A twenty-one-year-old generally healthy gentleman presented to the ophthalmology clinic with progressive reduction of vision over 3 years and family history of Best’s Disease. Best’s disease is a rare autosomal dominant congenital vitelliform macular dystrophy. Patients usually present with deterioration of central vision in the second decade of life and gradually worsening over the years. The disease is untreatable and low visual aids are used. Genetic and clinical counseling is accessible to affected individuals along with their asymptomatic relatives.

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Best’s disease is a rare autosomal dominant congenital vitelliform macular dystrophy\(^1\). This disease is caused by dysfunction of Bestrophin protein resulting in abnormal fluid and ion exchange in the retinal pigment epithelium (RPE) especially at the macula\(^2\)\(^\text{-}^4\). Hence, it leads to RPE swelling and subretinal accumulation of lipofuscin\(^5\).

It is a progressive macular disease which is usually detected in the second or third decade of life when central vision is affected (reduction of visual acuity, colour vision impairment, and metamorphopsia) but night vision is unaffected. In the fifth or sixth decade, the vision is severely impaired\(^6\). There is no specific treatment for the disease; the main aim is to enhance visual rehabilitation with low visual aids. In certain stages, such as secondary choroidal neovascularization or hemorrhage, it can be managed with direct laser treatment, intravitreal injection of bevacizumab and photodynamic therapy to prevent unnecessary complications\(^6\)\(^\text{-}^\text{10}\). This disease is avoidable by genetic counseling.

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The aim of this report is to present this rare disease which was not reported before in the Kingdom of Bahrain.