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Report of a case of malignant melanoma of the limbus (30-thy years of observation)

Przypadek czerniaka rąbka rogówki (30 lat obserwacji)

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Summary: Purpose: To describe a case of a malignant melanoma of the limbus.
Material and methods: A 49-year-old female patient with a suspected malignant limbic melanoma in the left eye. The first signs appeared in 1975, manifested as reddening of the left eye on the temporal side with markedly dilated conjunctival vessels. In 1993, the lesion became pigmented and nine years later its progression was noted. In May 2003, lamellar keratoplasty of the left eye was performed under general anaesthesia.
Results: Histologic assessment – spindle cell malignant melanoma. The entire tumour was resected and the cosmetic result was very good. The results of accessory diagnostic studies were within normal limits. Over a 28-month follow-up period no local recurrence or metastases to other sites were observed.
Conclusion: Patients with discoloration of the conjunctivae should be regularly evaluated by an ophthalmologist and an oncologist.
Słowa kluczowe: czerniak złośliwy rąbka, knaloplastyka warstwowa, melanocyty.
Key words: malignant limbic melanoma, lamellar keratoplasty, melanocytes.

Introduction

Malignant melanoma is a tumour arising from melanocytes, the cells containing enzymes required to synthesize the pigment melanin, which during embryogenesis displace from the neural crest to the skin, mucous membranes, central nervous system and the eye. About 10% of melanomas are localised in the eye. They arise from the uveal melanocytes in the choroid (80%), the ciliary body (12%), the iris (8%) or the conjunctiva (less than 1%), are the most common ocular malignancy in adults. The most common sites within the conjuntiva are the lacrimal caruncle and the semilunar fold. Malignant melanoma of the cornea, which most commonly arises from the limbus or the adjacent bulbar conjunctiva (1), is relatively rare.

Methods and material

A 49 years old Caucasian female patient was admitted to the Department of Ophthalmology, Medical University in Warsaw, in May 2003 with a suspected melanoma of the limbus in the left eye. She was otherwise healthy. In 1975, when pregnant, she noticed some reddening of the left eye on the temporal side with marked dilatation of the blood vessels and sought an ophthalmologist's opinion. Since then she had been regularly evaluated by an ophthalmologist. In 1993, the lesion (2x2 mm in size), became pigmented. The patient did not give her consent to surgical excision of the lesion and cryopexy was performed three times. For the next nine years no progression of the lesion was observed, but in 2002, the lesion was found to have enlarged (2x3 mm), and become darker. B-mode ultrasonography (32 MHz) of the left eye detected an echogenic homogenous lesion within the limbus, affecting the corneal epithelium, which also involved the bulbar conjunctiva, but did not invade the sclera. Another B-mode ultrasonography performed six months later confirmed progression of the lesion. Surgery was advised to which the patient gave her informed consent. On admission: visual acuity 20/20 bilaterally, IOP 21 mmHg bilaterally. The right eye: the anterior and posterior segments were normal. The left eye: a pale eyeball, a network of dilated conjunctival vessels on

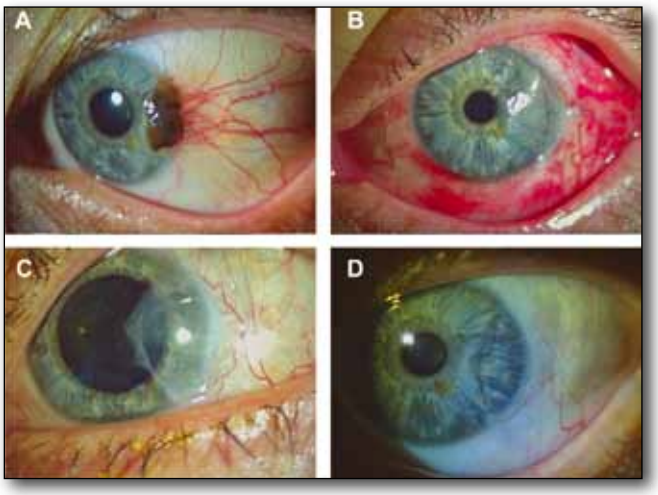

Fig. 1. The left eye: A – On admission. B – The first 24 h after surgery. C – One month after removal of corneal sutures, seven months after surgery. D – Twenty months after surgery.
Ryc. 1. Oko lewe: A. Przy przyjęciu. B. 24 godz. po operacji. C. 1 mies. po usunięciu szwów rogówkowych, 7 mies. po operacji. D. 12 mies. po operacji

