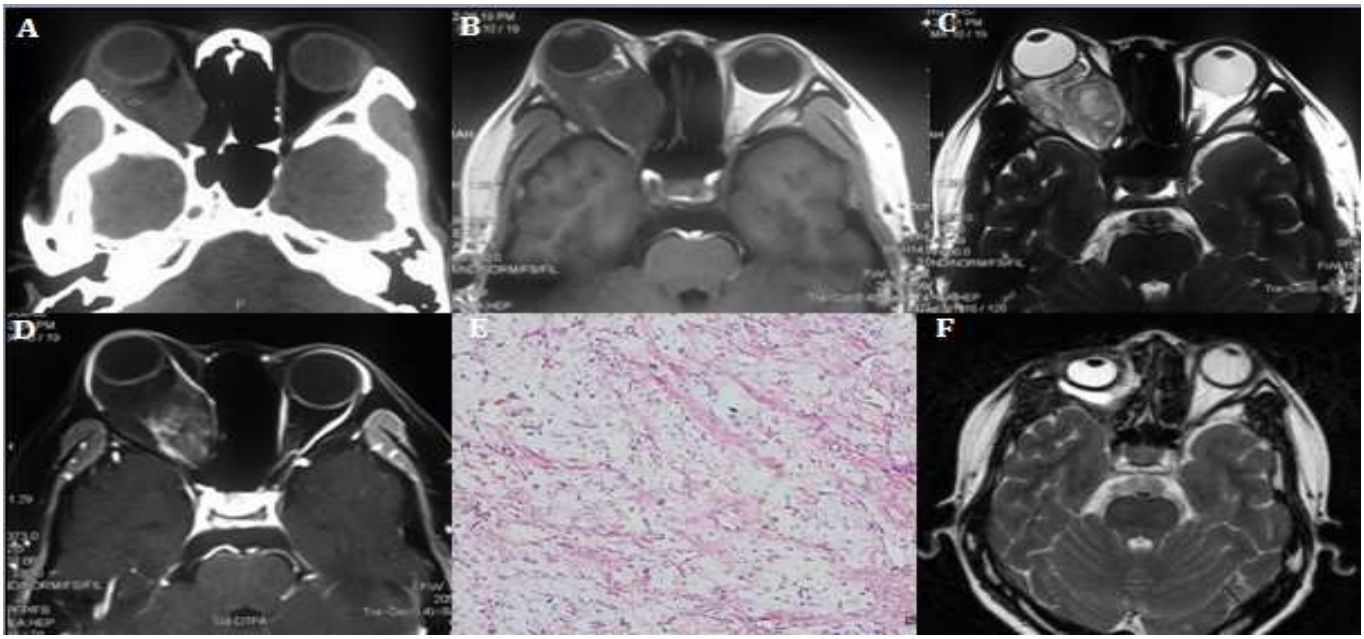

Figure 2: Same patient showing the evidence of recurrence of the tumor. (A) A isodensity mass with irregular border and small spots of low-density within the mass (mean CT value of -25 HU) is present in the right retrobulbar area. (B) On axial T1WI, the mass shows medium heterogeneous signal intensity. (C) On axial T2WI, the mass shows heterogeneous higher signal intensity. (D) On fat-suppressed and contrast enhanced image, the mass shows heterogeneously stronger enhancement than the previous one (Fig. 1E). (E) Medium- magnification (H and E, ×20) of the myxoid liposarcoma showing lipoblasts, vasculatures and myxoid background. (F) The post surgical image after treated with adjuvant radiation therapy. No evidence of recurrence is found after one year follow-up.

