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Understanding fluorescein angiographic signs of posterior pole choriocapillaritis.

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In recent years the term choriocapillaritis has been introduced to describe those uveal inflammations whose primary localization is the choriocapillaris. Some of those are not really new entities for the Ophthalmologist and they include: 1) Acute Posterior Multifocal Placoid Pigment Epitheliopathy; 2) Serpiginous Choroidopathy; 3) Harada's Disease; 4) Sympathetic Ophthalmia. Though little is known about their ethiopathogenesis, the diagnostic approach does not usually present particular problems.

Beside these entities, different uveal inflammations with primary localization in the choriocapillaris, but without the clinical aspects of the aforementioned diseases, have recently been described by many Authors [1,6]. The most important among these are the following: 1) Birdshot retinochoroidopathy (BSRC); 2) Multifocal choroiditis and panuueitis (MCP); 3) Punctate inner choroidopathy (PIC); 4) Diffuse unilateral subacute neuroretinitis (DUSN); (Table 1).

The BSRC, also called vitiliginous retinochoroidopathy, is usually seen in middle aged individuals. Females are more frequently interested than males. The fundus lesions consist of multiple yellow-white spots, scattered in the postequatorial choroid. Retinal vasculitis, vitreous reaction, macular edema and papilledema can be parts of the syndrome. In a high percentage these subjects are positive to the HLA-A29 antigen. BSRC is a chronic, slowly progressive disease where remissions are alternated with exacerbations. Usally, symptoms do not improve with steroid therapy. The pathogenesis seems to be autoimmune.

TABLE 1

Clinical features of «atypical» choriocapillaritis

	Birdshot retinochoroidopathy	Multifocal choroiditis and Panuveitis	Punctate inner choroidopathy	Diffuse unilateral subacute neuroretinitis
Median age	53	33	27	14
Male/Female ratio	1:2	1:3	females	3:2
Site of the lesions	bilateral, postequatorial	bilateral, ubiquitary	bilateral, posterior pole	rarely bilateral, posterior pole
Size (μm)	50 to 350	50 to 300	100 to 300	100 to 300
Vitreous reaction	+ + +	+ +	+ /—	+ + +
Vasculitis	+ + +	+ +	—	+ + +
Papillitis	+	+	—	+ + +
Subretinal neov.	rare	30%	40%	10%
Other signs	HLA-A29	AC reaction	moderate myopia	presence of a nematode in the outer retina

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The MCP was originally described by Nozik and Dorsch [2] and recently reviewed by Dreyer and Gass [3]. It is characterized by multiple gray-white spots scattered in the posterior pole and/or in the periphery. Vitreous cells and anterior chamber reaction are also frequently seen. The disease can be observed in young, otherwise healthy, females.

The PIC was initially described by Watzke and Coll. [4] who observed some small yellow-white spots in the choriocapillaris of the posterior pole in young females. It is usually associated with a moderate myopia. The ethiology remains unknown, so far. Morgan and Schatz [5] introduced a cumulative term of Recurrent Multifocal Choroiditis believing that PIC and MPC could be different clinical pictures of the same disease.

The DUSN is a syndrome characterized by vitritis, papillitis, retinal vasculitis, along with small white lesions at the posterior choroid. The pathogenesis should be conducted to the toxic effects produced by the presence of a nematode in the outer retina [6].

The aim of this study was to identify the angiographic patterns of these relatively new clinical entities, by retrospectively reviewing cases of choriocapillaritis observed in our Institute between 1983 and 1987.

The fluorescein angiographic changes can show variations according to the different stages of the disease (Table 2). In the acute stage some typical signs of active choriocapillaritis are evident with early blocked fluorescence associated with late staining at the level of the lesions. Sometimes, a sensory retina detachment can be visible. In the remission stage evidence should be given to the search for subretinal newvessels.

TABLE 2

Angiographic signs of choriocapillaritis

acute stage:
- — early blocked fluorescence at the level of the lesions
- — late staining of the lesions
- — subretinal pooling of the dye (sensory retina detach.)
- — other signs (related to vasculitis, papillitis, ...)

healing stage:
- — «window deffects» at the level of the lesions
- — subretinal pooling of the dye (subretinal newvessels)
- — other signs (related to cystoid macula edema, optic atrophy, ...)

