IRIS MELANOMA IN PAEDIATRIC PATIENT

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SUMMARY

Iris melanomas are very rare in children. The paper presents the case of a 12-year-old boy with iris melanoma of spindle-cell type A and pre-operatively associated with uncontrolled secondary glaucoma. Tumour resection was complicated by lens extraction and consecutive PC IOL implantation. Antiglaucomatous surgery was performed due to the persistently elevated intraocular pressure. Currently, the intraocular pressure is within the normal range and the patient has no signs of the presence of tumour cells or metastases.

Key words: iris melanoma, spindle-cell type, secondary glaucoma

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INTRODUCTION

Malignant melanoma occurs very rarely in childhood age. More than 50% of malignant melanomas in children are located on the iris.

In 80% of cases they are localised in the lower half of the iris. The base of the tumour is larger than 3 mm and protrudes more than 1 mm. Pigmentation of iris melanoma may be of varying degrees – from pronouncedly pigmented to amelanotic. A rarer variant is tapioca melanoma – amelanotic multinodular tumour [6].

Melanoma is manifested by marked progression and alteration of its shape, size, and pronounced vascularisation. In the case of suspicion iridectomy with excision of the tumour is necessary [3].

The following types of uveal melanoma are differentiated in Callender's histological and cytological classification: spindle-cell type A, spindle-cell type B, epitheloid and mixed. Callender's histological and cytological classification of uveal melanomas began to be used in 1931 in order to determine prognosis, and was more precisely modified in 1983 [1, 7].

The most common form is the spindle-cell, less malignant type. The epitheloid histological type has a worse prognosis. Growth of a diffuse melanoma through the trabecular meshwork or the settlement of tumour cells in the chamber angle may cause secondary glaucoma. A typical manifestation is the generation of a sector cataract [6].

CASE REPORT

At the beginning of March 2014 a 12 year old boy suffered a trauma to the right eye from a figure of a tin soldier, after which he experienced reddening of the eye and a deterioration of vision. He did not report to the district outpatient department of ophthalmology for an ocular examination until the end of March 2014. Due to suspected tumour of the iris he was immediately sent to the Department of Ophthalmology at Olomouc University Hospital for an examination. There he was diagnosed with suspected iris melanoma and secondary glaucoma, for which antiglaucomatous therapy was indicated.

At the district outpatient department the finding was monitored from a preventive ocular examination from February 2010, in which the ocular finding was bilaterally physiological. According to information from the boy's mother, the formation on the iris appeared as early as in 2011, and did not progress markedly (fig. 1, 2).

The patient was sent to our centre by our colleagues in Olomouc for further examination and subsequent treatment. The first examination at our centre was conducted at the beginning of April 2014. Uncorrected visual acuity (UCVA) in the right eye was 0.3, best corrected visual acuity (BCVA) 1.0. Both UCVA and BCVA in the left eye were 1.0. During hospitalisation we repeatedly measured increased intraocular pressure, on average 29 mm Hg, even during therapy with a triple combination of antiglaucomatous agents.

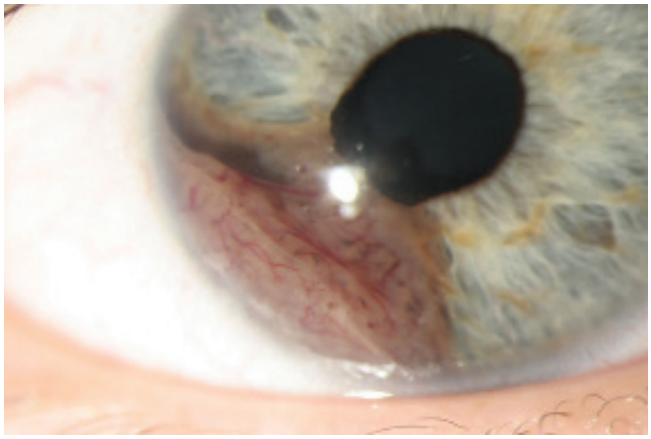

Fig. 1 Iris melanoma of right eye before surgery.

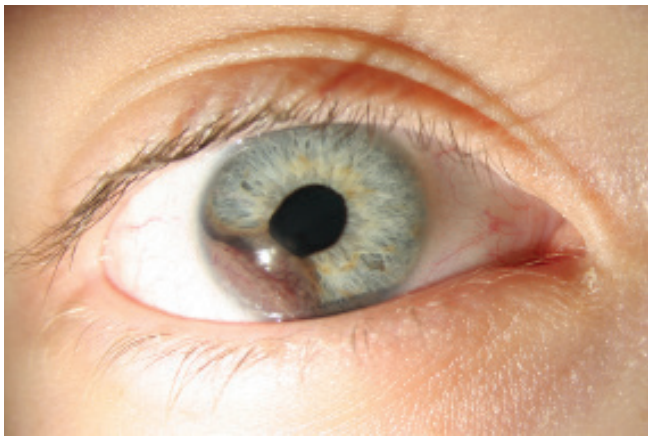

Fig. 2 Iris melanoma of right eye before surgery.

