Aim: To present an unusual case of limited form of Wegener's granulomatosis showing only bilateral ocular involvement without any other systemic manifestation.

Patient and methods: A 45-year-old female with right-sided ocular and orbital pain was diagnosed as right-eye scleritis. Treatment with steroids and anti-inflammatory drugs achieved remission of the disease. She had no history of any systemic disease.

Systemic examination did not reveal any abnormality. Abdominal ultrasonography was all within normal limits. Bilateral lung fields were clear on chest X-ray and there was no hilar lymphadenopathy. CT and MRI scans of the orbit were normal.

Three months after the first attack she developed bilateral scleritis that led to bilateral panuveitis and eventually to right eye exudative retinal detachment.

Results: Histologic and serologic findings were key in the final diagnosis. Five months after the first signs of illness she was diagnosed with Wegener's granulomatosis. Pars plana vitrectomy following right-eye retinal detachment was done. One year after the surgery she developed bilateral cataracts and systemic vasculitis involving symga, liver and cerebrum. After bilateral cataract surgery her vision improved. In last six months she had no local recurrence.

Conclusion: Wegener’s granulomatosis is a necrotizing vasculitis, which classically affects upper airways, lungs and kidneys. Early diagnosis and treatment is crucial to increase the survival rate for the aggressive and early medical treatment results in good outcome. Ophthalmic affection prior to systemic manifestations is much less frequent and could delay final diagnosis leading to postponed treatment and possible adverse outcome.