Myelinated nerve fiber is a developmental abnormality and occurs in 1% frequency. Unilateral large peripapillary myelinated nerve fiber may be associated with high myopia, anisometropia and amblyopia. Our aim is to evaluate an incidental case with unilateral high myopia and amblyopia. Seven-year-old male patient was referred to our clinic with suspicion retinoblastoma and lack of vision in the right eye. During fundus examination large myelinated nerve fibers were found around the right optic disc. Left fundus was normal. Eye movements were considered as normal. Myelinated fibers can lead to haze and visual deprivation in the retinal image. Such deficiency may contribute to myopia to the cause axial elongation during the critical stage of ocular development. On the other hand, axial elongation may cause tendency to nerve fiber myelination.