Ocular involvement in Whipple disease: three challenging cases

Testi Ilaria, Tognon Maria Sofia
Department of Neuroscience, Ophthalmology Unit, University of Padova, Padova, Italy
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Background:
Whipple disease (WD) is a rare but underrated, chronic, multisystemic condition, caused by the gram positive *bacterium Tropheryma Whipplei*. The most common symptoms include gastrointestinal disorders and intermittent polyarthralgias. Ocular manifestations may result from direct ocular involvement, primarily in term of uveitis.
We report three challenging cases of WD, with and without gastrointestinal symptoms, characterized by two different ocular involvements: surface and uveitis.

Cases:

Case 1:
The patient, native of Senegal, HIV+, CD4 400 cell/mmc, suffered from a chronic conjunctitivitis with a fibrovascular pannus involving corneal periphery and diffuse pigmented limbic nodules in both eyes. There was not intraocular inflammation. Patient complained of diarrhea, abdominal pain and weigh loss. The diagnosis of ocular Whipple disease was confirmed by a positive PCR in conjunctival scraping, after positive PAS staining of bowel biopsies.

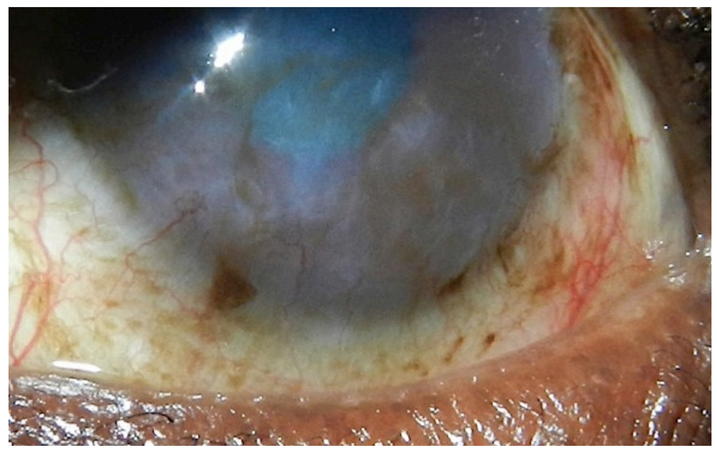

Figure case 1:
OS corneal fibrovascular pannus

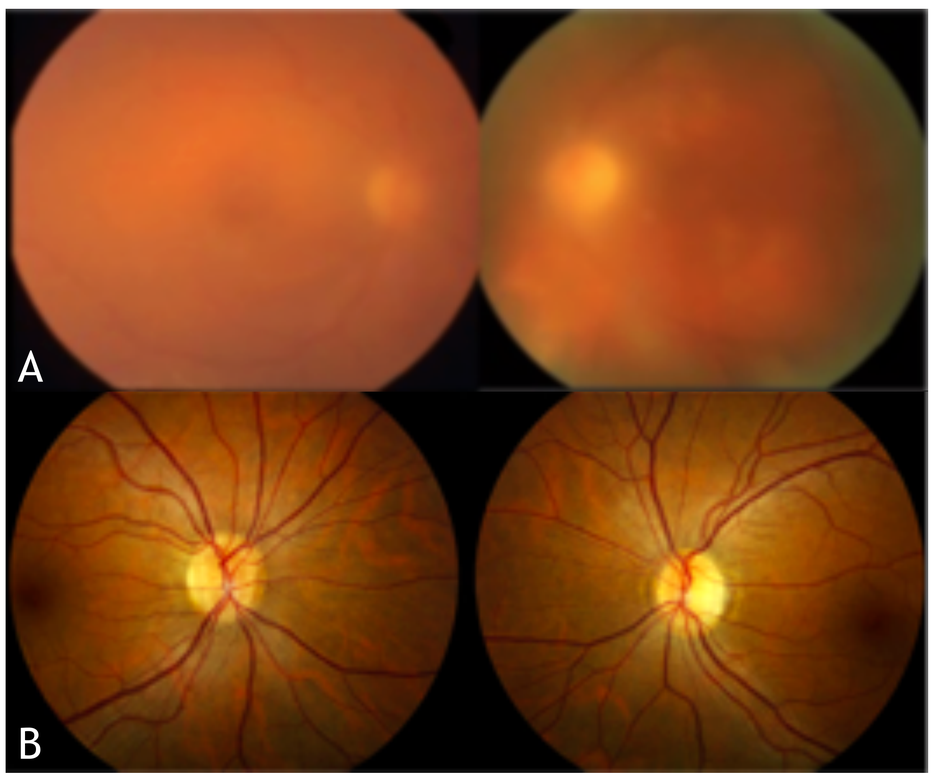

Figure case 2:
A, OU vitritis
B, OU resolution of vitritis after antibiotics treatment - only a temporal pallor of the optic disc in the left eye remained

Case 2:
The patient presented with abdominal pain, mesenteric lymphadenopathy and posterior uveitis, with intense bilateral vitritis. On suspicion of lymphoma associated with ocular masquerade syndrome, patient underwent laparoscopic biopsy, but the histopathological features excluded malignancy. PCR analysis of the lymphnodal sample revealed *Tropheryma Whipplei* DNA, leading to the diagnosis of WD with a likely ocular involvement. The patient was quickly treated with systemic antibiotics with significant ocular improvement, that confirmed the diagnosis of ocular WD.

