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Case Report

Pheochromocytoma "the great masquerader" presenting as hypertensive retinopathy: A case report

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ABSTRACT

At first glance, the ophthalmologist might seem to be invading a medical territory, not in their domain, when they write of Pheochromocytoma, but this is not true. Pheochromocytoma is an uncommon tumor that develops in chromaffin cells of the paraganglia or adrenal medulla which can give rise to malignant hypertension (systolic >200 mg Hg & diastolic >140). Pheochromocytoma-induced hypertension may present with many physical symptoms and signs, although sometimes the ocular features are the only clues that are present. Clinical suspicion of secondary hypertension should always be considered when fundus examination shows hypertensive retinopathy changes in a younger age group (18 years in our case). The patient, in our case, developed hypertensive retinopathy with macular star development due to Pheochromocytoma. It stresses the significance of a thorough search for a secondary cause of hypertension in young patients.

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1. Introduction

Pheochromocytoma is the term derived from the Greek words phios (dusky), chroma (colour), and cytoma (tumor). Pick created the name pheochromocytoma in 1972.[1] Pheochromocytoma is seen in 85% of adrenal tumors and 15% in extra-adrenal tumors. Paragangliomas are extra-adrenal pheochromocytomas. The tumors grow in specialized cells known as chromaffin cells that are found in the middle of an adrenal gland.[2] These cells secrete hormones, most notably adrenaline (epinephrine) and noradrenaline (norepinephrine). These hormones regulate various body activities, including heart rate, blood pressure (BP), and blood sugar. In reaction to a perceived threat, adrenaline and noradrenaline activate the body's fight-or-flight response. More of these hormones are released by a pheochromocytoma, and they are released even when there is no threat. The symptoms and signs are connected to sympathetic nervous system activation. Headache is a part of the classic triad (probably due to high BP or hypertension), tachycardia/elevated heart rate, and diaphoresis (excessive sweating, particularly at night, also known as hyperhidrosis).

Previously, many physicians referred to Pheochromocytomas as the 10% tumors. Recent developments in our understanding of Pheochromocytoma genetics have pushed physicians to dismiss the 10% rule as an oversimplification.[3] Patients should be checked for secondary causes of hypertension, including Pheochromocytoma, if they have the classic symptoms of the disease, present with severe hypertension or a hypertensive crisis, are younger than 20 or older than 50, have resistant hypertension, have a family history of the symptoms that are hereditarily associated with Pheochromocytoma, or have an incidental adrenal mass on imaging.

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2. Case Report

An 18-year-old girl presented us with the complaint of blurring of vision from the past two months, which was gradual in onset. She also had severe pain on the right side of the abdomen for 3-4 days. Past ocular and medical history were insignificant except for recently diagnosed diabetes mellitus three months back. She was having heat intolerance, panic attacks, and weight loss. She could not concentrate and had postural hypotension, due to which she left her college three months back. She had episodic elevations of BP. At the time of admission, she had a BP of 206/146 mm of Hg and a pulse rate of 124/min. On Ultrasonography, a well-defined hyperechoic mass of size 5.3x3.6 cm was seen in the right supra-renal region. On ocular examination, her visual acuity was 6/60 RE and 6/36 LE, and IOP was 20 and 18 mmHg. On slit lamp examination, the anterior segment was normal in both eyes. The pupillary reaction, both direct and consensual, was normal. On fundus examination, both macular eye fans were present with the blurring of nasal disc margins in RE, and multiple superficial hemorrhages in both eyes were found, as shown in Figure 1.

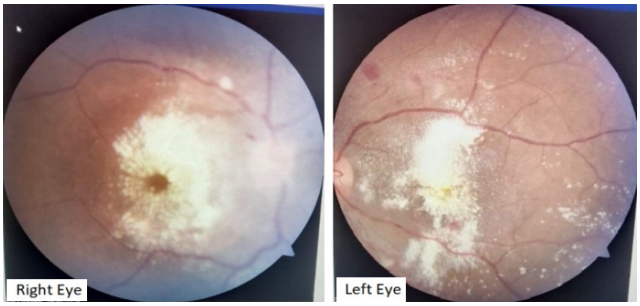

Fig. 1: Multiple superficial hemorrhages

On Optical coherence tomography-multiple hyper reflective shadows at the level of the inner nuclear layer and outer plexiform level were seen. In addition, retinal pigment epithelium alterations were present, with subretinal fluid and foveal contour being disturbed in Figure 2.

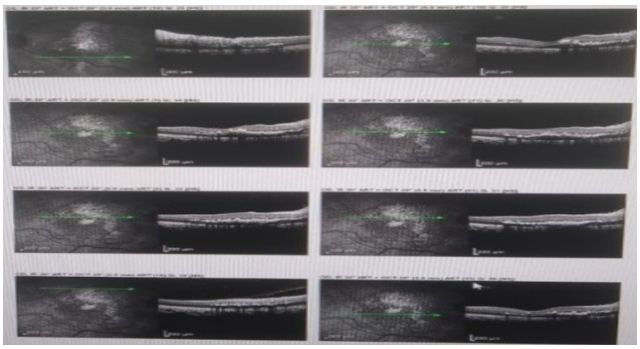

Fig. 2: Subretinal fluid and foveal contour

The patient was admitted, and two hourlies BP charting was done. BP control of the same was done with alpha and beta blockers and other antihypertensives as needed based on the monitoring report; with the control of BP, the patient showed improvement in visual acuity on the fifth day with the right eye 6/36 partial. Twenty-four-hour urine samples showed raised metanephrine levels, and she was planned for surgical removal after consulting an endocrinologist. Automated visual field testing 30-2 threshold SITA showed cecocentral scotomas, as in Figure 3.

