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CASE REPORT

Ocular Surface plasmacytoma

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ABSTRACT

Conjunctival plasmacytomas are rare tumours that may be solitary or related with multiple myeloma (MM), a malignant systemic plasma cell neoplasm that tends to metastasize. Plasma cell infiltration of bone marrow and presence of immunoglobulin is characteristic of MM. It may cause extraskeletal spread being the ocular tissue involvement very rare.

We report a case of a 57-year-old male patient affected of a caruncular mass that has increased of volume in the past two months. The patient was previously diagnosed of extramedullar affection of MM and he had no treatment in that moment. An excisional biopsy confirmed the diagnostic of a conjunctival plasmacytoma secondary to MM Immunoglobulin D (IgD). Second line systemic chemotherapy treatment produced total regression. However, ten months later the patient was diagnosed of a new expansive lesion originating from the right ethmoidal cells that infiltrated minimally the orbit.

Ocular surface plasmacytoma is an uncommon entity that has to be suspected in a known myeloma patient with a conjunctival mass growth, including the caruncle. The presence of conjunctival plasmacytoma may be a sign of recurrence and insufficient chemotherapy as showed our case. Therefore, regular ophthalmological examinations should be done to the myeloma patients without any treatment.

Introduction

Multiple myeloma (MM) is a monoclonal proliferation of abnormal plasma cells that produces an overproduction of clone immunoglobulins[1]. This abnormal plasma cells tends to metastasize and may cause this extramedullar mass called as plasmacytoma. It is classified as a malignant lesion and it represents only 3% of plasma cell tumour[2]. The more common extramedullary affected organs are the spleen, liver, lymph nodes and kidneys, being the ocular tissue involvement very rare[3]. However, the affection of the eye may be the first manifestation of a MM[4] in some reported cases.

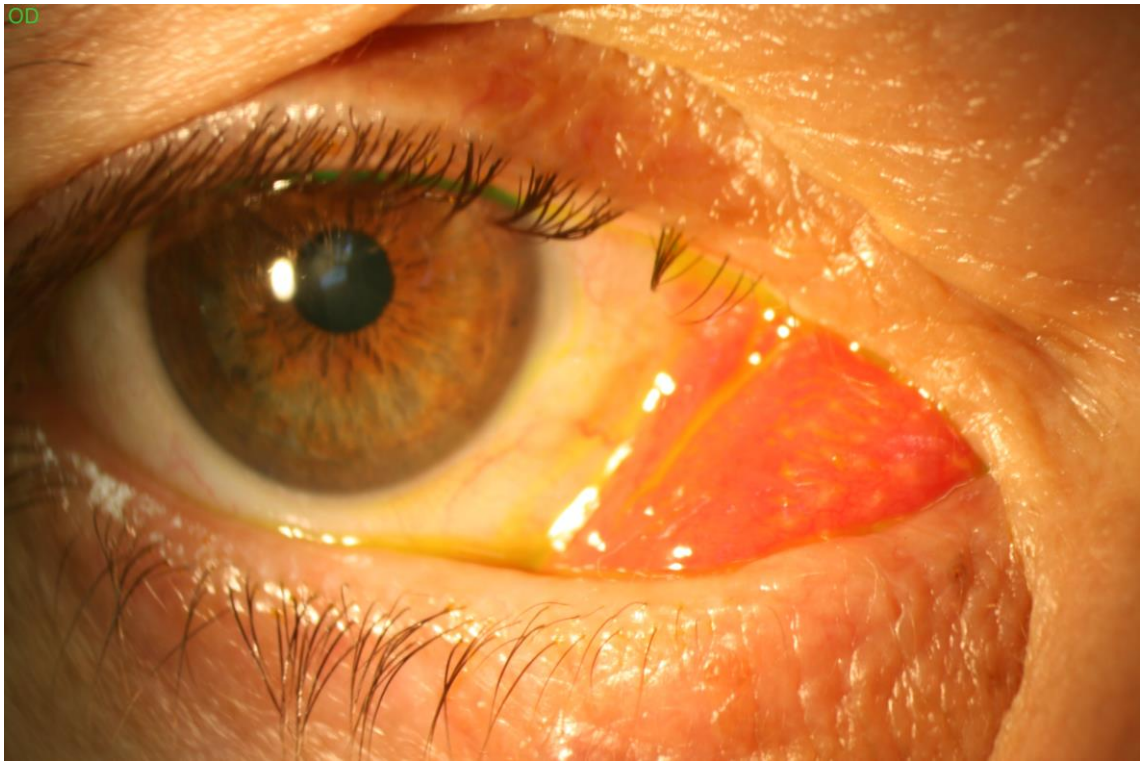
Conjunctival plasmacytomas may be solitary or related to MM[5]. It is usually presented as a mobile, painless, elastic, reddish mass involving a part of the conjunctiva including generally the fornix or bulbar conjunctiva[5,6,7]. However, other clinical presentations have been described in the literature[1,8]. To confirm the diagnostic, it is necessary an incisional or excisional biopsy with supplemental cryotherapy[9]. Histological grading (Grade I to grade III) depends on the maturity and degree of differentiation of the proliferating plasma cells and the frequency of the mitotic figures[10].