The early hypofluorescence at the level of an active lesion is an angiographic sign commonly seen also in other typical choriocapillaritis, such as placoid pigment epitheliopathy or serpiginous choroidopathy. This hypofluorescence is due both to a delayed filling of the affected choriocapillaris and to a masking effect of the deep choroid. The lesions appear as dark spots scattered in the fundus, the number being greater than expected by ophthalmoscopy (Fig. 1). This sign is common in the MCP,

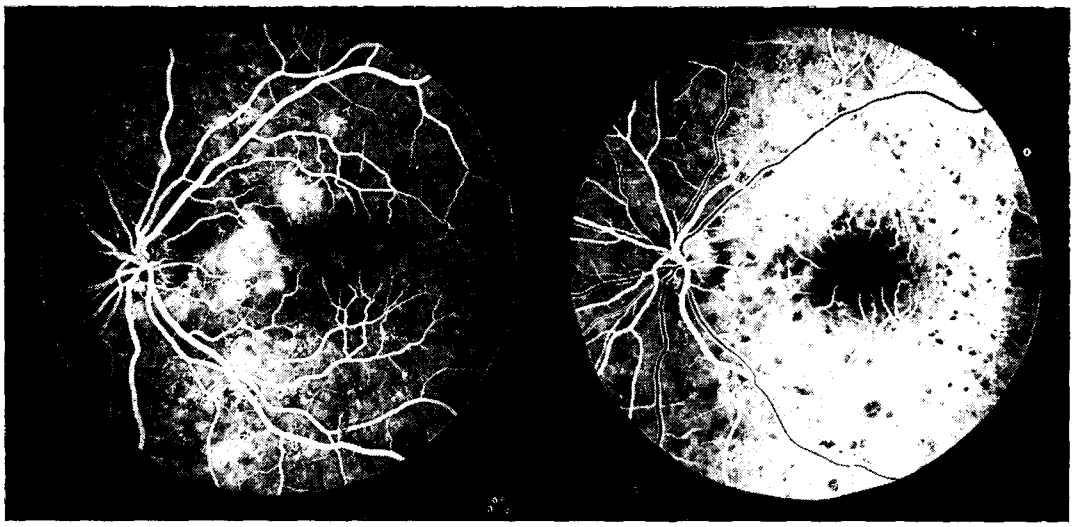


FIG. 1 — Recurrent multifocal choroiditis in the acute stage. Early phases (left) show multiple hypofluorescent spots. Some of these retain the dye in the late phases (right).

PIC and DUSN. In BSRC the lesions can be angiographically completely silent in these phases. In the following phases the dye begins to enter the lobules which progressively gain fluorescence. In the late phases the dye is abnormally retained in the affected choriocapillaris. Another important sign is related to the role of the retinal pigment epithelium (RPE) to maintain an intact outer blood retinal barrier. Occasionally, the RPE overlying the lesions can show an impairment of this function. Consequently, the dye from the choroid can pass through the RPE then leaking into the retina. This sign is frequently seen in the PIC, less frequently in the BSRC. Sometimes the leakage from the RPE can be so important that it creates a serous detachment of the sensory retina. After the venous phase, the dye begins to spread through the RPE and, by means of some pinpoint leaks, it abundantly pools under the retina (Fig. 2).

Other angiographic signs, which however should be considered not directly related to the lesion of the choriocapillaris, are secondary to vasculitis, macular

