Juvenile xanthogranuloma is a self-limited skin disorder of young children that uncommonly affects the eye. Juvenile xanthogranuloma has been described in adults, but reported intraocular involvement is extremely rare. We report a case of juvenile xanthogranuloma diagnosed in a 25-year-old man who was seen with nontraumatic hyphema and iridocyclitis. Diagnosis was made from a biopsy specimen obtained from a suspicious skin lesion. Topical and systemic steroids, radiation therapy, and finally immunosuppression were required to eliminate the iris tumor clinically and resolve the patient's recurrent symptoms.


Juvenile xanthogranuloma (JXG) is a granulomatous inflammatory condition of unknown origin most commonly seen in children. Intraocular involvement with JXG is uncommon, occurring in less than one half of 1% of patients with dermatologic involvement. The majority of patients with ocular lesions are young children typically younger than 1 year. Ocular lesions usually involve the iris but have also been reported in the orbit, optic nerve, retina, and choroid. Iridocyclitis, hyphema, and secondary glaucoma are frequent presenting signs. Intraocular JXG is extremely rare in adults. To our knowledge, only 7 previous cases have been reported in the literature (Table). We report a case of intraocular and dermal JXG in a 25-year-old man who was referred for spontaneous hyphema and uveitis with an iris mass.

REPORT OF A CASE

A healthy white 25-year-old man was referred initially in May 1994 for evaluation of nontraumatic iritis and hyphema. Findings on physical examination revealed a vascular, yellow-brown peripheral iris mass that involved several clock-hours of the inferior and nasal iris (Figure 1). Results from a medical workup, which included computed tomography of the orbits, were unremarkable. Evaluation of the patient's skin revealed several yellow-orange to yellow-brown pigmented nodular lesions on the face and scalp. A biopsy specimen was obtained from the most prominent lesion on the chin (Figure 2). Histopathologic findings were consistent with JXG, showing a characteristic sea of monotonous, small, foamy histiocytes with small nuclei, very little pleomorphism, and Touton giant cells (Figure 3).

The patient’s ocular lesion was treated with topical, systemic, and subconjunctival steroids over 2 months with regression but not resolution of the tumor. Four months later the patient returned with a symptomatic red eye and a small recurrent hyphema. He was not taking any medications during this time, having previously completed a tapering regimen of systemic steroids. The peripheral iris was diffusely thickened and infiltrated with tumor (Figure 4).

The patient was treated with 4 Gy radiation to the left anterior eye in November 1994, an additional 6 Gy in July 1995, and a final third course of 6 Gy in December 1995 because of persistent tumor, hyphema, and inflammation.

The patient was seen in follow-up in February 1996. Visual acuity was 20/60, secondary to axial posterior subcapsular cataract. There was persistent 1+ circulation of anterior segment cells. Peripheral anterior synechiae existed between the 4:30- and 7:30-o’clock position inferiorly and were as-
sociated with deposition of yellow material on the inferior cornea. The iris showed marked stromal atrophy, with several slightly elevated areas of vascular tumor. He was tapered off prednisone over 2.5 months.

The patient returned in June 1996 with recurrent symptoms of intermittent blur and pain. Tumor mass of the superior iris was more diffuse. Circulation of anterior segment cells was 4+. A yellow hypopyon was present. The patient refused systemic steroid therapy. Additional radiation therapy was not considered. Treatment with methotrexate, 15 mg/wk, was started as immunosuppressive therapy to replace systemic steroid therapy.

The patient was again seen in August 1996, and finally in November 1996. He was tolerating the methotrexate therapy without difficulty. He was asymptomatic for inflammation in his left eye. Visual acuity was unchanged at 20/60. Slit-lamp examination showed persistent inferior peripheral anterior synechiae related to old tumor inflammation, with marked atrophy of the iris and no evidence of tumor mass (Figure 5). He continues to take methotrexate, 15 mg/wk.

**COMMENT**

The histopathology of JXG lesions is characterized by a monotonous infiltration of normal appearing histiocytes with occasional appearances of other types of inflammatory cells, the most characteristic of which is the Touton multinucleated giant cell. DeBarge et al10 used immunohistochemical techniques to further characterize the histiocytic component of JXG lesions. S100, a brain-specific protein found in glial tissue and Langerhans cells, is consistently found in histiocytosis X. DeBarge et al did not find this marker in JXG. Juvenile xanthogranuloma varies from the histiocytosis X group of disorders in its clinical behavior and histopathologic differences that include negative staining for S100 antigen and the absence of Birbeck (Langerhans) granules on electron microscopy. Juvenile xanthogranuloma is now generally considered an abnormality of histiocytes belonging to the group of non-X histiocytoses.

In children, low-dose irradiation of 2 to 4 Gy given in fractionated doses has been successful in treating this tumor when it has not responded to steroid treatment.11-13 We could find no report in the literature of an adult treated with radiation or nonsteroidal immunosuppression.
Our patient responded favorably to low-dose methotrexate therapy. We acknowledge that his symptomatic and clinical improvement may have had a causal relationship to treatment received prior to starting therapy with methotrexate, or may have represented the natural course of the disease. Nevertheless, given the protracted nature of his symptoms, tumor persistence, and recurrence of inflammation when the systemic steroid regimen was stopped, we believe methotrexate was beneficial in mitigating his signs and symptoms. Low-dose methotrexate therapy may be a viable treatment option in adult patients with JXG who manifest intractable symptoms and persistence of tumor with conventional therapy.

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REFERENCES


Notes From Our Ophthalmic Heritage

A look at the past . . .

The egg in ophthalmology: The film or inner membrane of the hen’s egg has been employed in operative ophthalmic surgery. It has been used to prevent adhesions following lime burns and other injuries of the conjunctival sac. A fatty oil is also obtained by expression from the yolks of fresh eggs. Its chief ophthalmic use is, abroad, as a popular remedy for the removal of corneal opacities.