Case report

Circumscribed choroidal hemangioma diagnosed by ultrasonography
An retrospective analysis of 40 cases

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Abstract

Circumscribed choroidal hemangioma (CCH), a relatively rare benign tumor, can cause differential diagnostic problems by its atypical appearance at the time of presentation. Ancillary test such as fluorescein angiography, indocyanin green angiography, ultrasonography, computer tomography (CT), magnetic resonance imaging (MRI) bring not always additional information and their results are not always pathognomic. We retrospectively reviewed the clinical records of 40 patients with the ultrasonographical diagnosis of CCH compiled in the last 20 years. Ultrasonography appeared to be uniform in its lesion characteristics and reliable in the diagnosis. During a mean follow-up time of 7.4 years there were no clinical or posttherapeutical developments that made a correction of the diagnosis necessary.

Introduction

Circumscribed choroidal hemangioma (CCH) is a rare benign primary intraocular tumor with an unclear pathogenesis, probably already present at birth [1–3]. The tumor can be detected and well diagnosed with an accurate indirect ophtalmoscopic or biomicroscopic examination if present in its characteristic appearance of an orange-red or salmon colored smoothly elevated lesion with indistinct borders.

It is nearly always located in the temporal quadrants posterior to the equator two disc diameters or less from the fovea or optic disc [4–7]. The prolonged presence of the tumor frequently gives rise to secondary changes in the overlying retinal pigment epithelium (atrophy, proliferation, fibrous and osseous metaplasia) and in or on the neuroretina (cystic degeneration, neovascularization, serous detachment) [2, 6, 8, 9]. These changes, eventually combined with some enlargement of the lesion cause on the one hand the symptoms (blurred vision, metamorphopsia) and on the other hand diagnostic confusion by the atypical aspect and by the late time of presentation [10, 11]. In the past this could result in an enucleation of the eye because of a clinically expected choroidal melanoma [1, 2, 5, 12]. At the present time because of the increased awareness of the clinical features of CCH by the trained ophthalmologist and in some questionable cases the improvements in the diagnostic possibilities (fluorescein angiography, indocyanin green angiography, ultrasonography, computer tomography (CT) and magnetic resonance imaging (MRI)), the frequency of an erroneous diagnosis has been greatly reduced [4, 7].

Despite these modern techniques, the limited experience of the individual ophthalmologist with regard to this tumor still gives rise to hesitation and consequently extensive or even overdone diagnostic work-up. This study reports on these diagnostic problems and their resolution by an already well-known non-invasive technique with good results: ultrasonography.
Table 1. Ultrasonographic characteristics of CCH.

<table>
<thead>
<tr>
<th>Topographic</th>
<th>* dome shaped, close to disc, macula</th>
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<tbody>
<tr>
<td></td>
<td>* low profile (prominence mostly &lt; 6 mm)</td>
</tr>
<tr>
<td></td>
<td>* base/prominence ratio &lt; 3</td>
</tr>
<tr>
<td></td>
<td>* no choroidal excavation</td>
</tr>
<tr>
<td></td>
<td>* sec. ret. detachment: frequent</td>
</tr>
<tr>
<td>Quantitative</td>
<td>* high reflective (80-100%)</td>
</tr>
<tr>
<td></td>
<td>* regular</td>
</tr>
<tr>
<td></td>
<td>* if osseous metaplasia: attenuation/shadow</td>
</tr>
<tr>
<td>Kinetic</td>
<td>* no vascularity</td>
</tr>
</tbody>
</table>

Methods

We reviewed retrospectively over a period of 20 years (1974-94) the clinical records of the patients with the ultrasonographical diagnosis of CCH and searched for the histopathological diagnosis of CCH in all the enucleated eyes with a clinically suspected malignancy in the same period of time.

The ultrasonographic examination was performed with standardised A-mode (Kretztechnik 7200MA) and contact B-mode equipment (Bronson-Turner/Grumman, Ophthascan S/Biophysique Medical) with an unfocused 8 MHz and a focused 10 MHz transducer respectively. When the lesion was detected, the prominence, the internal reflectivity, the spike-regularity, the attenuation and the eventual signs of vascularity were examined with the A-mode instrument and method as described by Ossoinig [13]. With the B-mode technique the base, the shape, signs for calcification and choroidal excavation of the mass lesion were studied. The results were photographed (Polaroid film 667) for accurate documentation and measurements. When the tumor showed the ultrasonographic characteristics of Table 1, the diagnosis of CCH was made until recall.

Patients

During the observation period not a single eye was diagnosed as CHH by histopathological exam. 324 Eyes were enucleated, 318 of them harbouring a uveal melanoma. In 5 cases a single metastatic lesion and in one case extensive SHMD were the reason for an erroneous removal of the eye.