Biomicroscopically the anterior chamber from 6 to 9 o'clock was filled with a vascularised pigmented tumorous matter. The pupil had a size of 4 x 4 mm and slight deformation of circular shape around 7 to 8 o'clock, where ectropion of the pigment epithelium of the iris was located.

Upon an ultrasonic biomicroscopic examination an ovoid, slightly echogenic lesion was identified on the iris between 6 and 9 o'clock, reaching as far as the root of the iris. At its apex the lesion pressed against the endothelium, its base reached a size of 7 mm limbally and prominence was 2.8 – 3.4 mm, radially up to 4.6 mm. No manifest invasion into the ciliary body was evident.

Upon MR of the brain and orbit a discrete defect of the anterior chamber of the right eyeball was displayed, without post-contrast saturation, otherwise the finding was without further pathology.

Due to initial refusal of surgical treatment and subsequent illness, resection of the tumour was not performed until June 2014 (fig. 3).

Perioperatively the iris melanoma reached to the edge of the ciliary body. As already mentioned in the results of the ultrasonic biomicroscopy, invasion into the ciliary body was not evident. This was confirmed also during the operation. As a result quadrant iridectomy was performed without partial resection of the ciliary body. Resection of the iris was performed in the healthy tissue with a sufficient safety zone in sano. Subsequently, following the operation full body PET/CT was performed, upon which there was no presence of tumour residue in the coloboma, and complete resection of the tumour was confirmed.

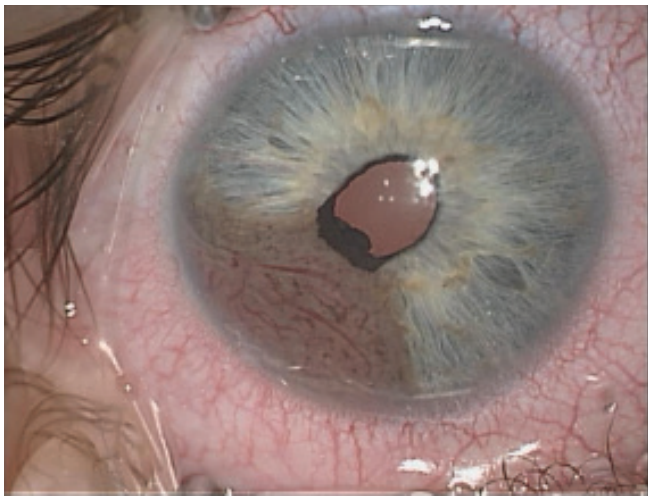

Fig. 3 Iris melanoma of right eye in operating theatre shortly before resection

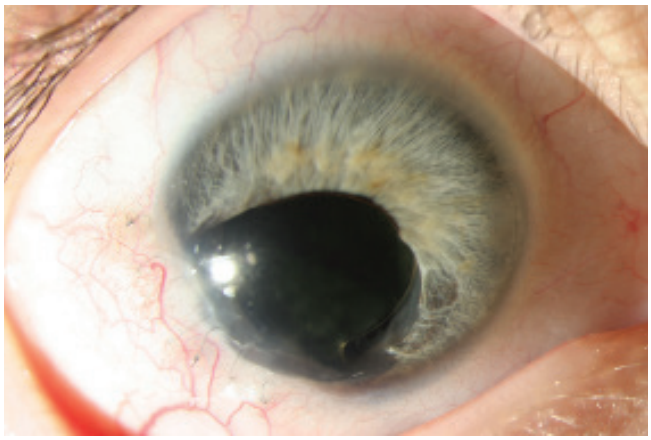

Fig. 4 Right eye after excision of iris melanoma and replacement of lens.

Upon quadrant resection of the iris tumour within the scope of the entire lower temporal quadrant, it was also determined that the tumour was growing into the anterior chamber of the lens. Partial peripheral cataract was also present in the given quadrant. As a result, after evaluation of the finding we performed anterior capsulorhexis, irrigation/aspiration of the lens and implantation of a PC IOL into the capsule (fig. 4).