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the temporal side, in the cornea, at the limbus an elevated, vascularized, pigmented tumour measuring 2.7x6.5 mm was extended outside the limbus and involved the cornea 2.5 mm from the limbus. The remaining cornea and other structures of the eye were normal, as was the posterior segment (Fig. 1A).

Lamellar keratoplasty of the left eye was performed under general anaesthesia. After surgery, topical corticosteroids and topical antibiotics were used. A histopathological examination of the resected tumour was performed.

Results

In the early postoperative period the visual acuity was 20/30, the temporal aspect of the eyeball was deeply injected, the graft in the temporal cornea as well as the own cornea were transparent, the other segments were normal (Fig. 1B). Histologic assessment: spindle cell malignant melanoma – pT1 pNx pMx, Breslov 0.7 mm (tumour cells stained with S100 and HMB45). Single tumour cells superficially invaded the cornea. The entire tumour was excised with a margin of healthy tissue (Fig. 2A, 2B, 2C).

One month post surgery, the visual acuity was 20/20. The graft became slightly opaque. After six months, extended network of conjuntival vessels on the temporal side and the graft of decreased transparency and with ingrowing blood vessels on the limbic side could be seen. The corneal sutures were removed and one month after that the cornea was transparent with subsiding neovascularization on the temporal side (Fig. 1C). No recurrence was found in 20-month follow-up (Fig. 1D).

Discussion

Malignant melanoma of the cornea may arise from primary acquired melanosis (PAM), a preexisting benign pigmented nevus or de novo (2). Sudden appearance of one or more nodules in a so far flat lesion and spread beyond Bowman's membrane with involvement of the corneal stroma, suggest malignant transformation of PAM (3). In 20% of cases, malignant melanoma develops from a pigmented nevus. Any lesion that increases in size, extending beyond the limbus is suspected. Primary conjunctival melanoma usually develops at the beginning of the fifth decade of life. In our patient the abnormalities, such as conjuntival congestion and dilated blood vessels were first noticed when she was in her early twenties, while pigmentation was observed after 18 years of follow-up. The standard treatment consists of surgical excision of the tumour with an appropriate margin of healthy tissue combined with cryo- or laser therapy. Pigmented tumours limited to the epithelium may be treated by resection of the epithelium with the lesion (3). A melanoma involving the superficial layers of the corneal stroma may be excised using superficial keratectomy combined with cryotherapy or lamellar keratoplasty, which is a safer and less traumatizing method than penetrating keratoplasty though technically more difficult. A lamellar transplant allows a regular, smooth surface and exercises a beneficial trophic effect on the cornea. The cosmetic result is very good. When the melanoma invades the cornea deeply, penetrating keratoplasty is required while enucleation of the eyeball or orbital exenteration are the most radical methods. The melanoma may spread haemato- or lymphogenously or via the lacrimal duct into the nasal cavity (2). When metastases are present, which may occur in any organ (most commonly the lymph glands, brain, liver and lungs) (4), palliative treatment and chemotherapy are used. In the case presented in this report, the results of all accessory diagnostic

