The Eye-Q Newsletter

The Eye in Systemic Disease

Introduction

Although it is easy to think of the eye and its various disorders in isolation and as distinct from the workings of the rest of the body, in truth, many systemic diseases can reflect themselves in visual symptoms and abnormalities on ophthalmic examination. From hypertension and diabetes to multiple sclerosis and medication toxicity, the eye can truly be a window through which we can see these conditions and their effects on our bodies. In this issue of the Eye-Q Newsletter we will briefly explore some of the more common medical illnesses and their ophthalmic findings.

Hypertension

Acute and chronic elevations in blood pressure can produce observable changes in the retinal vasculature and surrounding tissues. These changes are referred to collectively as hypertensive retinopathy. Patients with hypertensive retinopathy are almost always visually asymptomatic and the eye exam may be the first clue to the systemic disease.

The findings in hypertensive retinopathy all stem from hypertension-induced changes to the retinal microvasculature. The buildup of intimal plaque over time leads to a gradual reduction in the lumen size and eventually focal closure of the vessel. This narrowing of the visible blood column in retinal arterioles has been used to stage the severity of the disease and has prompted descriptive terms such as ‘copper and silver wiring’ for extreme degrees of narrowing. Focal closure gives rise to microinfarcts (cotton wool spots) and superficial hemorrhages (Fig.1). This occurs with diastolic readings of at least 110 mmHg. Since the arterioles and venules share a common adventitial sheath at points where they intersect, the thinner-walled venules can be pinched off to varying degrees by the adjacent thickened arteriolar walls resulting in arterio-venous crossing changes. In this situation the venule is seen to taper down as it intersects with the arteriole and may even disappear entirely. This has implications in the potential development of branch retinal vein occlusions.

More extreme elevations in blood pressure can produce a breakdown of the blood-retinal barrier and allow normally impermeable retinal vessels to leak blood products into adjacent retinal tissue. This results in retinal edema and hard exudates (lipid) on fundus exam. In advanced cases a macular star can develop (a ring of exudates from the disc to the macula). Optic nerve swelling can also be seen (Fig.2). Patients who develop papilledema from hypertension have malignant hypertension with blood pressure typically in the range of 250/150 mmHg. These changes can decrease visual acuity significantly. All of these changes can resolve slowly over time following the reduction of blood pressure.

Conditions related to hypertension such as carotid occlusive disease can also affect ocular perfusion and result in ischemia of both anterior and posterior segments of the eye. Emboli from carotid artery plaques can occlude the central or branch retinal arteries leading to loss of vision.
Diabetes

Diabetic retinopathy was covered extensively in the Eye-Q Newsletter vol 2 No 2 Fall 1997 issue. This remains the leading cause of blindness in North America in patents under the age of 65. Both Type I and Type II diabetics can develop retinopathy. The main features include increased permeability and occlusion of the retinal capillaries. This results in characteristic microaneurysm formation with dot and blot hemorrhages, exudates, cotton wool spots (nerve fiber layer infarcts), macular edema and retinal neovascularization (Fig.3). These neo-proliferative changes can further result in vitreous hemorrhage, scarring, traction retinal detachment, and blindness. The onset and severity of diabetic retinopathy is correlated to the length of time diabetes has been present and to the adequacy of glycemic control.

Regular ophthalmologic follow-up is essential for all diabetics in order to detect funduscopic changes early in their course. Early treatment may prevent vision loss from maculopathy and proliferative changes. Argon laser photoagulation for clinically significant macular edema as well as for proliferative changes has been shown in numerous studies to significantly reduce the rate of vision loss from diabetic retinopathy. Depending on the severity of the retinopathy, surgery such as vitrectomy and retinal detachment repair may be required.

Temporal Arteritis

Temporal or giant cell arteritis is a generalized inflammatory disease affecting large and medium-sized arteries that develops almost exclusively in patients older than 55 and affects women about twice as often as men. Many organ systems may be involved including intracranial arteries. Common associations include arthritis and polymyalgia rheumatica. In addition to the possibility of cerebrovascular accidents temporal arteritis can cause sudden, catastrophic loss of vision due to involvement of the posterior ciliary arteries causing anterior ischemic optic neuropathy (AION). If unrecognized, the fellow eye may also be affected within days to weeks. In addition to AION, temporal arteritis can also be a cause of oculomotor nerve palsies with resultant diplopia, ocular ischemic syndrome, and retinal artery occlusion.

