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Malignant Conjunctival Melanoma Developed On Primary Acquired Melanosis Of The Conjunctiva: A Case Report

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Abstract: Conjunctival melanoma is a rare malignant melanocytic tumor with a bad prognosis due to its recurrence and metastatic potential. Conjunctival melanomas "de novo" or occuring on a nevus have a better prognosis than melanomas associated with primary acquired melanosis, which is a major risk factor for local recurrence and metastatic spread. We report the case of a 51-year-old patient with no particular history who consulted for the first time for a swelling of the inner canthus of the left eye and whose excisional biopsy was in favor of a conjunctival melanoma developed on primary acquired melanosis of the conjunctiva, and who had initially benefited from conservative surgery with external radiotherapy followed by secondary exenteration due to a voluminous endo-ocular recurrence during follow-up.

Conjunctival melanoma, primary acquired melanosis of the conjunctiva, conservative surgery, exenteration.

I. INTRODUCTION

Conjunctival melanoma represents approximately 2% of malignant tumors of the eye [1]. The average age of onset is 60 years, the tumor occurs in more than 55-60% of cases on a pre-cancerous melanosis with cytonuclear atypia (primary acquired melanosis), in 20% of cases on a pre-existing conjunctival nevus, and in around 20-25% of cases the tumor occurs de novo [2]. The tumor may develop over the entire conjunctiva. The initial location has a prognostic impact. Therapeutic management of conjunctival melanoma presents a challenge of both local recurrence and lymphatic metastatic dissemination. We report the case of a 51-year-old patient with no particular pathological history who presented with conjunctival melanoma developed on primary acquired melanosis of the conjunctiva of the left eye.

II. OBSERVATION:

A 51-year-old patient with no previous pathological history presented with a raised brownish swelling in the inner canthus of the left eye that had been progressing rapidly over the past four months. Ophthalmological examination revealed a visual acuity at 10/10 P2 in both eyes, and slit-lamp examination revealed a raised, nodular, vascularized pigmented swelling measuring 20*12 mm developing at the level of the caruncle extending over the bulbar conjunctiva, the upper and lower conjunctival fornix and the upper tarsal conjunctiva [Figure 1].

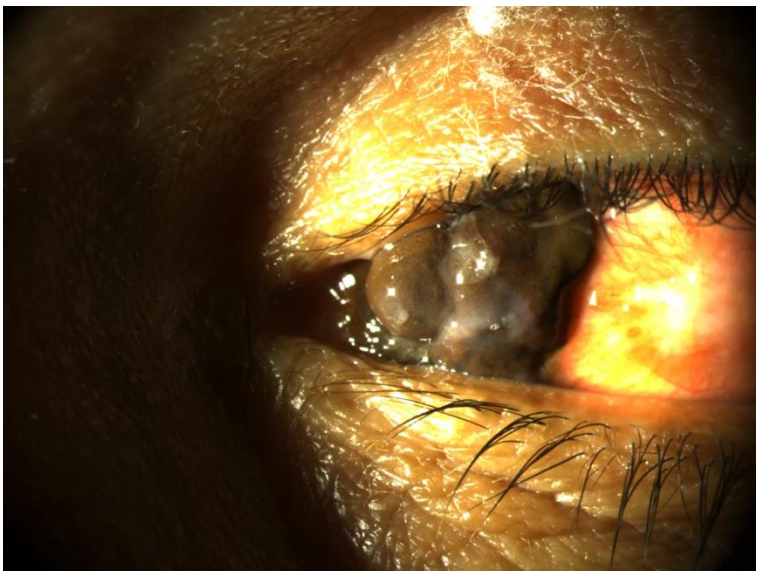

Figure 1 : raised brownish swelling in the inner canthus of the left eye

The examination of ocular surface of the left eye also revealed a extensive dusting of planar pigment on the bulbar conjunctival epithelium. The cornea was clear, the anterior chamber calm and deep, the iris was normal in pattern and color, and the fundus was free of suspicious lesions. On gonioscopy, the iridocorneal angle was open 360 degrees and free of suspicious lesions. Examination of the right eye was normal. General examination revealed a patient in good general condition, and palpation of the regional lymph nodes and parotid gland was completely unremarkable. However, the dermatological examination revealed hypochromic macules on the torso [Figure 2].

