An unusual presentation of lupus retinopathy

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We have described an unusual presentation of lupus retinopathy in the form of macular arterio-arterial and arterio-venular shunts with extensive macular ischemia as a presenting sign.

Key words: Lupus retinopathy, macular shunts, macular ischemia

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Systemic lupus erythematosus (SLE) is an autoimmune disorder characterized by damage of tissues and cells by pathogenic autoantibodies and immune complexes. At onset, SLE may involve only one organ system (additional manifestations may occur later) or may be multisystemic.[1] The ocular manifestations are varied and include involvement of the lids, cornea, secondary Sjogren's syndrome, conjunctival hemorrhages, sclera, retinal vascular disease with retinal hemorrhages; and neuro-ophthalmic lesions.[2,3] The retinal arterial occlusion is an unusual and serious ocular complication which may be sight-threatening.[4] We herein describe an unusual presentation of lupus vasculopathy in the form of macular arterio-arterial and arterio-venular shunts with extensive macular ischemia as a presenting sign.

Case History

A 31-year-old woman presented with the blurring of vision in her both eyes (left more than right) of one-month duration. Her best corrected visual acuity was 20/100 in the right eye and counting fingers at 1 m in the left eye. Anterior segment examination showed early posterior subcapsular cataract and few anterior vitreous cells in both the eyes. Intraocular pressure, Schirmer's test and anterior segment findings were within normal limits. Fundoscopy revealed diffuse arteritis with multiple cotton wool spots (CWS) in both the eyes. Additionally, arterio-arterial and aretrio-venular shunt was noticed across the macular area in the left eye [Fig. 1]. Fluorescein angiography (FA) demonstrated extensive areas of retinal ischemia and later leakage from the arterial tree and the optic disc in both the eyes. Extensive macular ischemia and arterio-arterial and arterio-venular shunts were noticed across the macular area in the left eye [Fig. 2].

Rheumatologist's opinion was sought due to her multisystem complaints and she was found to have polyarthritis, hepatosplenomegaly, oral ulcers and renal involvement. Systemic blood pressure was normal. Subsequently, by laboratory investigations she was found to be positive for rheumatoid arthritis factor, complement-reactive protein, antinuclear antibody and subnormal complement factor-3. Abdominal ultrasonography revealed right iliac fossa adenopathy and thickened bowel loops apart from enlarged spleen and liver. Based on medical and laboratory investigations a diagnosis of SLE was made and she was started on oral steroid therapy (prednisolone) on tapering doses (60 mg/day tapered by 10 mg every week for six weeks)) and posterior

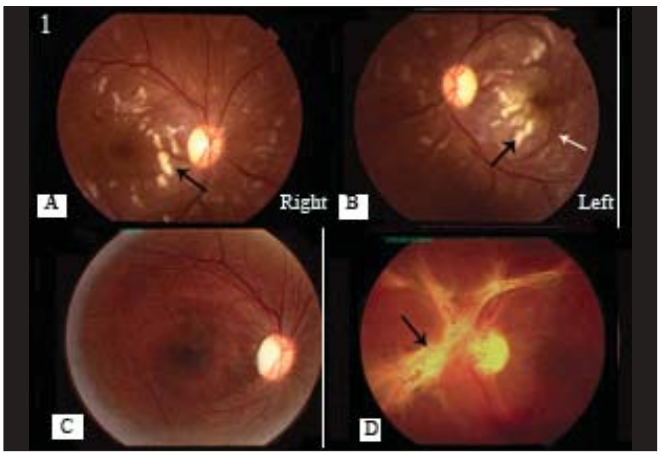

Figure 1 A-D: (A-B) Color photograph of both the eyes at presentation showing multiple cotton wool spots (black arrow) scattered all over posterior pole. Also notice the arterio-arterial and arterio-venous shunts across the macular area (white arrow) in the left eye. (C) Color photograph of the right eye at final visit showing complete disappearance of cotton wool spots. (D) Color photograph of the left eye at final visit showing extensive tractional retinal detachment (black arrow) and severe optic disc pallor

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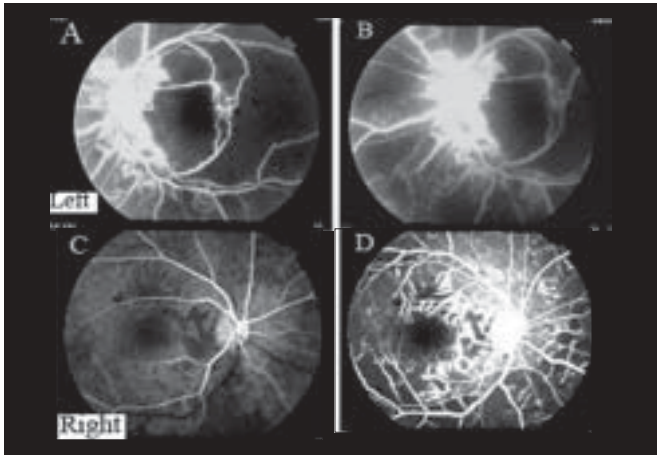

Figure 2 A-D: (A-B) Fluorescein angiograms of the left eye demonstrating extensive leakage and staining of the arterial tree and optic disc. Arterio-arterial and arterio-venous shunts along with extensive macular ischemia are clearly evident. (C-D) Fluorescein angiograms of the right eye showing extensive leakage and staining of the arterial tree sparing the foveal area and staining of the optic disc

subtenon (PST) injection of triamcinolone 20 mg in both the eyes at her first visit on outpatients basis. The PST was repeated after three weeks due to improvement in her retinal findings and laser photocoagulation was done in her left eye twice in her subsequent visits due to presence of multiple retinal neovascularization.

She was then examined periodically both by the retinal surgeon and rheumatologist and at the time of her last visit, 12 weeks after the initial presentation, her visual acuity had improved to 20/30 in the right eye but deteriorated to perception of light in the left eye. Ocular examination showed unchanged anterior segment in both the eyes whereas fundoscopy revealed complete disappearance of CWS and arteritis in the right eye and subtotal tractional retinal detachment up to equator with pale disc in the left eye [Fig. 1]. In view of the severe pale disc, no surgical intervention was advised.

Discussion

In a normotensive patient with SLE, retinopathy occurs due to the disease itself probably related to intrinsic endarteritis, narrowing and eventually occlusion of major arteries and arterioles with presence of multiple CWS.[5] In some patients, retinopathy is manifested by thrombotic occlusions of large arterioles, often associated with evidence of vasculitis and CWS.[6] The case described herein showed both these forms of lupus vasculopathy. In addition, a few other fundus findings were described in the literature which include segmented blood column often filled with white material, retinal hemorrhages and edema and extensive areas of retinal and disc neovascularization and vitreous hemorrhage.[7,8] However, the patient described herein had an unusual clinical finding in the form of arterio-arterial and arterio-venous shunts across the macular area with extensive macular ischemia which has not been reported earlier. Though the right eye of the patient regained near normal vision, the left eye could not be saved in spite of oral and periocular steroid therapy and laser photocoagulation. This report illustrates that ophthalmologists may play an important role in the diagnosis of SLE since ocular manifestations may precede or coexist with serious extraocular manifestations of the disease.

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