Vascular tumors of the retina are rare, benign lesions that may be associated with systemic disease, or may be an isolated finding. Retinal capillary hemangioma occurs in young patients between 10 and 30 years of age. The patient often presents with painless blurred vision and sometimes the mass is discovered on routine examination. Patients with capillary hemangioma may have an underlying von Hippel-Lindaus’ disease, specially in bilateral cases. The patient and family members should be screened for the same.

Characteristic fluorescein angiographic or echographic findings may be useful in securing the diagnosis. Treatment may consist of laser photocoagulation, cryotherapy, radiotherapy, vitrectomy, or a combination of these.

**Case Report**

A 23 year old male presented with painless and gradually progressive diminution of vision in the left eye for the past one month.

There was no history of floaters or flashes of light, no usage of thick spectacles, no fever or weight loss and no associated systemic illnesses. There was no significant past history and no history of similar complaints in any family member.

**Examination**

General and systemic examinations were within normal limits. Ophthalmic examination revealed a best corrected visual acuity of 20/20 in the right eye and 20/200 in the left eye(with -0.75 DC X 173°).

The cornea was clear and there were no anterior chamber or vitreous cells in either eye. The pupil were normal in...
size, shape and reacted briskly to light. Intraocular pressure measured in both eyes with Goldmann application tonometer was 13mm of Hg.

The right fundus examination was unremarkable. Left eye fundus examination showed massive collection of hard exudates at the macula with macular edema on 90 D biomicroscopy (Figure 1). Hard exudates were also present inferonasal to the disc and extending inferonasal up to the equator.

On screening the periphery, two pinkish yellow, well circumscribed, elevated vascular lesions measuring 5mm x 4mm were visualized in the inferotemporal quadrant in the mid-periphery (Figure 2). Similar but smaller lesions were present in the superotemporal quadrant as well (Figure 3). Both the lesions were supplied and drained by separate vessels, which were dilated and tortuous. The two lesions were also connected to each other by vascular channels.

On examining the patient, we made a provisional diagnosis of retinal vascular malformation and went on to further investigate the case.

Investigations

Fundus fluorescein angiography

The early arteriolar phase showed a prominent dilated feeder vessel. Within seconds the lesion was fluorescent and it increased in size as well as intensity (Figure 4,5).

Optical coherence tomography

Macular edema was confirmed on Stratus OCT, with a central macular thickness of 450 microns (Figure 6).

We advised an MRI brain and spine and an ultrasonography of abdomen to rule out hemangiomas at other sites. Urine analysis for vanillylmandelic acid was also recommended to rule out pheochromocytoma. All the above investigations were negative. Besides the above investigations a DNA analysis, for VHL gene of the patient and family members was also advised.
**Treatment**

Injection bevacizumab (Avastin) was given in the left eye in view of the macular edema. This was followed by frequency doubled Nd-YAG (532 nm) laser burns of large size (500 µm), low intensity and long duration (0.2 seconds). The laser burns were placed over the feeder vessels and also around and over the vascular tumor.

Four weeks post laser, the hard exudates had become confluent and clumped together (a sign of resolving macular edema). The OCT also confirmed the above finding. As the lesion still showed signs of vascularity, we decided to repeat the laser treatment (Figure 7).

Six weeks after the last laser sitting, vision of the patient had improved to 20/80. The vascular lesion had regressed significantly and the macular edema had also resolved considerably as viewed in the OCT (Figure 8).

We kept the patient on regular follow up. After ten months the patient’s visual acuity improved to 20/20 in the left eye and the vascular lesion had regressed completely with no macular edema (Figure 9).

The patient is kept on regular six monthly followup and is advised to get yearly systemic check-up to rule out von Hippel-Lindau disease.

**Discussion**

Capillary hemangioma usually affects young patients between 10 and 30 years of age. There is no predisposition for sex or race.

The tumors can be multiple in about one-third of cases.

Bilateral or multiple tumors imply the presence of underlying von Hippel-Lindau disease. There is a 45% risk for developing von Hippel-Lindau with a solitary hemangioma if age is less than 10 years.

Small tumors have a subtle rim of subretinal fluid or spotty adjacent exudation in the retina whereas the exudation with larger tumors can be remote from the tumor in the macula.

The features of von Hippel-Lindau disease include various combinations of retinal capillary hemangioma, cerebellar hemangioblastoma, renal cell carcinoma, pancreatic cysts and tumors and pheochromocytoma. All the above lesions should be ruled out in a patient presenting with capillary hemangioma.

Treatment of the retinal capillary hemangioma depends on the size and location of the tumor, clarity of media, and secondary features of the mass.

Tumors best suited for photocoagulation are those measuring less than 5mm in diameter and without substantial subretinal fluid. The goal is to occlude all feeder arterioles supplying the vascular tumor. Multiple laser sittings may be required in these cases.

Photodynamic therapy has been employed for medium sized retinal capillary hemangiomas that are too large to
treat with laser photocoagulation or those tumors in the juxtapapillary and macular region\(^9\). Retinal cryotherapy may be done in lesions present anterior to the equator with surrounding subretinal exudation.

Capillary hemangioma should be differentiated from vasoproliferative retinal tumor, which is a nodular gliovascular proliferation causing exudation and fibrosis. It is usually seen in the inferotemporal quadrant and anterior to the equatorial. This condition is idiopathic in 75% of cases or secondary to other ocular disease, such as uveitis in remaining 25% cases. These lesions are also supplied by feeder vessels, but unlike capillary hemangioma, the vessels are not dilated and tortuous\(^9\).

**Key Points**

- In young patients with exudates at the posterior pole, the periphery must always be screened.
- Repeated sittings of laser may be required in the treatment of retinal capillary hemangioma.
- Complete systemic examination should be done in all patients with retinal capillary hemangioma to rule out von Hippel-Lindau disease.

**References**