Malignant Melanoma of the Conjunctiva: about A Case and Review of the Literature

Laminou, L1*, Nouhou Diori Adam2, Magagi A1, Adamou Harissou1, Habou O1, Amza A2
1André Salifou University of Zinder (Niger), National Hospital of Zinder
2Abdou Moumouni University of Niamey (Niger), National Hospital A

Abstract: Purpose: The aim of this study was to describe the management of a case of malignant melanoma of the conjunctiva at the National Hospital of Zinder in Niger and review of the literature. Observation: This was a young adult, 46 years old, male, from Agadez, with an unfavorable socioeconomic status, with no particular history referred for an upper left eyelid tumour. On examination, his general condition was preserved. On inspection, a mass hanging from the free edge of the left upper eyelid was noted; resulting in total mechanical ptosis preventing examination of the condition was preserved. On inspection, a mass hanging from the free edge of the left upper eyelid was noted, resulting in total mechanical ptosis preventing examination of the anterior segment. Palpation of the orbital rim did not note anything in particular, the lymph node areas were free. On the ophthalmological examination, on the right the visual acuity from afar without correction was 10/10 with a normal biomicroscopic examination, on the left the visual acuity was counting the fingers at one meter and a biomicroscopy hampered by the ptosis. The assessment laboratory was normal, the orbitocerebral computed tomography showed the conjunctival origin of the tumor without notion of extension in the vicinity. Macroscopically complete excision was performed. Histopathological examination of the approximately 5 cm surgical specimen noted a malignant melanoma of the conjunctiva with impregnation of the margins (R1 resection). Adjuvant treatment with mitomycin C was instituted. The postoperative course was simple. After 6 months of postoperative follow-up, the ophthalmological examination was normal and the search for metastases was negative.

Conclusion: Malignant melanoma of the conjunctiva is a rare neoplasia that can mimic a palpebral tumor. Histopathological examination remains important for treatment and monitoring.

Keywords: Malignant melanoma, palpebral conjunctiva, mitomycin C, Zinder.

INTRODUCTION

Malignant oculo-orbital tumors are serious conditions, as they can jeopardize not only the visual prognosis, but also the vital prognosis [1]. Malignant melanoma of the conjunctiva is a rare neoplasia, representing 2 to 5% of ocular tumors and 7 to 10% of ocular melanomas with an incidence that varies according to race: per million inhabitants, it varies from 0.18% in melanoderms, 0.17% in Americans, 0.15% in Asians, 0.33% in Hispanics and 0.49% in non-Hispanic Caucasians [2-4]. In 50 to 70% of cases, it develops on acquired primary conjunctival melanosis (PAM) with atypia, but it can also occur on a healthy conjunctiva (de novo melanoma) or more rarely come from the degeneration of a pre-existing conjunctival nevus [5, 6]. Through this study, we describe the observation of a case of palpebral conjunctival melanoma diagnosed and treated at the National Hospital of Zinder.

OBSERVATION

He is a 46-year-old young adult, referred from the Agadez regional hospital center for an upper left eyelid tumor with treatment based on topical and oral anti-inflammatorie. The onset dates back to about 8 months with the appearance of a small mass on the posterior face of the upper left eyelid. This swelling gradually increased in volume, causing mechanical ptosis on the left with the impossibility of opening the
eye. It should be noted that no personal or family history was reported. On admission, the general condition of the patient was preserved. On inspection, there was a granite-like swelling, dark brown, hanging round about 5 cm in its horizontal axis and bleeding on contact, and also hyperpigmented tegumentary lesions on the face (Fig 1A). Lymphadenopathy was not found on examination of the lymph node areas. On ophthalmological examination, the best corrected far right visual acuity was 10/10th and counting the fingers 1 meter to the left (cf 1m). Biomicroscopic examination was unremarkable on the right. The ptosis being major on the left, this examination was difficult on the anterior segment. A radiographic assessment with orbitocerebral CT was performed, objectifying a tumor limited to the upper right palpebral region without local invasion (Fig 1B), a normal chest x-ray. The biological assessment made of an NFS with CRP, an HIV serology was normal. Exeresis, macroscopically complete intraoperatively, was performed (Fig 1C) with cauterization in the tumor bed allowing us to continue the rest of our ophthalmological examination under an operating microscope. The bulbar conjunctivas, the upper and lower fornix, the caruncle, the cutaneous face of the eyelid were free of any tumoral extension. Histopathological examination of the surgical specimen revealed a malignant melanoma of the conjunctiva (Fig 1D) with notification of infiltration of the margins. Because of this positive resection margins, adjuvant treatment with local chemotherapy with instillation of mitomycin C 0.02% eye drops was initiated. At 6 months postoperative, visual acuity without correction was 10/10 on the right and 9/10 on the left with a normal bilateral biomicroscopic examination (Fig 1E). Histopathological examination of the surgical specimen after skin biopsy revealed no precancerous lesion (Fig 1F).
DISCUSSION

