tumor size, extraocular extension and chromosomal abnormalities in the tumor tissue (primarily monosomy of chromosome 3 and abnormality of chromosomes 6 and 8) [170].

Diagnosis of choroidal melanoma

Choroidal tumors are identified and distinguished almost entirely (in 98% cases) on the basis of ophthalmoscopy (funduscope, Volk lens) and ultrasound US examination (10 MHz US and 35.50 MHz UBM). Recently DRI-OCT evaluation has proved useful in certain situations when a reliable diagnosis cannot be established. This evaluation helps to identify the actual nature of the lesion under review or to confirm an initial diagnosis. Additional data is provided by angiography (indocyanine and fluorescein angiography), autofluorescence and photography in red-free light [31, 103–107].

Characteristic features, diagnostic aspects and differentiation of malignant and benign intraocular tumors are necessary to make diagnosis. Choroidal melanoma may be recognized and differentially diagnosed using simple and readily available diagnostic methods [1–3].

The basic ocular examination comprising a measurement of visual acuity and intraocular pressure, slit-lamp examination of the anterior segment of the eye and dilated-pupil ophthalmoscopy helps to identify whether a tumor is present and if so, what its nature is. Making an appropriate diagnosis requires knowledge of morphological properties and location of the tumor in the ocular fundus, which is typical for some lesions, an ultrasound scan and characteristic manifestation of choroidal tumors in the DRI OCT image.

History of the present illness also plays an important role. In most cases, medical history is not characteristic but helps to initially guide the diagnostic process. A sudden loss of vision in a diabetic or hypertensive patient may suggest a hemorrhage into the vitreous chamber, a gradual deterioration of visual acuity in a patient aged 60 and more and a sudden loss of vision may indicate a subretinal hemorrhage secondary to AMD while a defect in the visual field is suggestive of a retinal detachment (including a secondary retinal detachment related to a choroidal tumor). Medical history of a systemic diseases (including tumors in the patient's family) may also be helpful in establishing a diagnosis.

There is a close correlation between the pigmentation of a choroidal melanoma and the type of melanin. Tumors of light colors contain more pheomelanin while intensively pigmented tumors (black, russet) contain almost exclusively eumelanin. Sometimes a single tumor displays a range of pigmentation. Tumors are usually clearly demarcated from the surrounding tissue, and the retina at the base of large tumors may be detached and, above the tumor, lifted. Tumor enlargement may be accompanied by a hemorrhage on the surface or at its base, and sometimes by a hemorrhage in the vitreous body.

The most important examination is A and B-scan ultrasonography US (10 MHz US), which helps to establish an accurate diagnosis and measure the tumor, assess the surrounding structures, and identify whether sclera is not ruptured and extraocular extension of melanoma UM is present [108].

An ultrasound scan also helps to precisely measure the thickness and width of tumor base in a longitudinal and parallel presentation, the parameters which are important for therapy planning and assessment of its effectiveness.

