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Case Report

Non-salivary and non-intestinal orbital adenocarcinoma

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ABSTRACT

The non-salivary and non-intestinal adenocarcinomas are glandular tumors that include cases of low grade with low metastatic potential and high-grade ones, with aggressive metastatic potential. We present a clinical case of a 47 years old patient, referred to an ENT specialist because of swelling of the inner corner of right eye. Computed tomography (CT) and magnetic resonance imaging showed a proliferation process. The histopathological examination of the tumor biopsy revealed a high grade, non-salivary and non-intestinal orbital adenocarcinoma. Afterwards we review the literature on the subject. The primary non-salivary and non-intestinal orbital adenocarcinoma is characterized by locoregional aggressiveness, making treatment difficult and often mutilating. The prognosis is generally unfavourable. The clinician must exclude a malignant process of lacrimal sac in any patient presenting with a mass of the inner corner of the eye, with the help of the CT associated with magnetic resonance imaging. At the slightest doubt, the biopsy must be obtained. The treatment is multidisciplinary.

Keywords: Adenocarcinoma, Non-salivary, Non-intestinal

INTRODUCTION

Primary malignant tumors of the lacrimal sac are rare, adenocarcinoma in particular.[1,2] The diagnosis of these malignancies is often delayed, which results from misleading symptomatology, marked by an internal canthal swelling that can be confused with a common dacryocystitis.[1,2] An early diagnosis is leading to suitable treatment results and better functional and oncologic prognosis.[1,2] We present a clinical case of non-salivary and non-intestinal orbital adenocarcinoma.

CASE REPORT

We present a case of a 49 years old patient referred to our clinic for ENT consultation because of a tumefaction of the inner canthus of the right eye that has been developing over one-year period. No risk factors were recorded in medical history. The patient lives in Algeria where he works as a teacher.

In the medical examination, we found a 10 mm large mass at the level of the inner canthus of the right eye associated with a moderate exophthalmia. Ocular motility and visual acuity were normal. There were no other ophthalmological symptoms (Figure 1). Nasal endoscopy showed no pathological findings and cervical palpation did not reveal cervical lymphadenopathy. The patient was in good general condition.

We performed a computed tomography (CT) which showed a lesion at the level of inner canthus of the right eye producing a mass effect on the eyeball with exophthalmos (Figure 2). The findings were suggestive of an inflammatory pseudotumor or lymphangioma.

A biopsy was performed under general anaesthesia, under a microscope. A right internal orbital incision showed a whitish mass, friable, without precise deep margins, adherent to surrounding structures. The periorbita seemed to be infiltrated and thickened. The ethmoid was intact. The histopathological examination of the removed tissue

revealed a poorly differentiated large cell carcinoma, in the paranasal orbital zone expanding from the lacrimal sac.

The patient continued the treatment in Algeria. He received radiotherapy at a total dose of PTV 56 GY. The patient was reconsulted at our clinic 15 months after the radiation therapy. The clinical examination showed a mass at the level of the internal angle of the right eye associated with exophthalmos. The skin over the lesion remained unchanged. Palpebrae were retracted (Figure 3). There was no cervical adenopathy, visual acuity and eye motility were correct and the general condition of the patient was good.

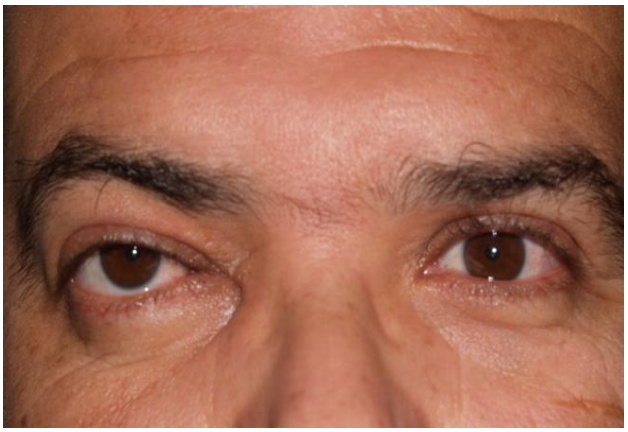

Figure 1: Clinical presentation at first consultation.

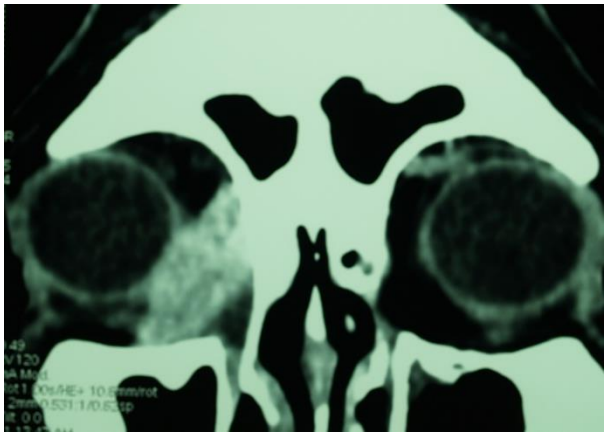

Figure 2: Orbital CT, coronal section: showing a right internal canthal mass without bone lysis.

