3.4 Classic Choroidal Neovascularization

Joachim Wachtlin

Epidemiology, Pathophysiology, and Clinical Presentation

- Classic and occult choroidal neovascularization (CNV) have to be distinguished. They can also appear in combined forms; depending on the relative size of the classic part, the condition is described as predominant or minimal classic CNV. Classic CNVs are considerably less frequent than the occult or mixed forms. The common definition in use today is based on the Macular Photocoagulation Study (MPS).

- The diagnosis of a classic CNV requires an angiography. The definition is clinically important, for the choice of therapy. A classic CNV can occur in exudative age-related macular degeneration (AMD), but also secondary to other chorioretinal diseases.

- In addition to actual CNVs, exudative lesions can contain other components, such as fibroed tissue, hemorrhage, pigmentation, or other features that may obscure the boundaries of the CNV. The term "lesion" can be defined as the entirety of the clinically and angiographically detectable changes, including all of the above-mentioned changes. The distinction between classic and occult and the estimation of size relate to the whole lesion (see also sections 3.7 and 3.13).

- A classic CNV is defined as a clearly visible and well-demarcated hyperfluorescence in the early phase, with increasing leakage in the late phase of the angiography.

- Newly formed fibrovascular networks which grow out of the choriocapillaris through Bruch membrane between the retina and the retinal pigment epithelium (RPE) are the basis for this angiographic picture. In angiography, classic CNVs are therefore better defined than occult CNVs, which principally proliferate under the RPE.

- The initial symptoms of classic CNV are metamorphopsia, deterioration in visual acuity, and central visual field defects.

- Ophthalmoscopic signs of CNV are grayish-white subretinal changes together with retinal edema, hard exudations, and subretinal and intraretinal hemorrhage. If the condition is not treated, progression with enlargement of the lesion and subsequent loss of photoreceptors will usually follow.

- The final stage is a subretinal fibrosis or disciform scar.

Fluorescein Angiography

- An early bright, well-demarcated hyperfluorescence that is caused by vascular proliferations between the RPE and the neuroretina is typical for a classic CNV.

- CNVs appear either as a network with clearly definable vessels or as a homogeneous structure. A small hypofluorescent edge is often visible. Evidence of a clearly defined vascular network is not mandatory for the diagnosis of a classic CNV.

- In the late phase increasing hyperfluorescence with clear leakage is visible, caused by the outflow of dye from the permeable vessels of the CNV.

- Leakage means that the intensity of the hyperfluorescence in the late phase increases and extends beyond the borders of the CNV visible in the early phase.

Diagnosis and Treatment

- A purely clinical distinction between classic and occult CNV is not possible, fluorescein angiography is essential for confirmation and differentiation.

- Angiography is not usually required if exudative changes cannot be clinically detected, if there is extensive hemorrhage, or if a disciform scar can be seen in the advanced form of the disease. In these situations no treatment options exist and the diagnosis can be established without angiography.

- Further categorization of CNVs is based on their position in relation to the center of the fovea (see section 3.7). Treatment recommendations are based on these classifications.

- In subfoveally predominant classic CNVs, photodynamic therapy is the treatment of choice, based on the findings of the Treatment of Age-Related Macular Degeneration with Photodynamic Therapy (TAP) study and Verteporfin in Photodynamic Therapy (VIP) study. In the Macular Photocoagulation Study, the use of argon laser coagulation is reserved for juxtafoveal and extrafoveal classic CNVs (see section 3.12).

- New treatment procedures include intravitreal or sub-scleral application of agents inhibiting angiogenesis and a combination of photodynamic therapy with either intravitreal application of triamcinolone or angiogenesis-inhibiting agents. The indications for combination treatment and the optimal choice of drug for specific CNV locations or sizes are currently being investigated in detail.

- Optical coherence tomography appears to helpful for treatment control and planning of repeat treatment.

References


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Fig. 3.4a–i Classic choroidal neovascularization

a Schematic representation of the position of a classic choroidal neovascularization between the neuroretina and the retinal pigment epithelium. R, retina; RPE, retinal pigment epithelium; C, choroid.

b Color photograph. The choroidal neovascularization is difficult to identify, and has the appearance of a light gray coloring in the foveal area. Some blood and several drusen are also visible.

c Early arteriovenous phase. The vessel network of the classic choroidal neovascularization appears as an early, well-defined and demarcated hyperfluorescence. It also features several small areas of hypofluorescence resulting from blockage of fluorescence from hemorrhage and hard drusen.

d Late arteriovenous phase. There is a marked increase in hyperfluorescence in the area of the choroidal neovascularization.

e Late phase. Leakage from the choroidal neovascularization. Fluorescein is flowing out of the vessels, and the area of increasing leakage extends beyond the vessel network that was visible in the early phase.

f Early arteriovenous phase. Another example of a classic choroidal neovascularization, consisting of a well-demarcated, homogeneous early hyperfluorescence. However, the vessel loops are not identifiable.

g Late phase. Clear leakage from the choroidal neovascularization is seen.

h Color photograph. The late stage of untreated exudative age-related macular degeneration. An extended disciform scar has formed, with subretinal fibrosis and hemorrhage.

i Late phase. Leakage and staining in the area of the extended fibrosis. This angiogram is not necessary, as the diagnosis could have been established clinically and angiography has no implications for treatment.