5th World Congress of IFHNOS and Annual Meeting of HNS

"Primary Adenocarcinoma of the Lacrimal Gland"

P Ruscito[1], L Urbani[1], L Sabino[1], M Bernardini[1], F Liberati[2]

[1]Otolaryngology, Head & Neck Surgery; [2]Pathology, Rieti Hospital, Italy

Introduction

The incidence of primary orbital tumors is rated between 7% and 10%. Primary adenocarcinoma represents about 3% of epithelial tumors of the lacrimal gland and its prognosis is usually poor: 5-year overall survival is about 50% (5). Considering the rarity and biologic aggressiveness of this malignant tumor and the aesthetic and functional problems related to its treatment, a wide range of treatment protocols are available in the literature, such as surgery, radiation therapy with/without chemo. The purpose of this presentation is to describe and analyze the challenges related to the diagnostic and therapeutic path of one case of primary recurrent adenocarcinoma of the lacrimal gland

Case report

A 64-year-old woman was admitted for left increasing recurrent exophthalmos and orbital pain, following two previous surgical treatments for adenocarcinoma of the lacrimal gland G3: two consecutive endorbital excisions of 2.1cm mass (cT2) and total conservative parotidectomy and modified radical neck dissection of levels 1-5 for parotid space extra-capsular single nodal metastasis (cN1-ECS+). At admission new staging was achieved through MRI and 18F-FDG PET/CT. The former evidenced recurrent 1.6cm extraconical nodular mass of the left lacrimal gland, infiltrating the lateral rectus muscle and displacing the eye, without bone involvement. It resulted as highly suspect for recurrent adenocarcinoma at morphological evaluation, gadolinium-enhanced dynamic studies and diffusion-weighted imaging (DWI). The latter confirmed the high metabolic activity at the primary site (Standard Uptake Value SUV: 8) and did not evidence regional and distant metastases. So the tumor was staged as r cT3, cN0 (previous pN1 ECS+), cM0 – St4°A. Salvage surgery of the primary and post-operative radiation therapy of T+N were planned.

With the aid of harmonic scalpel, in order to reduce bleeding, type IV orbital exenteration was performed, according to Meyer and Zaoli classification, which included eyelids, orbital soft tissues and was extended to the lateral wall of the orbit (zygomatic bone), preserving its orbital frame. Frozen sections of the orbital apex resulted free from disease. Reconstruction was achieved combining temporal mio-fascial flap transposition, through the zygomatic osteotomy, with skin graft apposition to the flap.

The Patient was discharged on the 6th post-operative, complication-free, day. Histopathological examination of the resected specimen demonstrated primary adenocarcinoma of the lacrimal gland poorly differentiated, 1.3 cm large, with high mitotic activity and tendency to angiolymphatic embolization, infiltrating the orbital soft tissue, comprehensive the lateral rectus muscle. It was staged as r pT3 (R0).

Post-operative radiation therapy consisted in intensity modulated radiation therapy (IMRT) to the primary site, ipsilateral parotid space and lateral neck, at the global dose of 59.6 Gy at the primary site and parotid space and to the ipsilateral neck (levels 1 to 3).

Six months after the end of adjuvant radiation treatment, the patient is alive without evidence of tumor recurrence at clinical examination and PET/CT scanning. At the present moment orbital prosthesis is going to be manufactured. In accordance with the patient's choice, titanium fixtures with abutments will be implanted to position the prosthesis.

Discussion

Related to the present report, the challanges in the surgical management of the present case of recurrent primary orbital carcinoma, arising from the lacrimal gland, are the following: 1. diagnosis and staging; 2. type of tumor-excision; 3. orbit reconstruction following exenteration: when and how it is required.

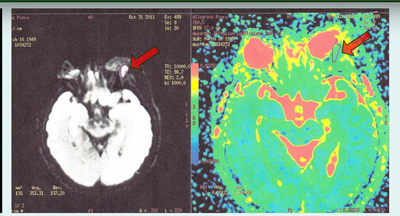
Diagnosis and staging. Diagnosis of orbital mass, in case of "buried" primary orbital lesion of the lacrimal gland and when there is no ocular mobility impairment, orbital pain or reduced visual acuity, consist in surgical trans-septal biopsy, preserving the peri-orbita and the orbital bone. The reason, for this standard procedure was avoided in the present case, is due to the fact that histology of the primary was known and that previous resections had likely left residual disease, so the suspect of recurrency was significant. MRI gadolinium enahanced dynamic study and diffusion-weighted imaging (DWI). Benign lacrimal gland tumors usually are in-homogeneously hyperintense on T2WI. Signs of cortical orbital bone compression are often evidenced in these tumors. Malignant tumors often show nodularity and infiltrative patterns on MRI. They frequently distort the globe and determine bony erosion. DWI was higly suspected roe recurrent malignant disease, even if only a few reports are available concerning DWI in the characterization of orbital tumors (4). 18F-FDG PET/CT evidenced hypermetabolic activity in the orbit with a SUV between 7.2 and 11.4, suggestive for adenocarcinoma (3). For these reasons it was avoided to violate again the orbital space, to achieve a diagnosis of recurrency hardly possible at frozen section examination, increasing the risk of tumor seeding.