A new CT scan was obtained after radiotherapy. It showed an extensive periorbital tumor, invading the orbital floor, the ipsilateral malar bone, and the right maxillary sinus. The right ethmoid sinus was completely free (Figure 4).

Imagining was supplemented by magnetic resonance imaging (MRI) of orbits and brain. The findings include a new proliferation process located on the right side expanding towards the right zygomatic arch, the anterior wall of the ipsilateral maxillary sinus, the soft parts of the infratemporal fossa, the subcutaneous fat of the lower eyelid and in reaching the orbit cavity without invading it. There was no brain invasion (Figure 5).

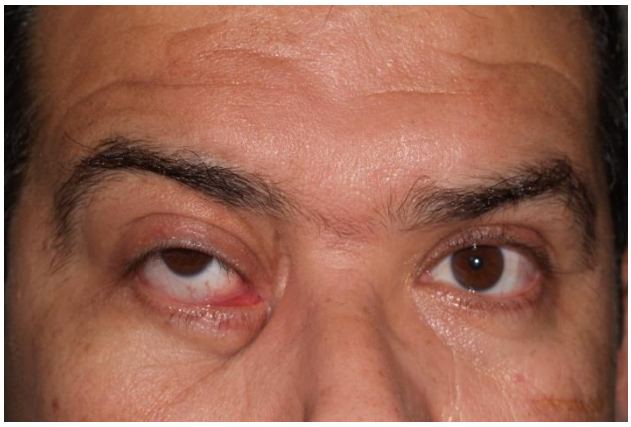

Figure 3: Clinical presentation after radiation therapy.

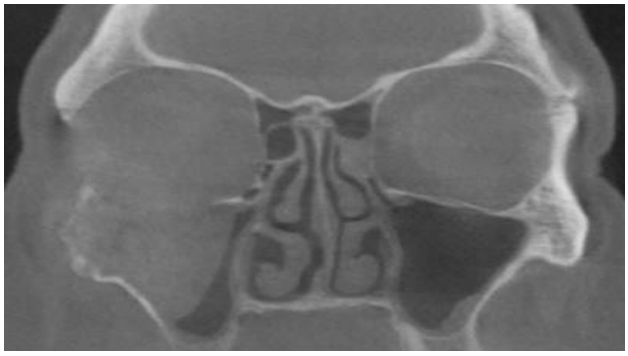

Figure 4: Coronal CT: periorbital tissue mass invading the orbital floor, the malar bone, and the right maxillary sinus.

Based on clinical data and imaging diagnostics, the patient was qualified for surgical treatment. The scope of the operation included a large tumor excision by external approach, with an incision under the orbit extended laterally to the right malar region followed by resection of the orbital floor, the lateral wall, the V2 nerve, the malar bone, anterior 2/3 of the maxillary sinus wall with preservation of the orbital content because it was tumor free (Figure 6).

The pathological examination of the specimen revealed a non-salivary and non-intestinal paranasal high-grade adenocarcinoma which histological characteristics were identical to those of the previous biopsy. The tumor expanded below the mucosa with an advanced trabecular bone infiltration, consisted of glands and cords of cuboidal epithelial cells with cytoplasm and atypical abundant nuclei, showing the characteristics of a poorly differentiated adenocarcinoma (Figures 7-9). The epithelium was not cylindrical and did not produce mucus so that it did not resemble an intestinal tumor

histologically, nor did the appearance recall a salivary gland tumor.

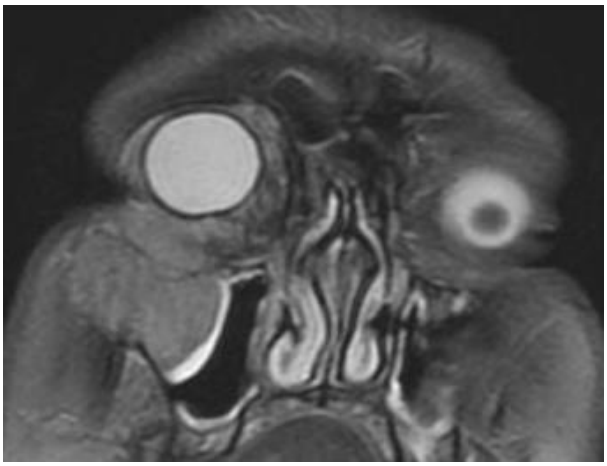

Figure 5: MRI, coronal section: proliferation process with lysis of the inferior and lateral orbital walls, the zygomatic artery invading the ipsilateral maxillary sinus, there is no cerebral involvement.

