Erdheim-Chester disease is a rare, systemic histiocytosis that involves multiple organ systems. A 39-year-old woman presented with diffuse skeletal pain, headaches, and bilateral optic disc swelling. A computerized tomography revealed bilateral enhancement of the optic nerves and lacrimal glands, with extensive soft tissue infiltration around the optic nerves. A lumbar puncture revealed high pressure with normal content. A biopsy from the tibia and lacrimal gland confirmed the diagnosis of Erdheim-Chester disease (ECD), a rare type of multi-organ histiocytosis. Treatment with oral acetazolamide, prednisone, and immunosuppressive drugs resulted in improvement in systemic symptoms and optic disc swelling. Two years later, while still on immunosuppressive drugs and with no ocular symptoms, optic nerve swelling was noted on a routine follow-up visit. Retinal examination revealed a new finding of bilateral large yellow-white choroidal lesion occupying the entire area between the disc and the macula with overlying subretinal fluid, the choroidal mass probably represents infiltrative lesions of ECD. Although ECD is known to involve different parts of the orbit and the periocular area, intraocular involvement is extremely rare, and to the best of our knowledge was reported in the literature only once.