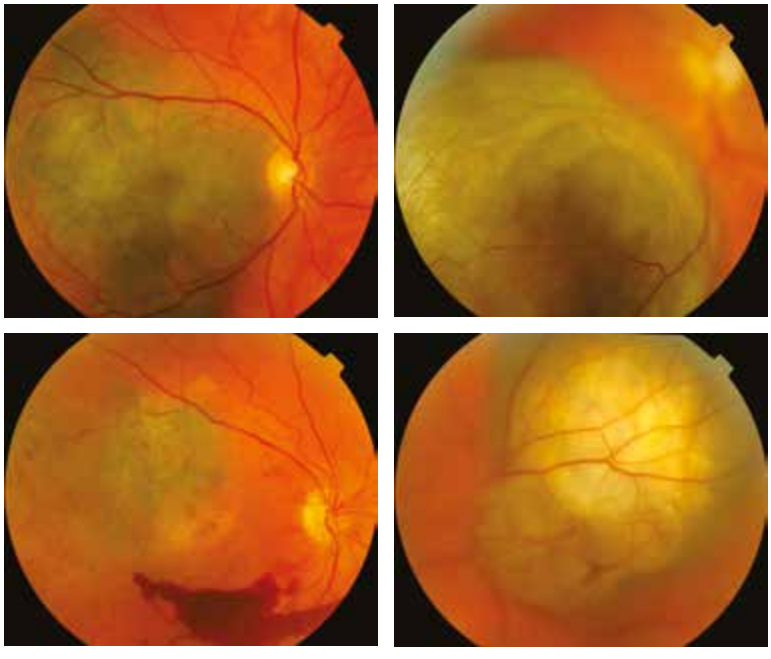
Choroidal melanoma, due to its homogeneous histological structure, shows a regular internal pattern and a low internal reflectivity in an ultrasonographic examination. Pulsating vasculature may be seen at the base of the tumor. A-scan shows a characteristic low reflectivity of the tumor with a high initial peak from the retina. Tumors may appear flat, dome-shaped or mushroom-shaped. Some melanomas have an irregular shape. Mushroom-shape, typical of large choroidal melanomas, is caused by a tumor enlarging above the retina after Bruch's membrane is ruptured.

A and B-scan ultrasonography is a valuable examination informing a differential diagnosis between uveal melanoma and nevi, choroidal metastases, choroidal melanoma, choroidal osteoma and a variety of other benign and malignant tumors. Choroidal hemangioma has multiple, small vascular caverns which form a regular structure and show a high internal reflectivity in an ultrasound US examination. Choroidal hemangiomas show a significantly higher reflectivity than uveal melanoma and higher reflectivity than metastatic tumors. Choroidal metastases have an irregular, chaotic arrangement of cells and varying distances between them which is shown as an irregular internal structure and a variable internal reflectivity in an ultrasound scan. Choroidal osteoma shows a very high reflectivity (resembling that of osseous tissue);

a B-scan presents an intensive shadow in the orbit. When an extensive extraocular invasion has been identified by means of ultrasonography US (10 MHz US), magnetic resonance imaging of the orbits is helpful for assessing its size. Orbital MRI is also recommended after enucleation of eyes with choroidal melanoma involving implantation of an orbital implant.

In cases which appear challenging for a diagnostician, additional methods may prove useful, including: SS-OCT (showing the choroid), transillumination (diaphanoscopy) helpful in diagnosing ciliary body tumors, infrared photography, autofluorescence and indocyanine angiography. The most critical issue for therapy planning and the patient's future is to distinguish a choroidal melanoma from choroidal metastases and benign tumors, choroidal hemangioma. At present, the ability to obtain SS-OCT images of choroidal lesions often helps to diagnose tumors of unclear origin.

6.185



6.186

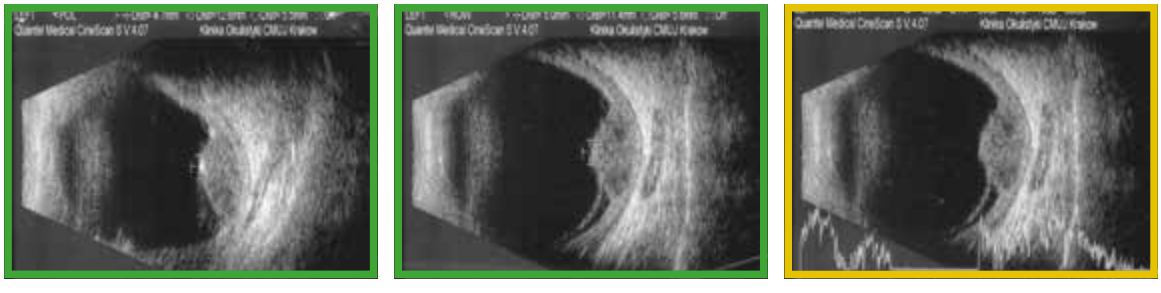
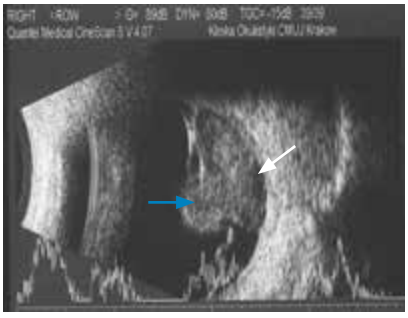
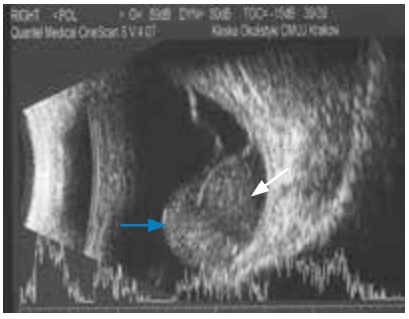