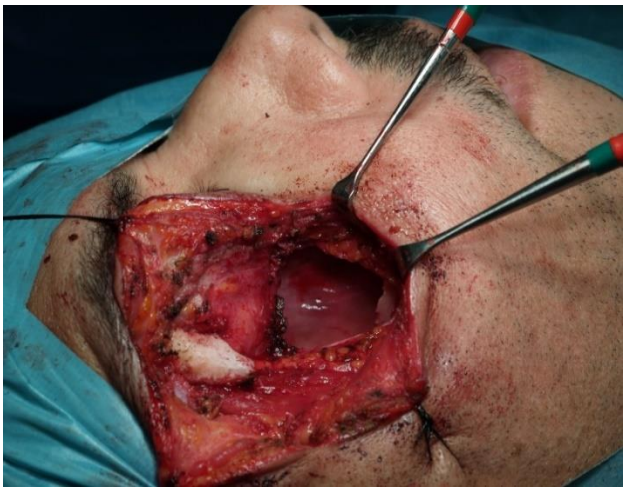

Figure 6: Intraoperative view.

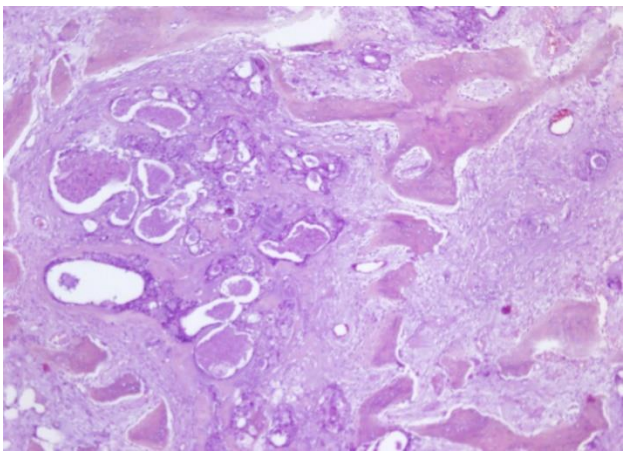

Figure 7: Aggressive adenocarcinoma infiltrating and destroying the maxillary bone.

The patient was consulted seven days post-operative. Postoperative imagining was ordered (Figure 10). MRI revealed a residual tumor located on the rest of the floor of the right orbit measuring 37×46 mm.

We made the decision that patient was eligible for first-line chemotherapy treatment which included three courses of doxorubicin 25 mg/m^2 J1, J2, and cisplatin 25 mg/m^2 J1, J2, J3 in 21 days intervals between courses. Control MRI exam showed a partial response with a 43% regression of the residual tumor.

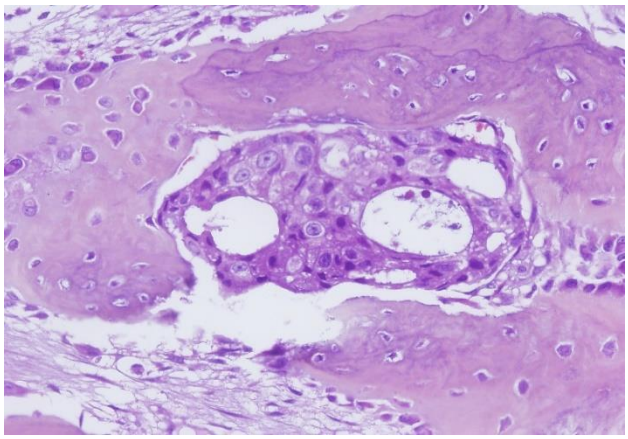

Figure 8: Details of an adenocarcinoma destroying and reshaping the bone.

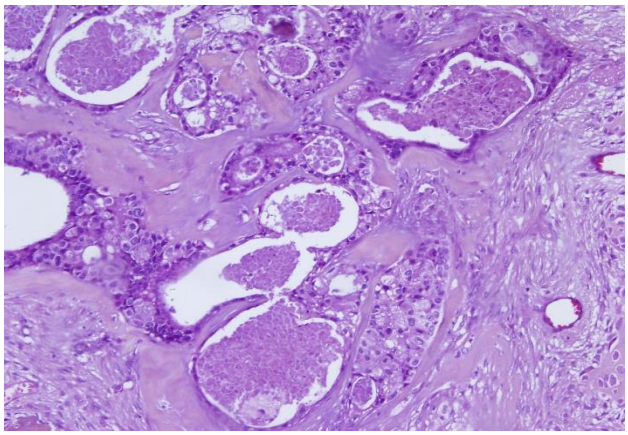

Figure 9: Non-salivary and non-intestinal adenocarcinoma.