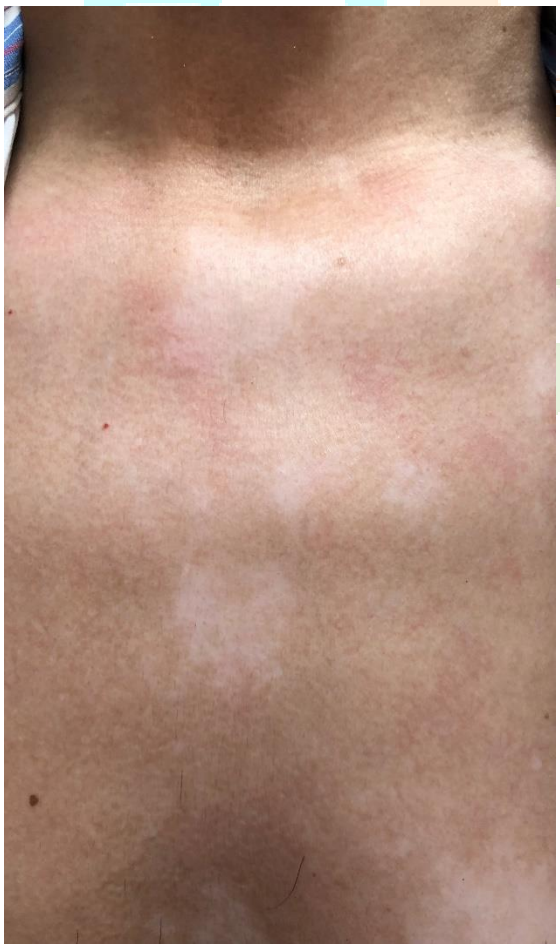

Figure 2 : hypochromic macules on the torso

The patient benefited from treatment combining carcinological surgical excision of the ocular tumour using the 'no touch' technique with wide 3mm safety margins under general anaesthesia, supplemented by amniotic membrane grafting after instrument replacement. The procedure was followed by excision of the inner third of the upper eyelid, completed by an external canthotomy freeing the eyelid, followed by an internal canthopexy with 6/0 prolene stitches [Figure 3].

Figure 3: Postoperative appearance of the left eye one week later

The surgical specimen, measuring 20*12*7 mm, was sent for anatomopathological study. Histological examination revealed a conjunctival mucosa containing a clearly malignant tumour. The tumor was melanistic in nature, characterized by a diffuse cellular layer made up of mostly spindle-shaped elements with scant cytoplasm, tattooed with melanic pigment. Nuclei are markedly enlarged, irregular, hyperchromatic and often nucleolated. The mitotic index was 3 mitoses/10 fields at high magnification, D0-062. The stroma is slightly inflamed, with no deep intravascular tumour emboli. Lateral excisional margins are healthy and increase to more than 2 mm. The deep limit increases to 4 mm. All of which concludes with an infiltrating melanoma of the conjunctiva [Figure 4,5]. The anatomopathological examination of the sample from the pigmented conjunctival area showed an epithelium devoid of any notable maturative abnormalities, partially abraded and resting on a chorion largely occupied by melanin deposits with no identifiable tumour cells and no suspicious lesions, suggesting an appearance conjunctival melanosis. Furthermore, the anatomopathological examination of the skin lesions on the torso showed discrete non-specific chronic inflammatory changes, with no suspicious lesions.