In this report, we present a middle-age adult with MM who presented a mass in the caruncle of the right eye. An excisional biopsy was performed with the result of a conjunctival plasmacytoma secondary to MM Immunoglobulin D with monoclonal component lambda light chain.

Case report

A 57-year-old male previously diagnosed of MM with extramedullar spread in pleura and testicles, was submitted to our department with a reddish nodular freshy, painless mass in the caruncle of the right eye. The lesion was progressively increased in size for two months and measured 8 x 6mm when he arrived (Fig.1). At that moment the patient had no treatment due to an acute cholestatic hepatitis secondary to the chemotherapy seven months ago.

Fig. 1: Clinical appearance of conjunctival mass in the caruncle of the right eye.



Previous ophthalmological history include myopic refractive procedure by means of-Lasik in both eyes. The visual acuity was 20/25 in the right eye and 20/20 in the left eye. The differential diagnosis included amelanotic nevus, myxoma, plasma cell granuloma, papilloma, pyogenic granuloma, lymphoma, conjunctival intraepithelial neoplasia and ocular surface squamous neoplasia[1] and conjunctival plasmacytoma among others. An excisional biopsy was decided under local anaesthesia.

Histophatologically, the lamina propria showed a diffuse infiltration by mature neoplastic cells. The morphology of the plasma cells showed a cracked chromatin core with the appearance of socker ball and the cytoplasm was reddish and eccentric with plasmacytoid morphology. Some abnormal plasma cells presented Dutcher bodies (Fig. 2a), nuclear inclusions of immunoglobulins. Immunohistochemical staining showed expression of antibodies to CD138 (Fig.2b), and lambda light chain (Fig.2c). The cells were negative for kappa light chain (Fig. 2d) staining and CD79. The conclusion of the analysis was the diagnosis of a conjunctival plasmacytoma with monoclonal component lambda light chain.

After the biopsy results, the patient was referred again to the haematologist and oncologist. They decided eight cycles of a second line chemotherapy treatment with DARAsc-kd (including daratumumab, carfilzomib and dexamethasone). Three months later, in the ophthalmic examination we observed that the lesion had disappeared completely. The conjunctiva and caruncle were normal appearance with no signs of mass recurrence (Fig.3). During ten months follow-up no regrowth of the lesions was observed.

Fig.2. A: Haematoxylin and eosin histopathologic appearance of the plasma cells (Magnification 60x). **B:** The cells expressing staining CD138 (Magnification 20x). **C:** Positive staining for lambda immunohistochemistry of plasma cells (Magnification 20x). **D:** Negative staining for kappa immunochemistry of plasma cells (Magnification 20x).