areas, which corresponded to the adipose tissue. The well-defined border was observed in the CT scans. But, surgery failed to find the capsule of the tumor. This may be related with the fact that the tumor compression created "nominal" border shown in the CT scans. MRI is very sensitive to soft-tissue tumors. T1WI, T2WI, fat-suppressed and gadolinium-enhanced scans can help to distinguish liposarcomas from other soft-tissue tumors by the detection of adipose components within the tumor, and are frequently reported. Ouni et al.,[9] analyzed twelve cases of myxoid liposarcoma and found that this type of tumors normally had well-defined borders. Lipoma may show similar clinical manifestations and imaging features making it difficult to distinguish from liposarcoma. However, the previous finding has suggested that the presen-ce of thick or nodular septa in the tumor is more suggestive of liposarcoma than lipoma.[10] In our case, we observed some apparent nodular septa of low signal intensity on MRI scans (Fig. 1D, and Fig. 2C). In addition, it is evident that CT and MRI scans can display with details of the liposarcoma. Though orbital liposarcomas are uncommon, we suggest that they should be considered in the differential diagnosis when some fat-density and nodular septa of low signal intensity are observed on CT and MRI scans, respectively.

Myxoid liposarcoma accounted for most of the orbital liposarcomas. They rarely recurred, and metastasis are very rare after surgical excision. The tumors were quite sensitive to radiotherapy.[5,9] Surgery is the therapeutic choice for orbital liposar-comas. It is recommended that subsequently loca-

radiotherapy is helpful to avoid the recurrence.[5]

In conclusion, primary myxoid liposarcoma rarely occurs in the orbit. Nevertheless, clinicians should consider it as a differential diagnosis when dealing with orbital masses. The treatment of choice for liposarcoma might be local excision of the tumor, accompanied by adjuvant radiation therapy and/or chemotherapy.

REFERENCES

1. Lee SY, Kim HJ, Park SY, Park YH, Chung SK. Myxoid liposarcoma involving the liver, subcutaneous tissue, and epidural space in a polycystic disease patient. Clin Nucl Med 2008; 33: 507-509.
2. Mridha AR, Sharma MC, Sarkar C, Suri V, Garg A, Suri A. Primary liposarcoma of the orbit: a reporta of two cases. Can J Ophthalmology 2007; 42: 481-483.
3. Liakakos TD, Troupis TG, Tzathas C, Spirou K, Nikolaou I, Ladas S, et al. Primary liposarcoma of esophagus: a case report. World J Gastroenterol 2006; 12: 1149-1152.
4. Cai YC, McMenamin ME, Rose G, Sandy CJ, Cree IA, Fletcher CD. Primary liposarcoma of the orbit: a clinicopathologic study of seven cases. Ann Diagn Pathol 2001; 5: 255-266.
5. Costas A, Castro P, Munoz JM. Primary orbital liposarcoma: case report and review of the literature. Int J Oral Maxillofac Surg 2001; 30: 170-172.
6. Drevelegas A, Pilavaki M, Chourmouzi D. Lipomatous tumors of soft tissue: MR appearance with histological correlation. Eur J Radiol 2004; 50: 257-267.
7. Garrity JA, Henderson JW, Cameron JD. Henderson's Tumors of Primitive Mesoderm, Striated muscle, Adipose Tissue and Smooth muscle. In: Garrity JA, Henderson JW, Cameron JD, editors. Henderson's orbital tumors. 4th ed. Philadelphia: Lippincott Williams and Wilkins; 2007. pp. 123–141.
8. Madge SN, Tumuluri K, Strianese D, Bonavolonta P, Wilcsek G, Dodd TJ, et al. Primary orbital liposarcoma. Ophthalmology 2010; 117: 606-614.
9. El Ouni F, Jemni H, Trabelsi A, Ben Maitig M, Arifa N, Ben Rhouma K, et al. Liposarcoma of the extremities: MR imaging features and their correlation with pathologic data. Orthop Traumatol Surg Res 2010; 96: 876-883.
10. Jaovisidha S, Suvikapakornkul Y, Woratanarat P, Subhadrabandhu T, Nartthanarung A, Siriwongpairat P. MR imaging of fat-containing tumours: the distinction between lipoma and liposarcoma. Singapore Med J 2010; 51: 418-423.