Coats' Disease

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Coats' disease is to be defined as exudative retinitis in childhood associated with congenital retinal telangiectasis. Cases of exudative retinitis in adults may only be labelled with Coats' name when the disorder has started in childhood as Coats' disease. The designation 'Coats lesion' for cases of exudative retinopathy in adults should be discontinued, whatever their aetiology.

The primary lesion is a congenital retinal telangiectasis. Arteriocapillary shunts cause abnormally high intravascular pressure which give rise to intraretinal accumulation of fluid, formation of cysts and degeneration of retinal elements resulting in lipid deposits. Leakage of exudate into the subretinal space provokes proliferation of pigment epithelium cells. These cells can dedifferentiate into 'ghost' cells and into fibre-producing cells. The latter, together with fibroblasts which have migrated from the retina, lay down subretinal fibrous tissue, which causes retinal folds and retinal detachment.

Discussion

Ten Thye: Coats described a disease of which the essential feature is a massive exudate behind the retina, and the ophthalmoscopic picture is a yellow or white area in the retina. This may be associated with anomalies of the retinal blood vessels or with a massive retinal detachment, and Coats inclines towards the idea that vascular anomalies are the primary cause, but according to him these may also be absent. By Coats' disease we understand a massive exudate behind the retina. As far as the aetiology is concerned, this may cover a variety of conditions.

Hagedoorn: If I have understood you correctly, an anomaly of the vessel wall is not assumed, but an abnormal communication between artery and veins, a sort of micro-Hippel-Lindau disease?

Manschot confirms this.