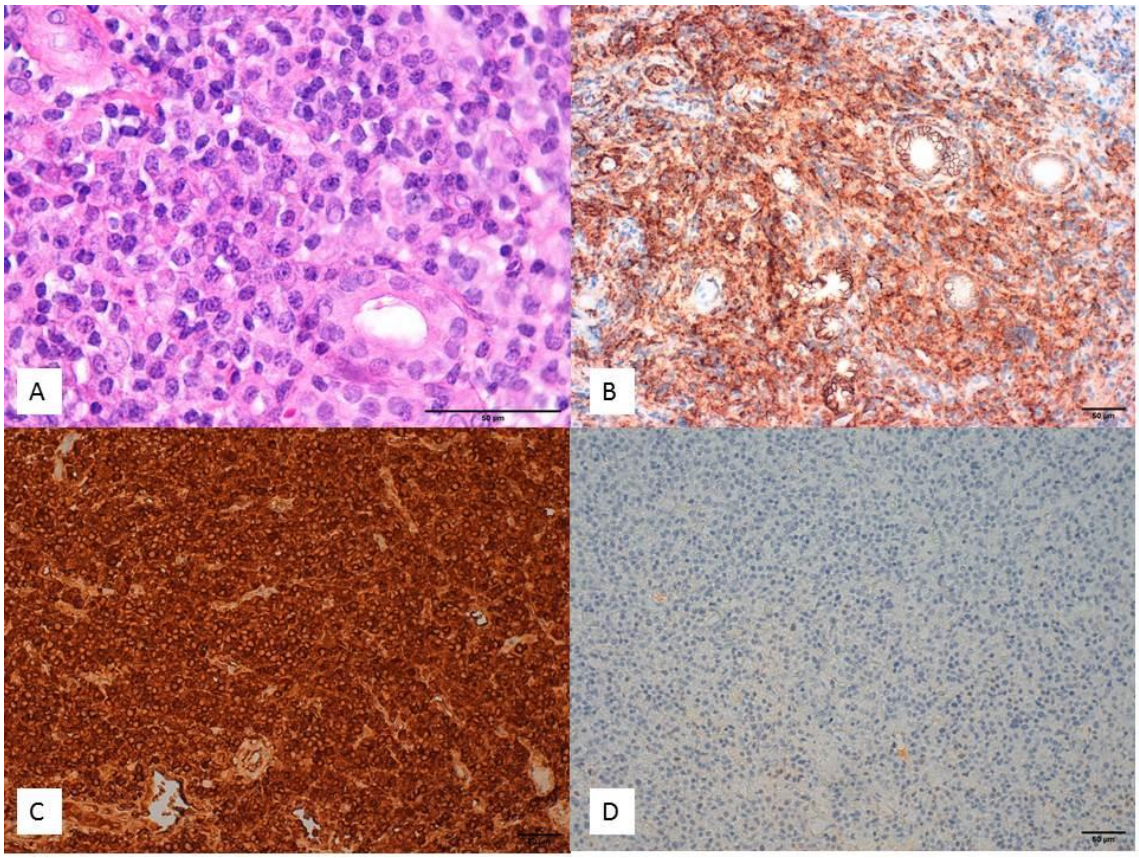
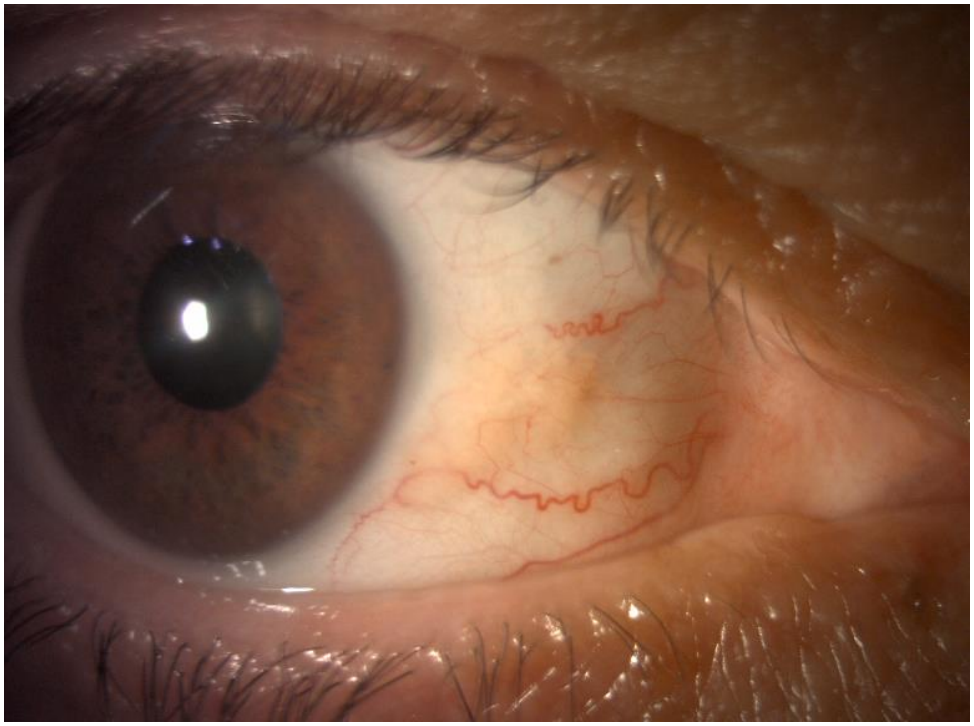