Figure 6.185. Choroidal melanomas with different degree of pigmentation.

Figure 6.186. Dome-shaped choroidal melanoma in 10 MHz US (green frames). Medium level internal reflectivity of the tumor and secondary retinal detachment in presentation (yellow frame).

6.187



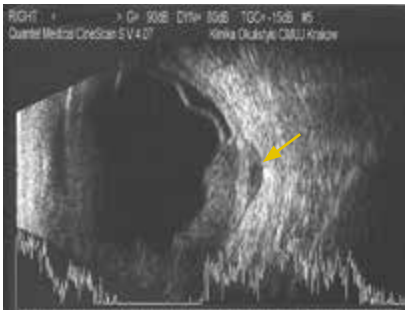


a

6.189

6.188

b



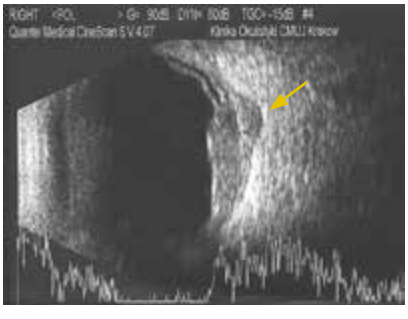


Figure 6.187. Mushroom-shaped choroidal melanoma in 10 MHz US. Initially dome-shaped tumor (white arrows), disruption of the Bruch's membrane (blue arrows).

Figure 6.188. Mushroom-shaped choroidal melanoma with secondary retinal detachment in 10 MHz US. Low internal reflectivity at the tumor base (white arrow) and medium internal reflectivity at the tumor apex (blue arrow).

Figure 6.189. Diffuse choroidal melanoma located in the posterior pole with extraocular extension: **a** – color photograph; **b** – 10 MHz US – diffuse flat tumor with extraocular extension (yellow arrows).