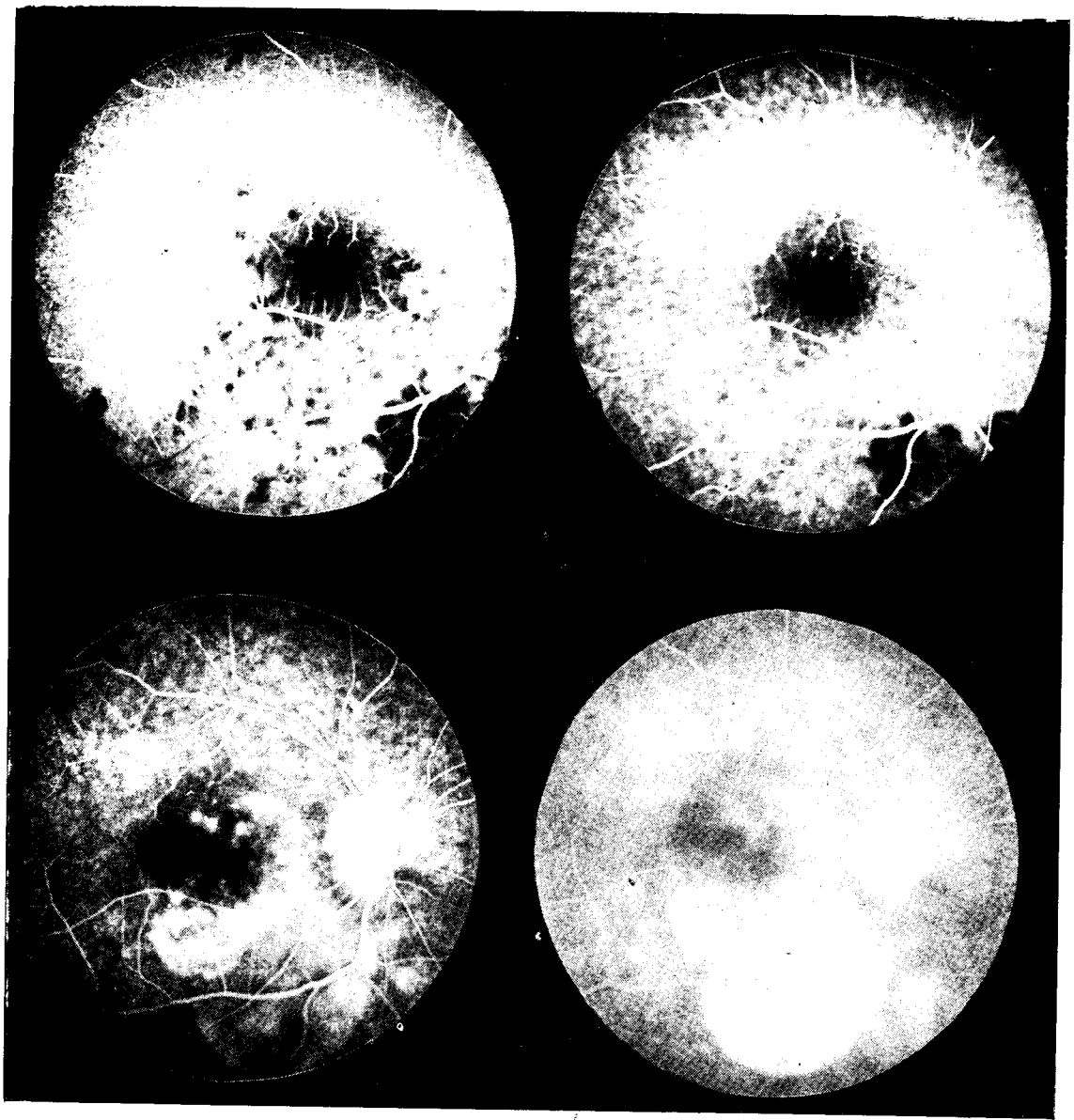


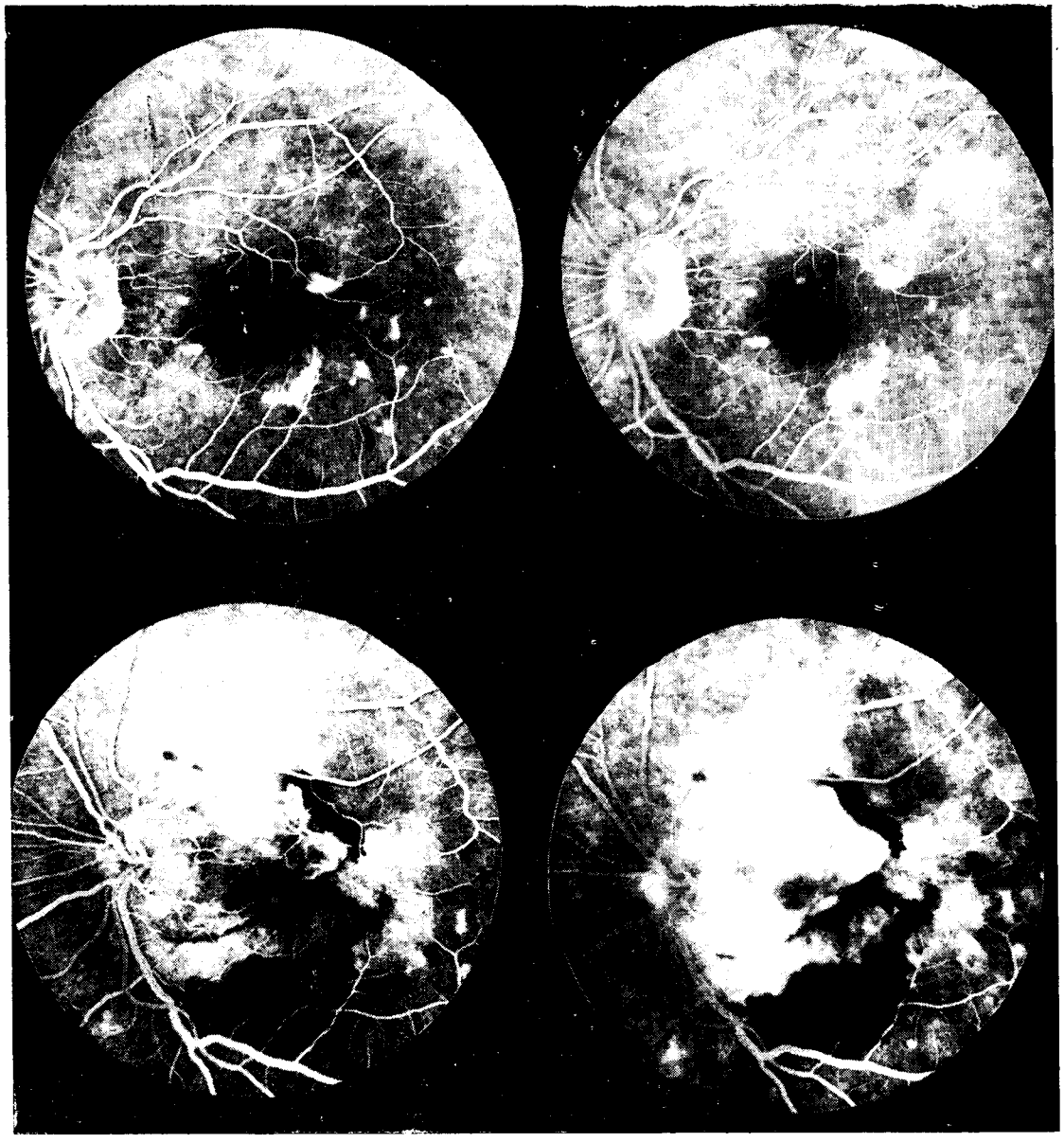
FIG. 2 — Punctate inner choroidopathy. Early phases show multiple hypofluorescent spots (top left). Some pin-point leaks become then visible (top right). A sensory retina detachment is evident (bottom left). The dye abundantly pools under the retina (bottom right).

edema, papillitis. Many of these can be seen in the BSRC and in the DUSN.

With the healing, the acute angiographic signs slowly disappear. More or less evident depigmentation occurs, which is responsible of the early hyperfluorescence of the old lesions, i.e. the classical «window deffect». Though the most part of the choriocapillary lesions shows this evolution, it is not uncommon that a lesion can disappear without any residual ophthalmoscopic or angiographic sign.

The most important complication of the healing process is the development of subretinal neovascularization, which is particularly frequent in the MCP and in the PIC. When subretinal newvessels appear, hemorrhagic detachment of the sensory retina rapidly occurs. Angiography can early detect subretinal newvessels, thus allowing a possible therapeutic approach (Fig. 3).

In conclusion fluorescein angiography gives unique informations on the structural changes which take place in the affected choriocapillaris and serves as a valuable instrument in monitoring the course of these types of choriocapillaritis.



IG. 3 -- Top pictures show a healed choriocapillaritis. After few weeks a large subretinal neovascularization is evident (bottom)

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