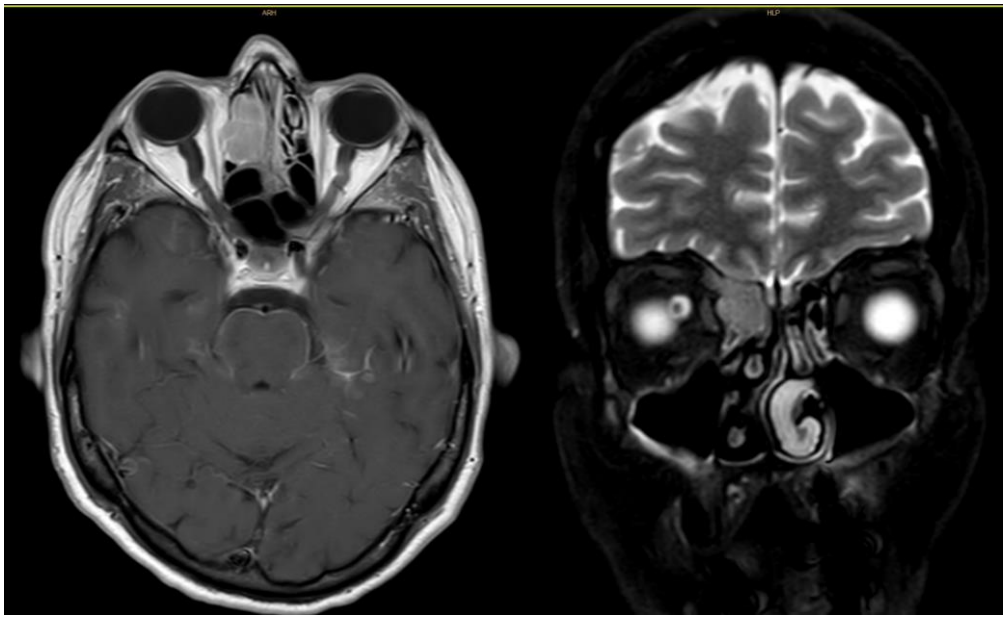


