Journal of Clinical & Experimental Ophthalmology

Farah et al., J Clin Exp Ophthalmol 2015, 6:1
DOI: 10.4172/2155-9570.1000388

Case Report Open Access

An Unsual Case of Eyelid Leiomyosarcoma and Orbital Invasion with MRI Findings

Edgard Farah[1*], Pierre-Vincent Jacomet[1], Mathieu Zmuda[1], Marc Putterman[2] and Olivier Galatoire[1]

[1]*Oculoplastic department, Fondation Ophtalmologique Adolphe de Rothschild, Ophtalmology, France*

[2]*Consultant, France*

[*]**Corresponding author:** Edgard Farah, Fellow in Oculoplastic department, Fondation Ophtalmologique Adolphe de Rothschild, Ophtalmology, Paris -75019, France; Tel: +33619354503; E-mail: edyfarah@gmail.com

Received date: Dec 02, 2014, **Accepted date:** Jan 20, 2015, **Published date:** Jan 23, 2015

Abstract

A 73 years old woman presented with 10 months history of inferior eyelid nodule, the biopsy revealed at the time a grade 2 leiomyosarcoma without metastasis, the excision was considered inappropriate and radiotherapy was performed. The patient was addressed for resistant ocular pain management grade 3 and persistence of the lesion despite all the treatments. The mass is now measuring 8 cm in maximal diameter. The MRI showed lesions that affect mainly the median extra orbital part. An embolization of the main artery that vascularise the lesion was performed, proceeded by wide exenteration. The patient died 4 months later of local recurrence in the orbital cavity and metastases. Leiomyosarcoma as a highly malignant character showed a furious resistance to chemotherapy, radiotherapy and embolization so wide radical exenteration from the beginning might give better chance at long term survival.

Keywords: Leiomyosarcoma; Orbit; Radiotherapy; Exenteration

Case Report

A 73 years old woman presented with 10 months history of inferior eyelid nodule without ocular symptoms, the histology of the biopsy revealed at the time a grade 2 leiomyosarcoma without metastasis. A multidisciplinary approach made by her surgeon, oncologist and radiologist recommended a radiotherapy with adjuvant chemotherapy as an initial therapy. The radiotherapy was performed with a dose of 30 Gy in 10 fractions 3 Gy per session during 3 months. An adjuvant treatment with chemotherapy 6 cycles (75mg/m2/cycle) was performed in interval of 3 months. The patient was addressed 3 months later to our department for resistant ocular pain management grade 3 and persistence of the lesion despite all the treatments. The mass is now measuring 8 cm in maximal diameter (Figure 1), the MRI showed a lesion that affect mainly the median extra orbital and extraconal part without infiltration of the intraconal fat nor the optic nerve (Figure 3), in the cerebral arteriography we can visualize the vascularisation of the lesion by the external carotid (terminal branches of infernal Maxillary artery) and the ipsilateral ophthalmic artery [1,2].

An embolisation of the main artery that vascularise the lesion was performed, proceeded by wide exenteration without adjunctive radiotherapy due to rapid clinical deterioration of the patient.

The diagnosis of leimyosarcoma is confirmed by histopathalogic examination that revealed grade 2 by clinical grade (Figure 2) cellularity +++, spindle shaped cells that have cigar-shaped nuclei, mitotic index 15 (x10), Necrosis (0) and high grade cellular atypia, and grade 3 of malignancy by AJCC staging system that was confirmed by the high histological grade and the tumor size (≥ 5 cm) [3,4].