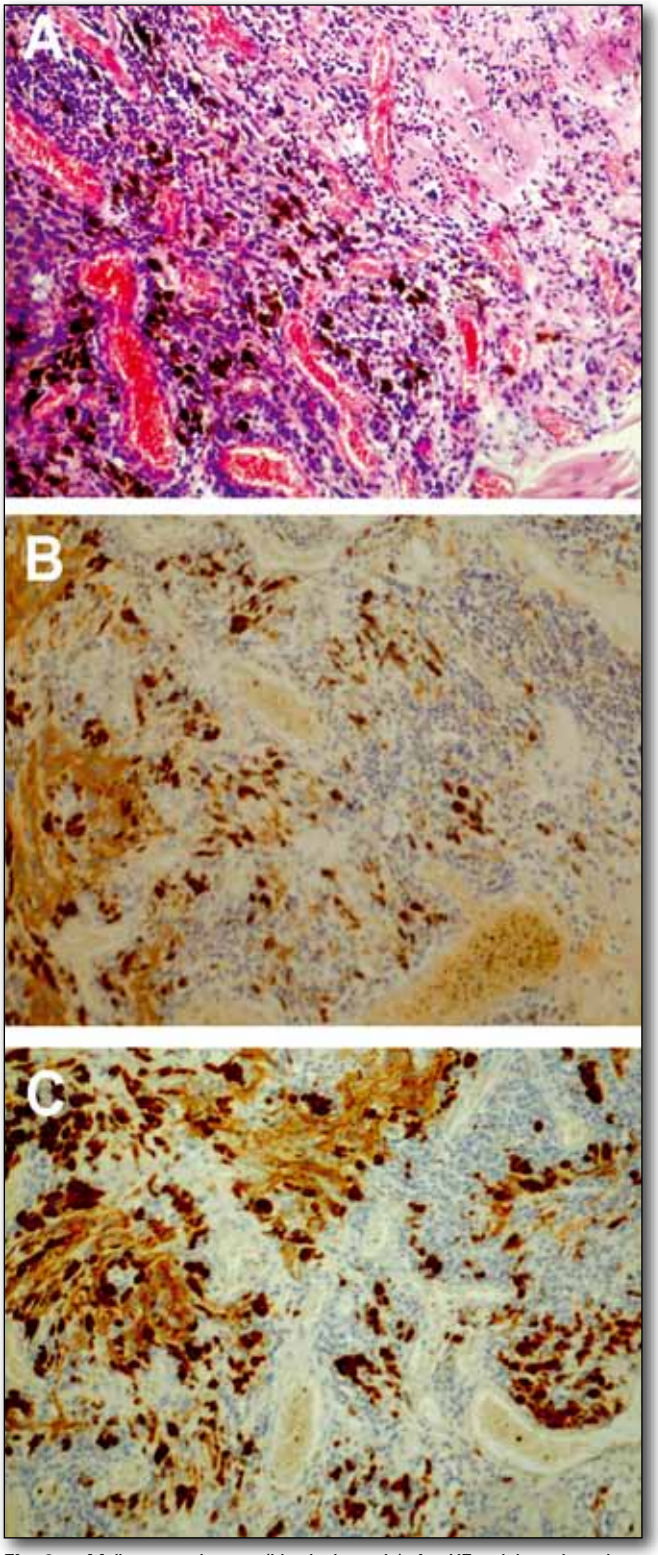

Fig. 2. Malignant melanoma (histologic study). A – HE staining – large lymphocyte infiltration, good prognosis. B – Staining with S 100 monoclonal antibodies. C – Staining with HMB 45 monoclonal antibodies.

Ryc. 2. Czerniak złośliwy (badanie histologiczne). A – barwienie HE – naciek z limfocytów olbrzymich, dobre rokowanie. B – barwienie przeciwciałami monoklonalnymi S 100. C – barwienie przeciwciałami monoklonalnymi HMB 45.

studies (NMR scan of the head, transaminase levels, chest X-ray,) remained within normal limits. The essential studies were performed every 6 months. Over a 20-months follow-up period, no local recurrence or metastases to other sites were observed.

Conclusion

The development of the condition over a long period time clearly demonstrates that patients with discoloration of the conjunctiva, should be regularly evaluated by an ophthalmologist and an oncologist.

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