25 Inflammatory Vascular Disease

25.1 Eales’ Disease

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Core Messages
- Eales’ disease is essentially a clinical entity, presenting as a recurrent vitreous hemorrhage in young adult males
- The clinical picture is one of an idiopathic retinal vasculitis (periphlebitis) and its sequelae
- Bilateral involvement in an economically productive age group makes it an important ophthalmic health care issue in South Asia
- Its exact etiopathology has evaded explanation, though the role of Mycobacterium tuberculosis, oxidative stress and immunological mechanisms have been studied
- Visual results with retinal laser treatment in patients with neovascularization and of vitrectomy in eyes having vitreous hemorrhage are good, if timely treatment is available
- Periodic follow-up is important given the progressive nature of the disease

25.1.1 History

Henry Eales, a British ophthalmologist, described this condition almost 125 years ago [14, 15]. Eales’ original description was of recurring retinal and vitreous hemorrhages along with epistaxis, headaches, variation in peripheral circulation, dyspepsia, and chronic constipation in young men. He felt it was a vasomotor neurosis, wherein constriction of the alimentary vessels resulted in compensatory dilatation of the vessels in the head, leading to bleeding. Though Eales was honored with the eponym for this disease, Wadsworth was the first to describe the presence of retinal inflammation, 5 years later [37]. Duke Elder considered Eales’ disease to be a clinical manifestation of many diseases.

25.1.2 Epidemiology

One in 200 – 250 general ophthalmic patients in India are affected, ten times more than in North America or Europe, where today it is a diagnosis of exclusion. The male preponderance is marked – almost 90% [10]. Bilateral affection has been reported in 50 – 90% of cases, depending on the study [26]. Most present in the 3rd decade of life (15 – 45 years). There is a definite predilection for the poor socioeconomic class and most patients come from rural communities. While the disease was reported globally at the beginning of the last century, improved sanitary conditions and standard of living have seen a gradual decrease in Europe and North America. Interestingly, Murphy et al. from the USA did not note a male preponderance in their series of 55 cases in America [27]. Today most cases are reported from Asia.

25.1.3 Clinical Features

Eales’ disease is an idiopathic, usually peripheral, invariably bilateral retinal vasculitis resulting in peripheral non-perfusion and neovascularization in a young otherwise healthy male population.

The presenting symptoms in most patients are the presence of floaters and painless diminished vision of varying degrees. On most occasions patients have had many such episodes. These are usually self-limiting in the initial phase, as reabsorption takes place when the bleed is small. Visual loss usually takes place due to recurrent vitreous hemorrhage and rarely due to vascular ischemia.

Clinical signs are mainly restricted to the posterior segment; rarely a patient may show the presence of minimal anterior chamber reaction. The presence of anterior chamber flare or cells is usually the first indicator of the development of rubeosis iridis, as the presence of new vessels is difficult to visualize in dark brown irides.

Clinically, the various features of Eales’ disease can be divided into signs of inflammation, signs of ischemia, and signs of neovascularization and its sequelae. In the normal course, they occur in this order; however, features from different stages may be
present in the same patient. Given the bilateral tendency of this condition, careful examination of the fellow eye almost always shows peripheral sheathing.

It is important to emphasize that the diagnosis of Eales’ disease is essentially clinical: no immunological, pathological or biochemical tests are available to make a diagnosis.

### 25.1.3.1 Signs of Inflammation

Peripheral periphlebitis or vasculitis is the hallmark of this disease. Perivascular exudates result in sheathing of the involved segment of the blood vessels. This can be localized, which can be referred to as cuffing (Fig. 25.1.1b) or involving a large segment of the vessels, called sleeving (Fig. 25.1.1a, c). Vessels with active vasculitis take up fluorescein (FA) dye, showing typical staining. It is important to note the extent of involvement of a vessel on FA far exceeds the ophthalmoscopic picture (Fig. 25.1.1d). Sometimes the vasculitis may involve one of the larger venous branches, resulting in a secondary branch retinal vein occlusion (Fig. 25.1.2a, c). Such secondary venous occlusions are characterized by sheathing of the involved vessels. Associated superficial retinal hemorrhages and edema occurring in this condition are often seen to cross the horizontal midline raphae temporally, which is not seen normally in branch retinal vein occlusion (BRVO) (Fig. 25.1.2b, c) [27]. Though not common, some eyes do show the presence of patches of fresh or old chorioretinitis, against the backdrop of retinal vasculitis (Fig. 25.1.3a–c).

### 25.1.3.2 Signs of Ischemia

Retinal ischemia due to vessel closure manifests in the same way as in other retinal vascular diseases [16, 35]. Superficial retinal hemorrhages in the nerve fiber layer are often seen in the area of sheathed vessels (Fig. 25.1.4). Interestingly, dot blot hemorrhages are rarely seen. In response to ischemia, venous changes such as beading and reduplication are seen (Fig.

![Fig. 25.1.1. a Multiple areas of active periphlebitis in the periphery, some associated with retinal hemorrhages. b Multiple cuffs of periphlebitis. c Sleeving of a large segment of vessels, laser marks seen. d FA of vein with active periphlebitis showing staining](image-url)