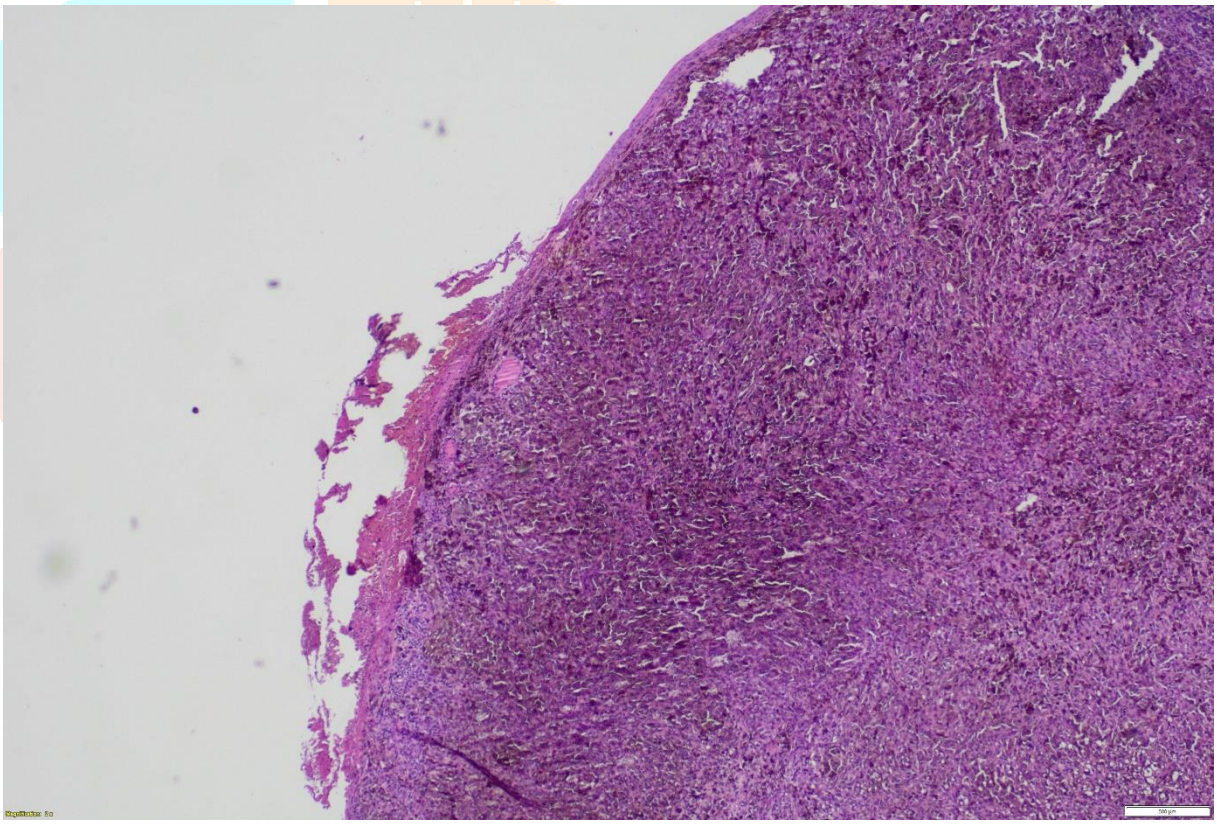

Figure 4: Histological section of conjunctival melanoma, magnification 100

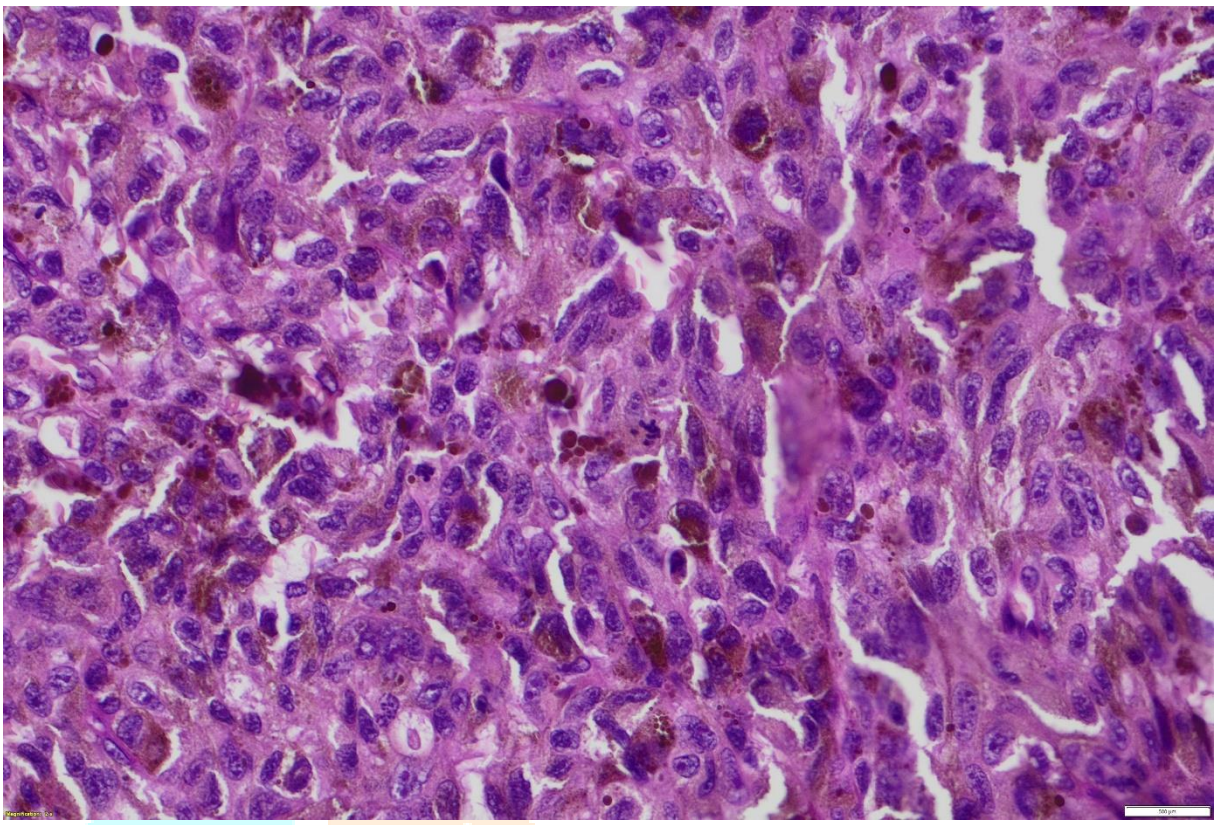

Figure 5: Histological section of conjunctival melanoma, magnification 400

The diagnosis adopted was that of an extensive caruncular tumour of invasive melanoma developed on primary acquired melanosis.
The extension assessment, including chest X-ray, abdominal ultrasound and PET scan, was unremarkable. Orbitocerebral MRI showed no endo-ocular extension [Figure 6].

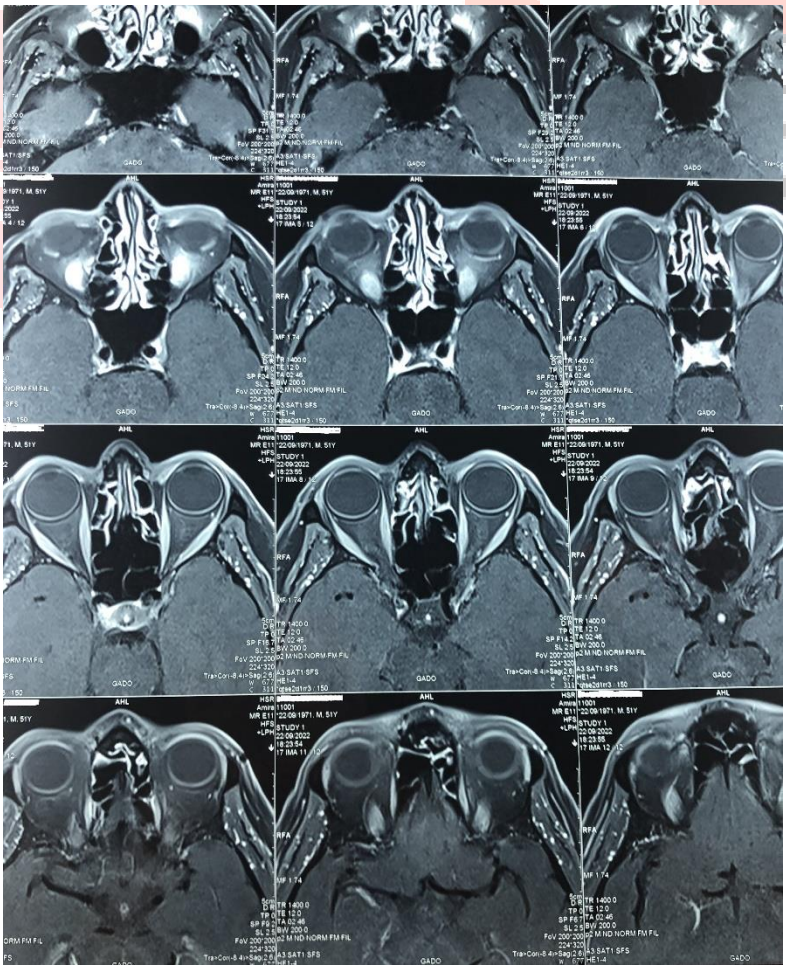

Figure 6: Orbital-cerebral MRI showing no endo-ocular extension

Complementary treatment was decided based on external radiotherapy, as proton therapy or brachytherapy were impossible due to the tumour's location. The evolution was marked by the occurrence of a homolateral recurrence with endo-ocular extension 8 months later. The patient was treated with secondary orbital exenteration with filling of the cavity with a muscle flap and adjuvant radiotherapy [Figure 7].

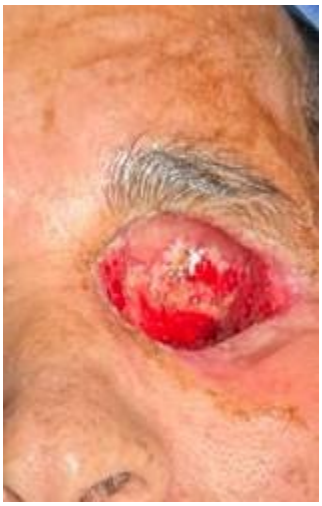

Figure 7: Postoperative appearance after secondary exenteration of the left eye

A full extension assessment was repeated, showing no metastases.

