HKJO Quiz

Answer

Bilateral macular scar due to congenital toxoplasmosis, with the fovea being spared on the left side.

(Question on page 37)

Discussion

Human toxoplasmosis is caused by the protozoa *Toxoplasma gondii*. The infection is a zoonosis, and members of the cat family are the definitive host. It may occur as either a congenital or an acquired form.[1] Congenital infection is the result of ingestion of a *Toxoplasma gondii* cyst by a previously unaffected pregnant woman followed by transplacental transmission of the organism to the fetus. Cysts may be found in the tissue of animal meats or as oocysts in cat feces or litter as well as in the soil. Infection in the first trimester of pregnancy is associated with neonatal convulsions, intracerebral calcifications, and retinitis. When the infant is affected in later trimesters, normally only retinitis develops.[2] If a pregnant woman becomes infected with *Toxoplasma gondii*, there is a 40% chance that her infant will be affected.[1] The prevalence of congenital toxoplasmosis has been estimated to range from 1:1000 to 1:8000 live births in the USA and 1:1000 live births in France.[3,4]

Chorioretinal scars are estimated to occur in approximately 80% of congenitally-infected newborns.[2] The chorioretinal scarring is usually heavily pigmented and associated with areas of chorioretinal atrophy. It is bilateral in 85% of cases and frequently involves the macula. The predilection for the posterior pole of the eye may be related to the end-artery anatomy of the fetal macular circulation.[5] Toxoplasmosis acquired after birth rarely results in chorioretinitis.[1]

More severely affected children may have active chorioretinitis with an accompanying vitritis and anterior uveitis. In children with mild infection, the disease may become apparent only later in life when retinochoroidal scarring is detected on ophthalmoscopic examination. This may occur during evaluation for strabismus, at routine school vision check, or during a routine ocular examination.[1]

In patients suffering from toxoplasmic retininochoroiditis, up to 40% will have permanent unilateral visual loss to 20/100 or less. Nearly 90% will be caused by a lesion in the macular region.[5]

The differential diagnosis of congenital toxoplasmosis includes macula coloboma, cytomegalic inclusion disease, herpes simplex chorioretinits, or foci of retinoblastoma. The diagnosis is usually based on the appearance of the characteristic lesion. Serology tests for *Toxoplasma* infection are supportive of the diagnosis but not definitive. The seropositivity in a population increases with the age of the individual. Marked regional differences have been reported in the prevalence of seropositivity among different countries, presumably due to differing dietary and living habits. [1]

Old, healed, congenital ocular toxoplasmosis can sometimes recur in otherwise healthy individuals, usually between the ages 10 and 35 years when the cysts rupture and release hundreds of tachyzoites into normal retinal cells. Secondary changes such as vitritis and anterior uveitis may occur as a result of a hypersensitivity response.

In an immunocompetent patient, not all active lesions require treatment because small peripheral foci are frequently self-limiting and innocuous. Treatment is indicated for patients in whom the lesion threatens the macula, papillomacular bundle, or optic nerve head, or when the vitritis is severe. Treatment is also indicated for immunocompromised patients.[6] The classic treatment regimen for ocular toxoplasmosis consists of pyramethamine, sulfadiazine, and corticosteroids. Clindamycin is also used by some centers. While all these treatments have demonstrated activity in animal models and efficacy has been reported in uncontrolled studies, there are no randomized, controlled studies to demonstrate the superiority of one regimen over the others. Surgical techniques, including photocoagulation, cryotherapy, and vitrectomy, have also been shown useful in selected cases.[1]

References

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