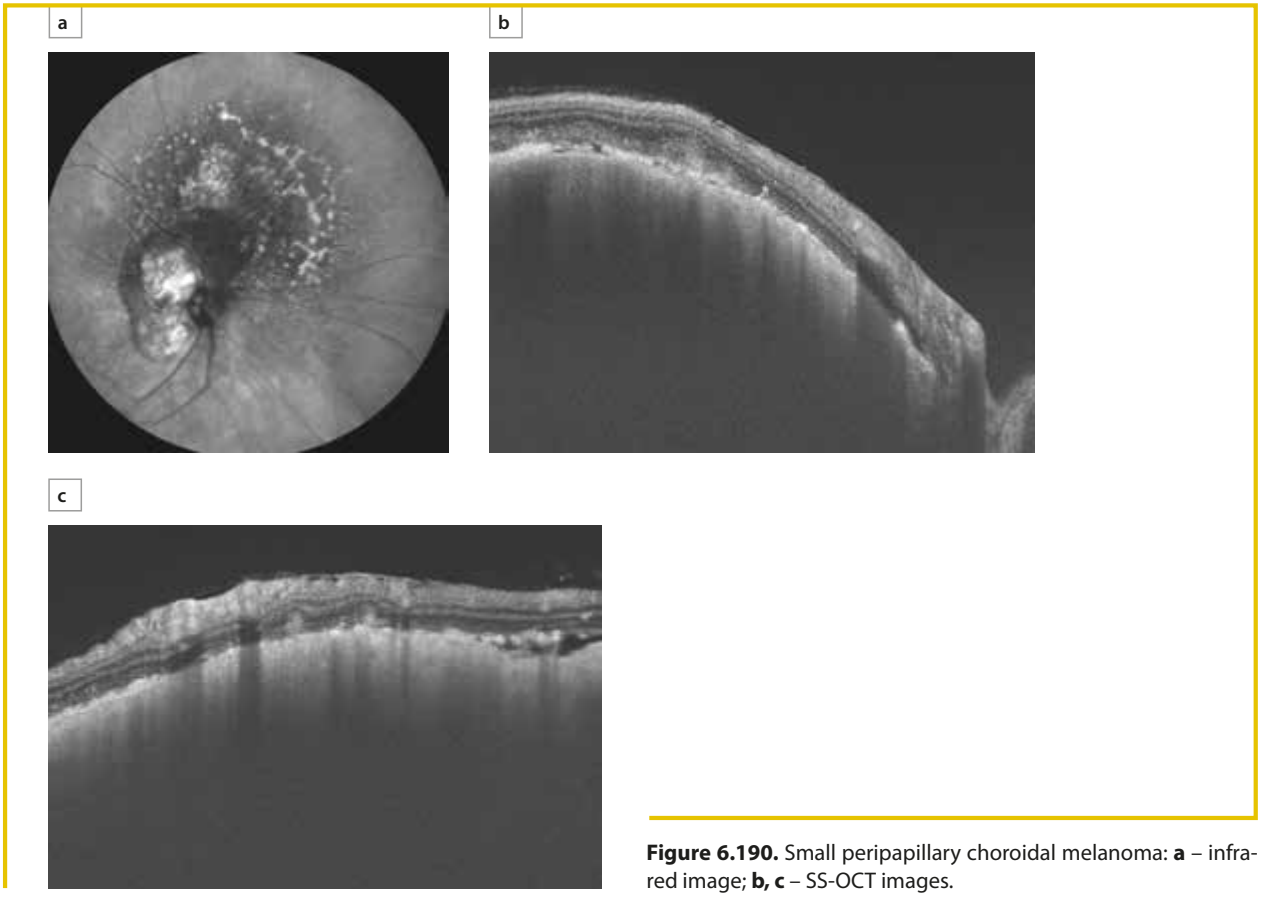


Figure 6.190. Small peripapillary choroidal melanoma: **a** – infrared image; **b, c** – SS-OCT images.

Ultrabiomicroscopy (UBM)

When diagnosing neoplasms of the anterior segment of the eye (iris and/or ciliary body), high-frequency ultrasonography US (35 and 50 Mz), called ultrabiomicroscopy (UBM), is very useful and conclusive. It provides a high-resolution image, which makes it possible to obtain a detailed image of a tumor, and paths of extension through the sclera and surrounding structures, and to measure the tumor.

A differential diagnosis of intraocular tumors by means of ultrasonography with a 35-50 MHz probe should take into account:

- tumors of iris and ciliary body;
- tumors of peripheral choroid;
- cysts of iris pigment epithelium and ciliary body.

Indocyanine angiography (ICGA)

ICGA is much superior to fluorescein angiography in imaging the vasculature of a choroidal melanoma. This is attributable to the different light absorption properties of these two dyes. In addition, indocyanine is absorbed by serum proteins and does not leak through choriocapillaris fenestrations, thus helping to visualize the vasculature. ICGA is considered to be a supplementary examination in a differential diagnosis of a small melanoma. Maximum fluorescence of vessels is achieved after ca. 18 minutes from an intravenous dye administration.

Choroidal melanomas display a heterogeneous fluorescence (hypo-, iso- or hyperfluorescence) depending on the intensity of tumor pigmentation. Choroidal metastases present a homogenous diffuse hyperfluorescence with a late isofluorescence. Choroidal hemangio-