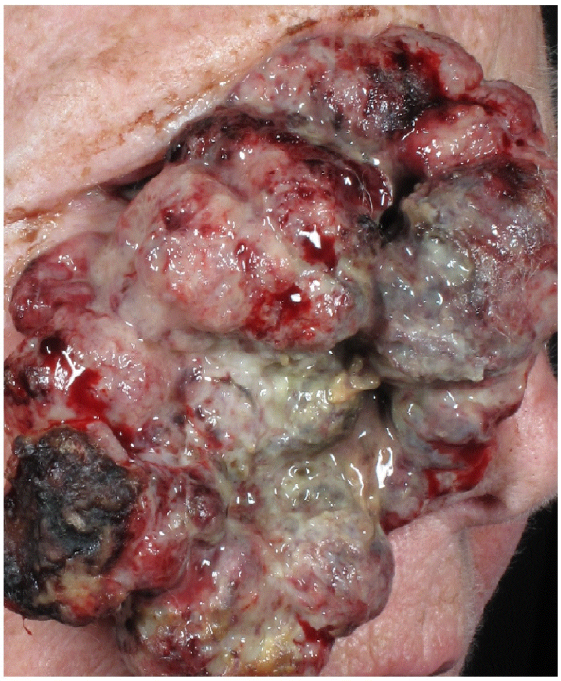

Figure 1: Multi-lobulated, well vascularised mass measuring 8 cm on maximal diameter.

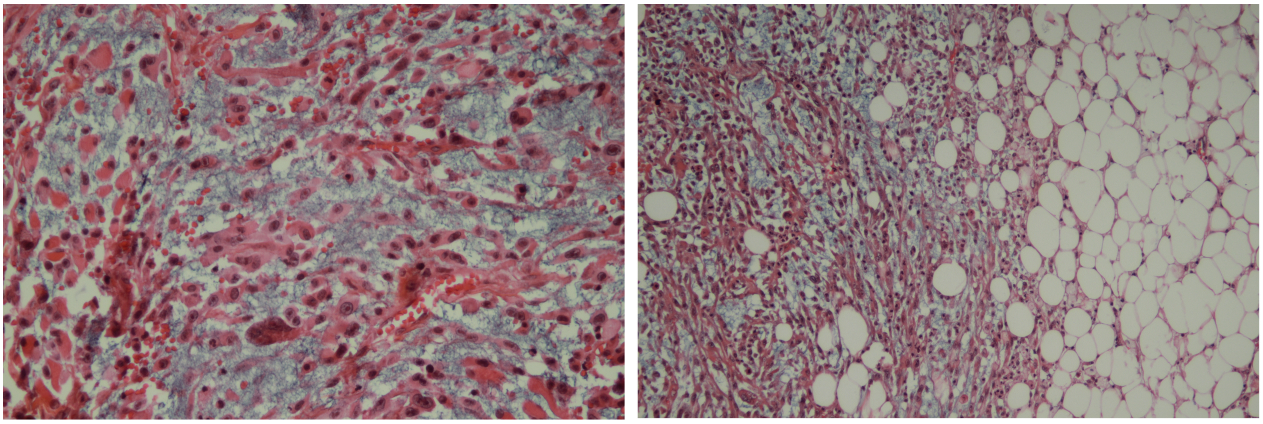

Figure 2: Histopathology images of High grade leiomyosarcoma fig a (x 10), fig b (x20), tumor showing spindle-shaped neoplastic infiltration.