Characteristic clinical symptoms and signs of temporal arteritis include headache, malaise, fever, weight loss, ear pain, jaw claudication, and tenderness over the temporal arteries and scalp. Ocular symptoms include amaurosis fugax, sudden visual loss (partial or complete), and diplopia. AION may be preceded by transient visual obscurations for a variable period of time. With onset of acute vision loss there is evident swelling and often hemorrhage of the optic disc. Visual field testing may show an altitudinal or arcuate defect. Vision may range from 20/20 to NLP (no light perception).

The erythrocyte sedimentation rate is typically markedly elevated. A high index of suspicion is required, however, since the ESR may be normal in 10% of patients and classic symptoms may be absent. Confirmatory diagnosis often requires temporal artery biopsy, but this, too, may be negative. The ESR normally increases with age and is higher in women than in men. The normal value can be approximated by dividing the age by 2 in men or adding 10 to the age before dividing by 2 in women. Anemia also elevates the ESR so the patient’s hematocrit must also be taken into account.

Although no treatment has been shown to be of value in reversing these vision changes once AION has occurred, it is necessary to begin treatment with high dose steroids as soon as the diagnosis is entertained in order to protect the fellow eye. Because of the high risk of involvement in the second eye temporal arteritis should be managed as an ophthalmic emergency.

Optic Neuritis

Optic neuritis affects predominantly women between the ages of 15 and 45. It is characterized by unilateral acute or subacute visual loss, often associated with retrobulbar
pain or pain with eye movement. Generally, there is also diminished brightness sensitivity, decreased color saturation and a central or paracentral scotoma in the affected eye. Other types of field loss can also occur including arcuate and altitudinal patterns. There is almost invariably an afferent pupillary defect acutely. In two thirds of patients the inflammation is retrobulbar and the optic disc appears normal (“the patient sees nothing and the doctor sees nothing”). In the remaining third the inflammatory process is anterior and optic nerve swelling (papillitis) is evident on fundus examination.

Vision typically worsens over the first week and then recovers gradually over several months. Initial acuity ranges from 20/20 to no light perception. Seventy-one percent recover to 20/20 and 95% to 20/40 without treatment within 1 year. Despite this many patients experience residual abnormalities in brightness sense, color vision, pupillary reactivity, contrast sensitivity, optic disc appearance, and visual evoked potentials. Uhthoff’s phenomenon (transient decrease in vision with exercise or elevation of body temperature) occurs in one half of patients after recovery.

The diagnosis is based on clinical findings and patient history with emphasis on recent viral illnesses, neurologic dysfunction, and family history of multiple sclerosis. Neuroimaging can be done in atypical cases (fat suppressed gadolinium-enhanced MRI). This may show focal areas of enhancement in white matter tracts including the optic nerve (Fig.5) indicative of demyelination.

Optic neuritis in children differs from that in adults. It is frequently bilateral and usually postviral and may be associated with meningoencephalitis.

The association of optic neuritis with multiple sclerosis (MS) has become more clearly defined in recent years. Long term studies have shown that over decades the risk of an optic neuritis patient developing MS is greater than 60%. The risk may be greater in women and with occurrence at a younger age. Most MS patients have evidence of optic nerve disease, although only 55% have had a clear history of optic neuritis. MRI abnormalities in the brain are found in about half of patients with clinically isolated optic neuritis. Many do not have neurologic signs or symptoms.

The Optic Neuritis Treatment Trial which ended in 1991 showed that treatment with oral prednisone to be of no benefit in typical cases of optic neuritis and may even predispose patients to further attacks. High dose intravenous methylprednisilone with subsequent oral steroids led to a slightly faster visual recovery but did not change the final visual acuity compared with placebo and did not affect the rate of development of MS over a 3 year period. The decision to use steroids must be made after careful assessment of their potential risks and benefits.