The ultrasonographic signs of CCH were seen in 40 patients, 17 female, 23 male, with 18 times a tumor location in the right and 22 times a tumor location in the left eye. At the moment of the tumor detection their age ranged from 15 to 79 years (mean 49.2 yrs).

Most of the patients (83%) presented with the symptoms and signs of the tumor in the 4th to 6th decade, 50% in the 5th decade.

Results

Of the 40 patients referred to our department 21 were send by a general ophthalmologist and 19 came from different university clinics. The provisional diagnosis was 'tumor' in 23, CCH in 12, retinal detachment in 3, central serous retinopathy in 1 and cataract in 1 case after general ophthalmological examination.

In only 2 of our patients, the tumor was nasally adjacent to the disc. In all the other patients the lesions
were located in the temporal quadrants close to the fovea and the disc or partially subfoveal. The main reason for diagnostic confusion was in 9 patients the color of the lesion (pale, white-grayish), in 8 patients secondary pigment alterations and in 4 patients an extensive retinal detachment. In 1 patient the media were opaque. With the additional information of the fluorescein angiography there was still uncertainty about the diagnosis in 14 cases. In 2 patients a computer tomogram (CT) and in 4 patients a magnetic resonance image (MRI) was made but not helpful for the diagnosis. Ultrasonography showed in all cases the same ultrasonographic pattern as described in Table 1 and shown in Figs 1 and 2. The tumor-prominence varied from 1.5 to 6 mm (mean 2.8 mm), the tumor basis from 3 to 13.5 mm (mean 7.2 mm). The ratio tumor basis/tumor prominence was < 3 in all cases.

Enlargement of the lesion was observed in 2 cases, detection of superficial calcium deposits (osseous metaplasia RPE) in another 2 patients. The maximum prominence measured in our series was 6 mm.

During the mean follow-up period of 7.4 years (2 months–20 years) there was no clinical or posttherapeutical development that made a correction in the diagnosis necessary. No patient of this relative young population was lost for follow-up.

Discussion

The clinical diagnosis of CCH is not always so straightforward as sometimes suggested in the literature [4, 10]. The very limited experience of the individual ophthalmologist and the chronic secondary changes created by the benign tumor in the surrounding tissue are responsible for the diagnostic problems.

Ancillary diagnostic tests are, either not uniform in their results (visual fields, fluorescein angiography), a burden to the patient (P32), or only employed in a limited number of patients [ICG, CT, MRI]. Although fluorescein angiography is helpful in identifying CCH (a coarse vascular pattern of fluorescence in the (pre-)arterial phase and a diffuse multiluculated pattern in the outer polycystic degenerated retina in the later stages) changes in the overlying RPE may interfere in such a way with its characteristic angiographic pattern that they are not always pathognomic for the lesion [14–16]. Indocyanine green, unlike fluorescein does hardly leak from the choroidal vessels, giving rise to a sometimes even very characteristic angiographic pattern in CCH [12, 26]. Despite better results the experience with ICG is very limited. The same holds true for CT and MRI. In the differentiation of intraocular tumors in adults the results CT are poor despite high resolution thin slice techniques, enlargement of images and contrast enhancement. The small differences in density do not allow tissue-typing [17]. The non-ionizing MRI technique including gadolinium contrast enhancement is useful in tumor detection and tumor extension but of very limited value in tumor differentiation, especially when the paramagnetic effect of melanin is absent. The data acquisition time of this expensive examination is relatively long while the evaluation of more cases is necessary to determine if the preliminary observations until now are consistent. With CT and MRI only masses with a prominence > 3 mm are clearly visible. The radioactive phosphorus uptake test (P32), gives significantly lower uptake counts in CCH than in comparably sized melanomas, but it is an invasive test, which requires a peritomy and special equipment. It is used
less frequently today because of the better recognition of intraocular lesions with non-invasive techniques and of the more definite diagnostic techniques such as fine needle biopsy in the questionable cases.

Ultrasonography enables the detection of mass-lesions with a prominence of > 0.3 mm. When the prominence is > 1.5 mm CCH can be differentiated from other tumors because the ultrasonographic characteristics are uniform and quite different from other intraocular mass-lesions such as choroidal melanoma (Fig. 3) [shape: dome or mushroom, reflectivity: low to medium, vascularity and choroidal excavation frequently positive]. In choroidal metastasis the B-mode echogram shows mostly a broad based choroidal infiltration of variable prominence and reflectivity level. Only a choroidal naevus can give the same echogram as observed in a slightly elevated CCH. There is a lot of experience with this inexpensive examination technique that can always be near the ophthalmologist’s hands.

In the 40 patients we followed for a long time it appeared reliable.

In conclusion for our series. Only two examinations – ophthalmoscopy and ultrasonography – could ascertain the most probable diagnosis of CCH in all the cases.

References