Type of resection. no evidence-based treatment recommendation is available in the literature concerning the treatment of carcinoma of the lacrimal gland. The challenge of orbital surgery for malignant lesions is represented by combining a radical free detectable margin resection and function preservation (eye-sparing surgery or exenteration) (2). Limited primary tumors may be radically treated through eye-sparing approaches. Infiltrating or extensive carcinoma requires wide resection, consisting in orbit exenteration (5,6). It has been classified by Meyer and Curioni into 4 classes (1):

Type 1: eyelid skin and conjunctiva are preserved;
Type 2: the eye, the conjunctiva and soft tissue are excised sparing only the eyelid skin;
Type 3: both eyelids and orbital structures are resected;
Type 4: exenteration comprehensive of both eyelids, soft tissue and extended to involved bone structures.

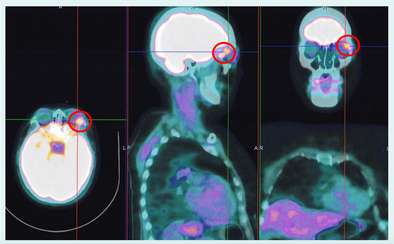
In the present case Type 4 exenteration was performed considering the recurrent disease, the infiltration of orbit soft tissue and the close contact to orbit bone.

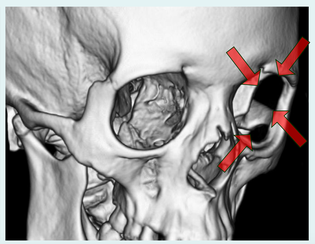
Orbit reconstruction. Reconstruction after orbital exenteration ranges from no-reconstruction to microvascular free-flap transfer. The aims of one-time, concomitant reconstruction are aesthetical, anatomical (to divide orbital space from anterior skull base and paranasal sinuses), and to give adequate cover and fast healing to exposed orbital bone structures, which will be irradiated. Theorically only in some definite cases of exenteration type 4 require reconstruction. All other cases may achieve sequential, spontaneous healing. The fact is that some form of reconstruction is necessary in most cases, considering the high rate of postoperative irradiation following exenteration, which must be started in a 45-60 day, post-operative period. Complete second healing may be longer and the not adequate to protect bone from radiation local side effects. Moreover aesthetic rehabilitation through eye or orbital prosthesis generally results better when neo-orbit soft tissues are regular and trophic. In the present case, the choice to reconstruct the orbit through a pedicled flap, a temporal mio-fascial flap, pulled through the zygomatic osteotomy, preserving the orbital frame, tried to join the necessity to achieve a good reconstruction, in terms of aesthetic outcome and mass of tissue transposition, with the use of an easy and fast to-be-harvested flap.



Imaging

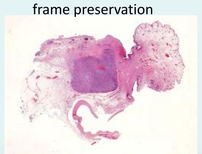
MRI evidences 1.6cm extraconical nodular mass of the left lacrimal gland, infiltrating the lateral rectus muscle and displacing the eye, without bone involvement. Gadolinium-enhanced dynamic studies and diffusion-weighted imaging (DWI) are highly suspected for recurrent adenocarcinoma. 18F-FDG PET/CT scanning confirmed the high metabolic activity at the primary site (Standard Uptake Value SUV: 8) and did not evidence





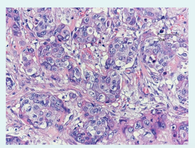
Surgery

1. Pre-op frontal view
2. Pre-op lateral view with incision lines marked
3. Frontal view of the orbita after exenteration and temporal mio-fascial flap transposition, through the zygomatic osteotomy
4. Post-op CT 3D reconstruction showing the zygomatic bone osteotomy and orbital frame preservation



Pathology

5.6. Infiltrating epithelial neoplasm composed of solid sheets of large cells with marked nuclear atypia and scattered mitotic figures. Limited areas of necrosis and focal ductule formation were seen. No evidence of a pre-existing pleomorphic adenoma was found



Post-op

60day-post-radiation MRI (7) and frontal view (8)

Conclusion

The surgical treatment of orbital tumors is a challenging field of head and neck oncology, for the value of the hosted organs and structures, the wide range and rarity of tumors, which may arise in its narrow space, and its allocation between skull base and sinu-maxillary complex. No evidence-based guide-lines are available, so its treatment recommendations come from single institution reports. At the state of the art, radical, free margin resection and postoperative radiation therapy give most chances of loco-regional control. The surgical planning must consider the importance to perform definite time postoperative irradiation and future prosthetic rehabilitation. This means that reconstruction is generally required and has to be reliable and bearable for the patient, in order to reduce the risks of sequela and complications, which could delay post-operative irradiation.

References

1. Croce A, Moretti A, D'Agostino L et al: Orbital exenteration in elderly patiens: personal experience. Acta Otorhinolaryngol Ital, 28, 193-9, 2008
2. Janecka I, Housepian E, Trokel S et al: Surgical management of malignant tumors of the lacrimal gland. A J Surg, 148(4), 539-41, 1984.
3. Jeong H-S, Chung MK, Son Y-I et al.: Role of 18F-FDG PET/TC in management of high-grade salivary gland malignancies. The J. Nucl. Med. 8, 1237-44, 2007
4. Sepahdari AR, Aakalu VK, Setabutr P, Shiehmorteza M, Naheedy JH, Mafee MF. Indeterminate orbital masses: restricted diffusion at MR imaging with echo-planar diffusion-weighted imaging predicts malignancy. Radiology 2010; 256:554–564
5. Skinner HD, Garden AS, Rosenthal DI et al: Outcomes of malignant tumors of the lacrimal apparatus. Cancer, 2801-10, jun, 2011
6. Weizman N, Horowitz G, Gil Z, Fliss DM: Surgical management of tumors involving the orbit. JAMA Otolaryngology Head & Neck Surg, 841-846, 2013