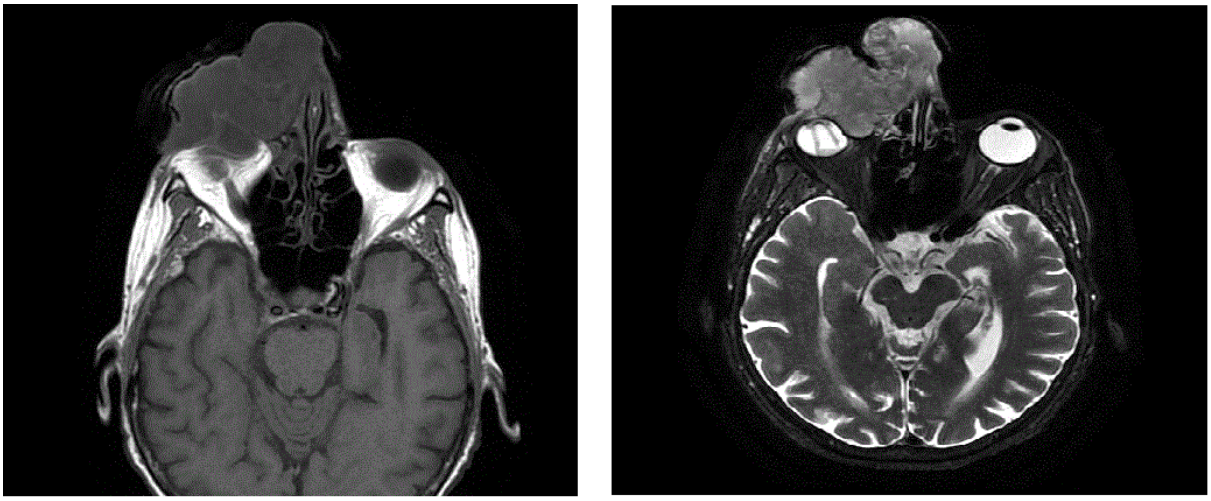

Figure 3: MRI shows a left orbital mass (left) transaxial T1 weighted MRI image demonstrates an 8 cm mass that is hypointense relative to cerebral cortex with peripheral contrast enhancement. (Right) transaxial T2-weighted MRI the mass is isointense relative to cerebral cortex.

Immunohistochemistry test showed that the tumor cells were positive for antismooth muscle actin, CD68 and negative for s-100 proteine, CD 30, desmine, caldesmone.

According to Myajima et al. the tumor size and AJCC stage in leiomyosarcoma are the most reliable prognostic parameters for survival rate [5]. The patient died 4 months later of local recurrence in the orbital cavity and metastases in the near ganglions, lungs and liver.

Discussion

Leiomyosarcoma is a rare neoplasm of smooth muscle, a primary myogenic tumor in the orbit is suspected to be vascular smooth muscle [3]. Folberg et al. described orbital leiomyosarcoma secondary to radiation for bilateral retinoblastoma [6]. Hou et al. suggested that MRI can provide more important information then CT and Ultrasound [1], on T1 weighted MRI the tumor was hypointense and on T2 it was Isonintense relatively to cerebral cortex with peripheral contrast enhancement and extraconal location of the mass (Figure 3).

In the literature the surgical wide local resection was the treatment of choice, Jakobiec et al. recommend exenteration with the removal of any involved bone required because of the high infiltrative nature of leiomyosarcoma [4] and it is possible that asymptomatic metastases may have occurred before exenteration so a meticulous search for any metastasis especially lung metastasis should be conducted before any treatment.

Our case illustrate, that leiomyosarcoma as a highly malignant character showed a furious resistance to chemotherapy, radiotherapy and embolization so wide radical exenteration with adjunctive radiotherapy from the beginning might give better chance at long term survival.

References

1. Hou LC, Murphy MA, Tung GA (2013) Primary Orbital Leiomyosarcoma : A Case Report With MRI Findings. American J of Ophtalmol 408-410.

2. Yeniad B, Tuncer S, Gonul P, Ozgur M, Ozenc M, et al. (2009) Primary Orbital Leiomyosarcoma American Society of Ophtalmic Plastic And Reconstructive Surgery 154-155.
3. I-Chan lin, Chen-Tu Wu (2005) Primary Orbital Leiomyosarcoma. Ophtalmic Plastic And Reconstructive Surgery 451-453.
4. Ted Jakobiec FA, Howard GM, Rosen M, Wolff M (1975) Leiomyoma and leiomyosarcoma of the orbit. Am J Ophtalmol 80: 1028-1042.
5. Miyajima K, Oda Y, Oshiro Y, Tamiya S, Kinukawa N, et al. (2001) Clinicopathological prognostic factors in soft tissue Leiomyosarcoma : a multivariate analysis Histopathology 40: 353-359.
6. Folberg R, Cleasby G, Flanagan JA, Spencer WH, Zimmerman LE (1983) Orbital Leiomyosarcoma After Radiation Therapy for Bilateral Retinoblastoma. Arch Ophthalmol 101: 1562-1565.