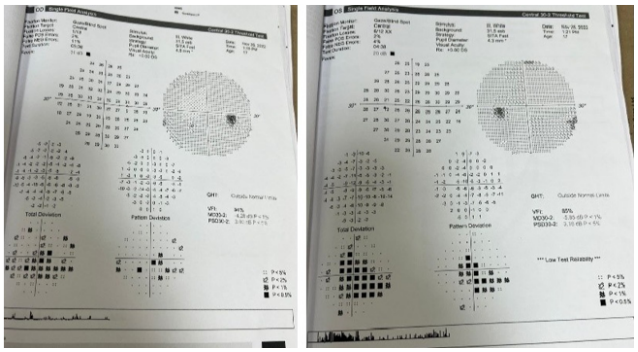

Fig. 3: Ceco central scotomas

3. Discussion

Hypertensive retinopathy is often characterized by retinal vascular changes in the retina in the event of increased BP. Though changes are most frequently seen in the retina, but choroid and optic nerve can also be involved,[4] A significant factor in secondary hypertension that might result in hypertensive retinopathy is Pheochromocytoma. Once therapy is initiated at an early stage, as in our instance, alterations in such circumstances are frequently reversible, and the condition remains stable.

Retinopathy, which manifests as arteriolar constriction, occlusion, ischemia, and subsequent smooth muscle necrosis, is typically the first sign of malignant hypertension. Following this vasodilation, the loss of autoregulation and the downstream transfer of elevated BP may occur. A failure of autoregulation occurs when perfusion pressure changes outside a crucial autoregulatory range. In other words, auto-regulation does not always shield retinal vessels. The retinal tissue may experience ischemic damage if the perfusion pressure exceeds (malignant hypertension) or falls below (arterial hypotension) the critical range,[5] There are four overlapping and occasionally non-sequential phases that make up the pathophysiology of hypertensive retinopathy: a vasoconstrictive phase, an exudative phase, a sclerotic phase, and consequences of the sclerotic phase.

In our case, the patient had a macular star, an uncommon finding caused by exudates collecting in the retina's outer plexiform layer (or Henle's layer). As the exudates

accumulate in the outer plexiform layer, the orientation of neurons in the foveal region, or central part, of the macula becomes much more oblique, resulting in the star pattern.[6,7] The collapse of the inner blood-retinal barrier causes retinal hemorrhages. The course of the nerve fiber layer is followed by bleeding, resulting in flame-shaped hemorrhages and bleeding within the retina (intra-retinal hemorrhages).

For various reasons, acute rises in BP significantly impact the choroid, which feeds blood vessels to the retina rather than the retina itself. Because choroid vessels have a relatively short course with little branching, autoregulation is not as good as retinal vessels. Acute BP increases because of choroidal ischemia and retinal pigment epithelium necrosis.[8–10] The retinal pigment epithelium is responsible for maintaining retinal attachment, and its loss results in the buildup of subretinal fluid, as shown in our instance.

4. Conclusion

Pheochromocytoma-induced hypertensive retinopathy, although rare, is an important cause of visual and systemic morbidity. Timely diagnosis is important, and hormonal investigations and immunohistochemistry play a significant role. The changes are reversible with resection of the tumor, and the visual prognosis is good in the early stages. This case is unique as it highlights the importance of systemic examination. If hypertensive ocular findings are seen in young patients, data about the same is limited to a few case reports.

5. Conflict of Interest

None.

6. Source of Funding

None.

References

1. L P. Das ganglioma embryonale sympathicum. *Berl Klin Wschr*. 1912;49:16–22.
2. Manger WM, Gifford RJ. Pheochromocytoma. *J Clin Hypertens (Greenwich)*. 2012;4(1):62–72. doi:10.1111/j.1524-6175.2002.01452.x.
3. Jr WY. Endocrine hypertension: then and now. *Endocr Pract*. 2010;16(5):888–902. doi:10.4158/EP10205.RA.
4. Henderson AD, Bruce BB, Newman NJ, Biousse V. Hypertension-related eye abnormalities and the risk of stroke. *Rev Neurol Dis*. 2011;8(1-2):1–9.
5. Pelino CJ, Pizzimenti JJ. Essentials of hypertension: high blood pressure remains a major health issue worldwide, with profound–and often silent–multisystemic effects. *Rev Optom*. 2014;151(9):88–90.
6. Hayreh SS, Servais GE, Virdi PS. Fundus lesions in malignant hypertension: V. Hypertensive optic neuropathy. *Ophthalmology*. 1986;93(1):74–87. doi:10.1016/s0161-6420(86)33773-4.
7. Hayreh SS, Servais GE, Virdi PS. Fundus lesions in malignant hypertension: IV. Focal intraretinal periarteriolar transudates. *Ophthalmology*. 1986;93(1):60–73.
8. Suzuki M, Minamoto A, Yamane K, Uka J, Aoki S, Mishima HK, et al. Malignant hypertensive retinopathy studied with optical coherence tomography. *Retina*. 2005;25(3):383–4.
9. Bourke K, Patel MR, Prisant LM, Marcus DM. Hypertensive choroidopathy. *J Clin Hypertension*. 2004;6(8):471–2.
10. Lee AG, Beaver HA. Acute bilateral optic disk edema with a macular star figure in a 12-year-old girl. *Survey Ophthalmol*. 2002;47(1):42–9.

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