Histologically iris melanoma was confirmed, spindle-cell type A, prognostically favourable variant. A microscopic and immunohistochemical examination of the resected tissue of the size of 4x3x2 mm was performed by the Institute of Pathology at the University Hospital in Brno and the Institute of Pathology and Molecular Medicine of the 2nd Faculty of Medicine, Charles University and Motol University Hospital. With regard to the confirmed diagnosis, the patient underwent a complex oncological examination and remains to date within the care of the Department of Paediatric Oncology.

After surgery we supplemented full body PET/CR, MR of the brain and orbit and complex laboratory examination. None of these methods determined pathology, all the results were negative. Mutation of the gene BRAF, NRAS, TP53 was not demonstrated, but positive mutation Q209L of gene GNAQ was determined. The tumour markers were negative. On the basis of the results of cytogenetics and molecular genetics, the oncologists did not indicate chemotherapy or other adjuvant therapy.

Postoperatively and upon combined antiglaucomatous local and general therapy, it was not possible to compensate intraocular pressure long term. At the beginning of October 2015 we performed trabulectomy with mitomycin C and

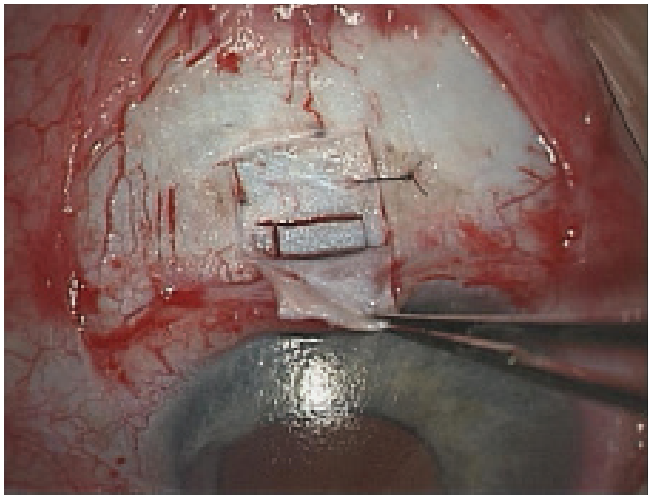

Fig. 5 Right eye during course of 2nd operation – trabulectomy with mitomycin C and basal iridectomy.

Fig. 6 Right eye after 2nd operation – trabulectomy with mitomycin C and basal iridectomy.

basal iridectomy due to decompensated glaucoma and progression of glaucomatous changes on the disc of the optic nerve (fig. 5, 6).

At present there is normotension upon local antiglaucomatous therapy, and BCVA of the right eye is 0.1. Even despite general and local preoperative and postoperative corticoid therapy, fibrotisation of the posterior capsule of the lens occurred. As a result it shall be necessary in future to perform sparing posterior capsulotomy.

The patient is without signs of the presence of tumour cells or metastases.

DISCUSSION

Iris melanoma in childhood age is a rare disorder, nevertheless in 1951 Duke and Dunn published a study of 28 eyes with iris melanoma, of which 3 were diagnosed in children [2].

In 2012 authors from Thomas Jefferson University in Philadelphia compiled a retrospective non-randomised study on 317 eyes with iris melanoma. The study included 24 children, in which a child was defined as a person aged under 20 years. There was no statistically significant difference in sex, laterality, localisation or metastasising in any of the age groups. There was a statistically significant difference in the age groups in terms of therapy. In children local resection was performed more frequently, and in adults radiotherapy was more frequent. In paediatric patients the base of the tumour was smaller, with a lower incidence of secondary glaucoma, and tapioca melanoma was diagnosed significantly more often in children [9].

In 2009 authors from the Leiden University Medical Center in the Netherlands published an interesting case report of a boy treated from the age of 12 years for melanoma in the lower quadrant of the iris, spindle-cell type A. Following the performance of local resection of the iris melanoma, sector iridectomy was performed. A phakic iris implant was produced to order, which reduced postoperative photophobia, and postoperative UCVA was 1.0 [4].

In differential diagnostics it is necessary to exclude spindle-cell nevus, adenoma of the pigment epithelium of the iris, leiomyoma, melanocytoma, iris metastases and juvenile xanthogranulomatosis.

A range of Czech and Slovak authors have dealt with the issue of uveal melanoma [5, 8, 10, 11, 12], but to date the rare case of iris melanoma in children has not yet been published in the Czech and Slovak literature.

CONCLUSION

Despite the good prognosis for iris melanomas, quick and correct diagnosis is of essential importance for patients. Larger size of a tumour of the iris represents one of the unfavourable prognostic factors. Upon correct diagnosis and timely total resection, malignant iris melanoma of spindle-cell type A has a very good prognosis, without the necessity of enucleation of the eyeball and further oncological treatment. However, inter-disciplinary co-operation and regular monitoring of the patient is necessary.

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