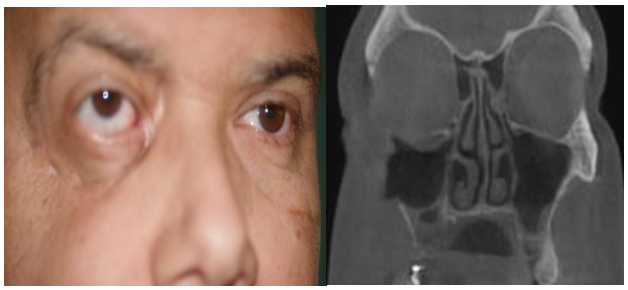

Figure 10: Clinical presentation and CT after chemotherapy.

DISCUSSION

The epidemiology of non-salivary and non-intestinal orbital adenocarcinoma seems to be poorly understood and its prevalence may be superimposable to that of rare malignant sac tumors.[1,2] It is generally considered that there is no ethnic nor gender predominance even though some authors have found that female are in majority.[2] Malignant lacrimal sac tumors usually occur in patients after the age of 50 but can be found in younger patients as well, just like in the presented case with a relatively young patient.[1,3]

The most common symptom is swelling of the internal canthus, which may or may not be associated with exophthalmos. These signs are almost always unilateral.[1,4] The classic epiphora is rarely found at an early stage. The misleading presentation and lack of specific clinical signs often explain the delay of diagnosis.[4] The clinical manifestations are in fact related to the obstruction of the nasolacrimal duct and thus may, in the first place, suggest more common diagnoses such as stenosis of the nasolacrimal duct or dacryocystitis.[4] However, certain aspects must be considered suspicious. The appearance of tears flow stained with blood or tumor fragments appearing spontaneously or after applying pressure on the lacrimal sac, must bring to mind the diagnosis of a malignant tumor. The association with an internal canthal swelling is also suggestive especially if this swelling is persistent.[5]

Most authors agree on the importance and complementarity of CT and MRI.[1-3,6] These two imagining studies allowed us to suggest the diagnosis and facilitate the surgical treatment and the post-therapeutic follow-up. MRI is very suggestive of malignancy and also allows evaluation of intra-orbital margins (relationships with the internal rectus muscles, oblique and orbital fat).[4]

Malignant tumors of the lacrimal sac are most often of an epithelial origin and squamous cell carcinoma is largely predominant. Adenocarcinoma and adenoid cystic carcinoma are less common. Adenocarcinomas develop mainly from the mixed or serous glands present in the lacrimal ducts.[3-5] In our patient, there was a particular type of adenocarcinoma: non-salivary and non-intestinal adenocarcinoma. These adenocarcinomas are usually found in the sino nasal region, which makes the presented case unique. There are three main types: the salivary gland type, the intestinal type, and the non-salivary and non-intestinal type.[7,8] The latter is the most diversified and composed of cases in which morphology does not fit easily into the two previous categories. From the prognostic point of view in this group, we can distinguish between low grade and high-grade adenocarcinomas, classification based on architecture, nuclear characteristics and mitotic activity.[7,8] The ethmoidal and maxillary sinuses tend to be more often affected than other locations.[7,8] However, in our patient, the clinical findings, imagining and histopathological examination favoured a primary malignant tumor of the lacrimal sac extending to the maxillary sinus and the right malar bone. In most cases, these malignant cells would rather come from ethmoid sinous or from another distant organ.

The treatment of non-salivary and non-intestinal adenocarcinoma, like in all orbital malignancies, is multidisciplinary.[6] The therapeutic path depends on the histological type, tumor expansion and the age of the patient, considering that it is a burdensome and mutilating treatment.[5,9] The scope of the surgery ranges from complete tumor excision to orbital exenteration with the removal of adjacent structures. Complete surgical excision is often associated with adjuvant radiotherapy.[7,9] This is the therapeutic approach we considered in our patient; however, he had already received a course of radiotherapy. We, therefore, decided to implement adjuvant chemotherapy following the surgery.

The prognosis of malignant tumors of the lacrimal sac is generally unfavourable and depends on the histological type.[6] In our case, this statement is insufficient to judge the prognosis. However, the results of chemotherapy are very encouraging because after three courses of the doxorubicin protocol associated with cisplatin we managed to obtain a partial response by decreasing the residual tumor by nearly 43%. We, therefore, decided to continue with the same protocol.

CONCLUSION

Non-salivary and non-intestinal primary orbital adenocarcinoma is very rare. Like all orbital malignancies, it is characterized by locoregional aggressiveness, making treatment often difficult and mutilating for the patient. The clinician must exclude a malignant process of lacrimal sac in any patient presenting with a mass of the inner corner of the eye, with the help of the CT associated with magnetic resonance imaging. At the slightest doubt, the biopsy must be obtained and the treatment is multidisciplinary.

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