III. DISCUSSION:

Primary acquired melanosis is a pathology of the conjunctiva which presents in the form of a more or less dense, more or less extensive, unilateral plane pigment dusting on the bulbar and palpebral conjunctival epithelium, at the level of the fornix, the caruncle and the semilunar fold. These lesions correspond to a proliferation of intraepithelial melanocytes, and may stabilize or spread to the cutaneous side of the eyelids, or invade the cornea [2,3].

For this reason, strict clinical controls are recommended at least once a year by the same examiner, and even more frequently in the case of lesions larger than 7.5 mm by 10 mm [2].

This examination should include careful inspection of the bulbar and palpebral conjunctiva, with the upper eyelids everted and the lower fornix exposed. it is also necessary to complete by palpating the eyelids in search of nodules as well as the pretragial lymph node areas. In cases of primary acquired melanosis localized in the inner canthus, some authors recommend an ENT examination to detect any associated melanosis of the nasal mucosa [3].

The risk of transformation of primary acquired melanosis into conjunctival melanoma is very low, it is of the order of 1 in 400,000 cases of primary acquired melanosis [2]. However, in 70% of cases, it is the primary acquired melanosis that gives rise to conjunctival melanoma.

The presence of cytonuclear atypia within a primary acquired melanosis constitutes a risk factor of malignant transformation into conjunctival melanoma of the order of 50% [2,3], which is why it is recommended to biopsy any primary acquired melanosis with an inhomogeneous and extensive character.

In case of a positive biopsy, surgical treatment, cryotherapy or local treatment with antimitotic mitomycin eye drops may be required to reduce the risk of degeneration [4].

Conjunctival melanoma is an uncommon ocular tumor, even rarer than choroidal melanoma.

Treated conjunctival melanoma frequently recurs. The frequency of recurrence estimated between 51% and 56% of cases at 10 years [5]. Recurrences are located either at the initial tumour site, or distantly due to the migration of malignant cells along the lacrimal rivers [6].

The occurrence of metastases is mainly related to the mean maximum thickness of the tumour, the number of recurrences, and their rate is estimated at 26% at 10 years according to studies [5].

The pretragial, submandibular and cervical lymphadenopathy which are most often invaded must therefore be systematically palpated during the clinical surveillance examination.

The first prognostic factor is the topography of the conjunctival melanoma [2]. Localization in the bulbar conjunctiva in the area of the palpebral cleft and in the sclero-corneal limbus is the most frequent, accounting for 57% of cases, and has a better prognosis [5, 6], probably because the lesion is more easily visible and therefore detected early. In contrast, conjunctival melanoma located in the fornix, caruncle or palpebral conjunctiva poses a problem of delayed diagnosis and treatment.

The second prognostic factor is the nature of the treatment [5]. Treatment is usually conservative, combining surgical resection and radiotherapy of the episcleral margins and deep plane. Amniotic membrane grafting after conjunctival resection is a technique that both facilitates healing and monitoring thanks to its transparency [7]. Adjuvant treatment such as radiotherapy or local treatment with an antimitotic such as mitomycin is justified due to the low excision margins seen with the conjunctival situation of the melanoma, in order to reduce the risk of recurrence [8].

The combination of surgery and radiotherapy has a better prognosis than surgery alone [6].

For some authors, the third prognostic factor is the histological type of melanoma [6]. In multivariate analysis, the only factor statistically influencing the risk of local recurrence and the risk of metastatic dissemination is the association with a primary acquired melanosis [9].

Treatment of conjunctival melanoma is usually conservative. Exenteration is sometimes indicated in cases of locally advanced tumor inaccessible to conservative treatment and/or associated with primary acquired melanosis and/or if recurrences are voluminous or if their number is significant. This enables local control of the cancerous pathology, but no benefit of exenteration on patient survival has yet been demonstrated.

IV. CONCLUSION:

Primary acquired melanosis being susceptible to degeneration must be considered as a precancerous lesion. It is essential that all patients with primary acquired melanosis should receive regular ophthalmological surveillance. In case of doubt, recourse to biopsy must be systematic; in the presence of atypia, an excision must be carried out otherwise radiotherapy or local treatment with mitomycin C. The association of melanoma with acquired primary melanosis is a major risk factor for local recurrence and metastatic spread. Hence the importance of strict follow-up every 3 to 6 months for 2 years, then every 6 months to 1 year for 5 years, with semi-annual lymph node ultrasound and annual PET scan. Treatment of conjunctival melanoma is currently conservative, but exenteration may be considered depending on the number of recurrences, or in the case of massive tumors exceeding any conservative therapeutic resource.

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