Case 3:
The patient suffered from gait unbalance, dysarthria and arthralgias associated with a ten-years history of bilateral panuveitis (endothelial keratic precipitates, anterior chamber cells and severe vitritis, partially organized). Reporting also sporadic oral ulcers, a diagnosis of neuro-Behcet's was first made. Considering the worsening of ocular inflammation and a progressive optic atrophy in the right eye after intravitreal and systemic steroid therapy, a diagnostic vitrectomy was performed and a positive PCR for *T.Whipplei* DNA was found in vitreo, leading to the diagnosis of neuro-Whipple disease, complicated with optic neuropathy, in the absence of any gastrointestinal symptoms.

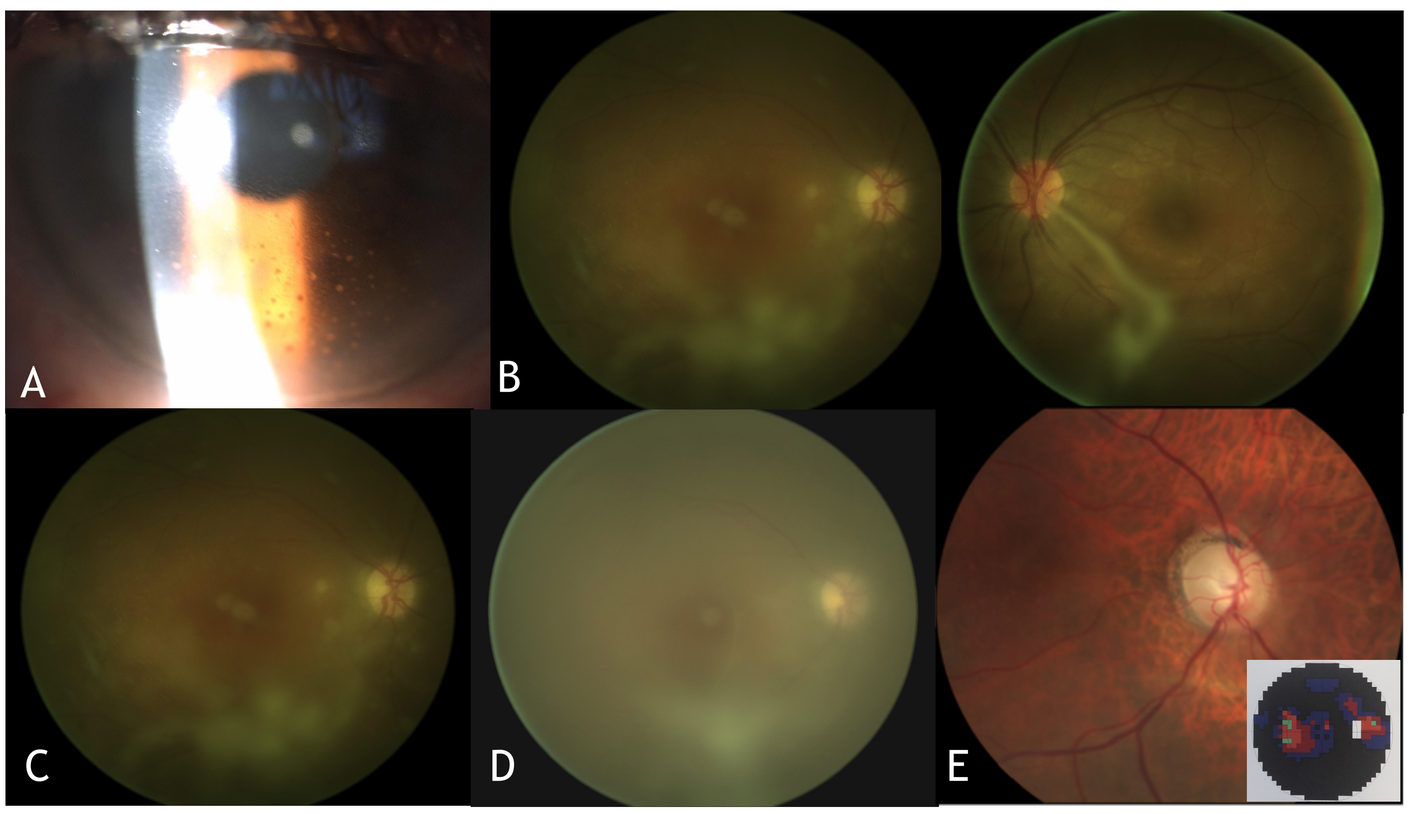

Figure case 3:
A, OD endothelial keratic precipitates and anterior chamber cells
B, OU severe vitritis, partially organized
C and **D,** OD vitritis pre and post intravitreal steroid therapy
E, OD outcome: resolution of vitritis - optic atrophy for delayed treatment with a completely lost visual field

Comments and Conclusions:
Ocular inflammation is a possible feature of WD, usually occurring late in the course of the disease; most of the patients with ocular involvement have also an history of gastrointestinal symptoms. Nevertheless, WD is underrated and it should be ruled out in atypical chronic uveitis mimicking a Behcet's disease or a lymphoma, even in the absence of any gastrointestinal symptoms.
Uveitis is typical in WD, but also an isolated ocular surface involvement is possible. Cytologic and molecular analyses of ocular samples can lead to the diagnosis. An early detection and a ready treatment of the disease are crucial to avoid a poor visual prognosis, as in case 3.