Fig. 3: The appearance of conjunctiva and caruncle three months after excision biopsy.



Nevertheless, the patient was summited again at our department for an episode of blurred vision of the right eye diagnosed as retrobulbar optic neuritis. The magnetic resonance (MR) was performed and showed leptomeningeal spread with a new expansive lesion in the right superior ethmoidal cells that infiltrated minimally the adjacent orbital flat, next to the muscle superior oblique (Fig.4). These findings were compatible with a recurrence of MM involving the orbit. The patient was admitted to the hematologic department for new treatment based on triple intrathecal therapy (TIT) and third line chemotherapy.

Fig.4. A: Axial T1 view of a MR scan showing an expansive mass in the right superior ethmoidal cells infiltrating minimally the adjacent orbital flat, next to the muscle superior oblique. **B:** Coronal T2 view of a MR scan of the lesion.



Discussion

MM represents 1% of all cancers[11] and 10% of the hematologic malignancies[12]. Although rare, it can affect any ocular structure and it may be the first presentation of the disease[4,13]. Rare ocular signs of plasmacytoma include corneal crystals. Moreover, macular serous detachment and retinal hemorrhage have been reported[14]. Ocular manifestations are separated into 2 groups; those secondary to hematologic abnormalities associated with plasma cell dyscrasia and those that are the direct result of tumour growth in ocular structures[1] as showed in our case. The presence of an extramedullary plasmacytoma according to the National Comprehensive Cancer Network is one of the diagnostic criteria of MM and requires regular examinations[15]. The prognosis of the patients with MM is generally poor, with a 5-year survival rate of 45%[15].

Conjunctival plasmacytomas related to MM are exceptional and it may be suspected when a mass growth near the caruncle of the conjunctiva. It is usually presented as a diffuse or localized slightly elevated smooth or multinodular freshy mass[16]. It is generally pink or reddish as described in our case. However other clinical presentations have been described in the literature, such as Ben Artsi et al. that reported a case of a 33-years old female diagnosed of MM 7 years earlier treated with chemotherapy and two autologous stem cell transplantations (ASCT). The patient presented a history of two months gradually enlarging painless salmon-patch lesion in the temporal bulbar conjunctival. It was biopsied and diagnosed of plasmacytoma. She was treated with a melphalan and a third ASCT. The patient became refractory to the treatment and systemic MM progressed[8].

Lymphoid tumors are usually located in the fornix or bulbar conjunctiva. No infiltration of the cornea has been described in the literature[5,6,7]. Sometimes the limbus may be affected with a mild vascular supply or larger dilated conjunctival vessels apparent in larger tumors[9]. Bradley et al. described a case of a patient with known MM who presented two subconjunctival haemorrhages obscuring conjunctival plasmacytoma[1]. For this reason, it is recommended a close follow-up when recurrence subconjunctival haemorrhage appears in a patient with MM because it may be the initial presentation of a plasmacytoma.

Other locations of plasmacytoma in the eye related to MM may be the orbit[17] and exceptionally the iris[5]. Thuro et al. evaluated 30 patients with an ocular plasmacytoma, 60% were diagnosed with MM before and 37% were diagnosed immediately after the presentation of the ocular plasmacytoma. These, shows the close relationship between MM and orbital plasmacytoma and the relevance of the early diagnosis for a systemic screening. Also, they described 4 distinct anatomical patterns; 1) 50% bony plasmacytoma affecting the supratemporal orbit, epidural space and temporal fossa; 23% with discrete orbital plasmacytoma; 3) 13% infiltrative plasmacytoma originating for a sinus, as our case shows; 4) 13% originating from the orbital floor and infiltrating facial soft tissue. All patients had local control of the orbital lesions; nevertheless they have poorer surveillance than the median suggesting the presence of orbital involvement a worse prognosis factor[17].

The standard treatment for systemic MM is based on the radiation of the local lesions, systemic chemotherapy, and bone marrow transplant[5]. The presence of extramedullary manifestations of MM such as orbital plasmacytoma may be the first and only sign of insufficient chemotherapy[18]. In our case, the patient presented a conjunctival plasmacytoma showed up after seven months without any chemotherapy treatment. The presence of an ocular surface plasmacytoma should be a sign of recurrence and insufficient chemotherapy. Therefore, it seems reasonable to do periodic ophthalmological examination when a patient with MM does not have any treatment for a long time.

Conclusions

Conjunctival plasmacytoma is an uncommon entity. Although it can have other clinical presentations, it has to be suspected in a known MM patient with a freshly reddish conjunctival mass growth, including the caruncle as described in our case. Excisional biopsy and histopathology analysis is required to confirm the diagnosis and systemic chemotherapy may be an effective treatment. The presence of conjunctival plasmacytoma may be a sign of recurrence and insufficient chemotherapy as showed our case. For this reason, a regular ophthalmological examination should be done to the patients with MM without any treatment.

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