 Conjunctival malignant melanoma is a rare but potentially devastating tumour, for which epidemiological studies in the United States and Finland have shown a doubling of its incidence in 30 years, varying between 0.54 and 0.80 per 1,000,000 inhabitants per year [7, 8]. It is a tumor that occurs at any age, but with a predilection interval between 40 and 60 years. The average age of diagnosis of melanoma of the conjunctiva varies between 54 and 67 years, but much lower in Asians and Africans [9] as is the case of our patient whose age of discovery was 46 years. It rarely occurs in children (4% before the age of 20). In 2019, Balzer and al. reported 32 cases of conjunctival melanoma before age 18 and Jenny and al. one case in an 11-year-old girl [10, 11]. The preferred site is fair-skinned adults around 60 years old, and only exceptionally occurs in melanoderm like our patient. Clinically, there are two types of conjunctival melanosis: ethnic melanosis, which concerns 92.5% of black subjects and which presents itself in the form of bilateral brown pigmentation affecting the perilimbal region, the interpalpebral conjunctiva and attenuating at the fornix. It is a physiological state in the melanoderm population that degenerates only exceptionally. The second type of melanosis is primary acquired melanosis (PAD), which has the appearance of pigmented areas ranging from golden yellow to dark brown. Unlike ethnic melanosis, this is unilateral and is acquired in adulthood, as is the case with our patient. The most recent clinical prognostic classification is the 8th edition of the American Joint Committee on Cancer (AJCC) TNM classification. This classification has a good predictive value with regard to the risk of metastatic spread and mortality [12]. Currently there is no standardized management of malignant melanoma of the conjunctiva due to its rarity, but also due to the lack of large-scale studies on this pathology and there is no consensus as to adjuvant treatment and this same in the best equipped health structures [13]. No touch surgery is the gold standard. It consists of removing the tumor as far as the healthy part without touching the tumor with a safety margin of 2 to 3 mm at the level of the bulbar conjunctiva, a wider margin of 3 to 4 mm at the level of the fornix, palpebral or pretarsal conjunctiva and a margin of 5 to 6 mm when other locations are suspected [14]. In the case of conjunctival melanoma involving only the palpebral conjunctiva without involvement of the fornix like ours, the ideal surgical treatment consists of full-thickness resection of the eyelid. But in the case of very advanced melanoma or multifocal or multi-recurrent lesions, orbital exenteration, although still controversial, may be necessary to locally stop the melanoma [15, 16]. The localization of the tumor was in 92% at the level of the bulbar conjunctiva, including 61% in the temporal quadrant [17], and when it was located at the level of the palpebral conjunctiva, it mimicked a palpebral tumor making the diagnosis even more difficult, as is the case with our patient. As adjuvant treatment, several authors recommend cryotherapy at the margins of the excision while others prefer radiotherapy, or chemotherapy such as mitomycin C at 0.02 % or 0.04%, 5-fluorouracil (5- FU 1%), interferon alpha 2b (INF-α2b, 1 million IU/ml), the toxicity of which was considered low and used in the event of a lack of response to cryotherapy, and mitomycin C [18]. We had used mytomycin C at 0.02%, which was the only product available and financially accessible to our patient after tumor excision with a satisfactory result. Conjunctival melanoma is a multi-recurrent tumour, difficult to treat and with a poor prognosis [19]. The mortality rate varied between 7% and 32% patients over 5 years [2] and between 13 and 38% at 10 years [2]. According to the authors, the local recurrence rate varies according to the studies and the type of treatment. At 5 years, it was around 24% when complementary proton beam radiotherapy was associated, 45% if cryotherapy was complementary to surgery [17, 5]. Recurrences occur much more in patients treated with surgery alone than in those whose treatment was coupled with cryotherapy, radiotherapy or local chemotherapy. We had not noted any recurrence in our patient; this could be attributable to the very limited monitoring period. Regional metastases occur to the periauricular and submandibular lymph nodes with a parotid predominance and distantly to the liver, brain, lungs, gastrointestinal tract and skin [2, 5]. Desjardins et al., [9] observed in a retrospective series of 56 cases only one case with metastases at the time of diagnosis. Metastases were reported in a quarter of cases and appear late in the course of the disease, generally a few years after the initial diagnosis, between 3 and 15 years in some authors [3, 20]. We had not yet observed any metastases in our patient, probably due to the relatively short monitoring period.

CONCLUSION

 Malignant melanoma of the conjunctiva can mimic a palpebral tumor. This observation underlines the importance of an anatomopathological examination in front of any ocular tumor in order to determine the precise histological nature and to provide appropriate treatment even in a situation of precarious resources. Monitoring for the purpose of detecting a second cancer should be advised for patients affected by this rare tumor.

Conflict of Interest: The authors declare no conflict of interest.

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