CASE REPORT

Sudden Vision Loss in a Young Patient with Pulmonary Hypertension, A Rare and Unusual Finding

Saba Alkhairy[1] and Saad Aslam[1*]

[1]*Department of Ophthalmology, Dow International Medical College, DUHS, Karachi, Pakistan*

Abstract

Loss of vision associated with pulmonary hypertension in the pediatric age group has been rarely reported in the literature and often goes unnoticed for a long period. Pulmonary hypertension may manifest as shortness of breath limiting the activity of the individual, palpitation, headaches, easy fatiguability, dizziness been the most common symptoms encountered. However, primary pulmonary hypertension would not present itself instead secondary to other diseases like congenital heart disease, lung disease, connective tissue disorders or genetic diseases. Our case demonstrated a 17-year old individual with a primary complaint of progressive loss of vision in the left eye leading to complete vision loss. The patient experienced dyspnea two years back which exacerbated affecting his normal routine activity. His Echocardiography revealed severe pulmonary hypertension and was started on appropriate medications. His vision worsened and slit-lamp examination revealed multiple hemorrhages. Further investigations with FFA and OCT revealed significant hypoperfusion and significant elevation of the retina of the left eye. Anti-VEGF injections were given in an attempt to restore vision, but it resulted in no further improvement. Our case emphasizes the importance of prompt ophthalmologic examination in Pulmonary Hypertension.

Keywords: *Familial pulmonary hypertension, macular edema, central retinal vein occlusion.*

INTRODUCTION

Pulmonary Hypertension is defined as increased pulmonary arterial pressure which in childhood presents even at rest [1]. Pulmonary Hypertension is a rare and life-threatening disease of both pediatric and adult age groups. The etiology of PH has been classified by the World Health Organization based on the mechanism of an underlying disease. However, the most common cause of Pulmonary Hypertension is Left Heart Failure. Untreated Pulmonary Hypertension leads ultimately to death [2]. Increased venous back pressures dilate the ocular veins, increasing the IOP increasing the risk of ciliary detachment, venous occlusions and several other Ocular complications [2, 3]. Treatment of PAH is usually done systemically aimed to reduce Pulmonary Hypertension thereby reducing the risk of complications leading to organ damage . Ocular manifestations due to Pulmonary Hypertension has not been published extensively. We present the case of a patient who presented with loss of vision due to Central Retinal Vein Occlusion secondary to Pulmonary Hypertension.

CASE PRESENTATION

We are presenting the case of a 17-year old male patient who presented in the Ophthalmology Out-Patient department 4 weeks back with the complaint of a sudden loss of vision in the left eye. The patient experienced blurring of vision in the left eye more compared to the right eye which gradually increased leading to complete loss of vision. It remained unnoticed until the patient developed difficulty reading at school. On further questioning, it was revealed that the patient had developed dyspnea two years back. The patient never developed any symptoms at all before this event. The patient experienced a significant increase in dyspnea making him short of breath with little exertion. Family history revealed that a paternal female first cousin also has pulmonary hypertension which was detected a few years back.

On visual assessment, his best-corrected vision was 6/12 OD and 1/60 OS without any further improvement on Snellen's Chart. Slit Lamp examination showed a normal anterior segment with a clear cornea, without any flares or cells, the pupil was round and regular with a normal lens as well as no cells in vitreous were visualized. The right eye fundus was normal with a normal optic disk and a healthy macula with the vasculature. Whereas the left eye had a normal anterior segment with a clear cornea however the fundus exam revealed pre-retinal and intraretinal hemorrhages, dilated and tortuous veins and macular edema (**Fig. 1A**). Goldmann Applanation Tonometry was performed on a slit lamp and intra-ocular pressures were found out to be within normal limits in both eyes. To further investigate, a Fundus Fluorescein Angiography with an SD-OCT was performed which showed normal right eye with normal retinal thickness and a normal FFA clear of hemorrhages, or hypoperfusion (**Fig. 1B**). However, the left eye Fundus Fluorescein Angiography images revealed areas of hypoperfusion in all four quadrants

**Corresponding author: Saad Aslam, Department of Ophthalmology, Dow International Medical College, DUHS, Karachi, Pakistan;*
Email: saad.aslam1@hotmail.com
Received: April 07, 2020; Revised: December 07, 2020; Accepted: December 09, 2020
DOI: https://doi.org/10.37184/lnjpc.2707-3521.2.11

(**Fig. 1C**). SD-OCT of the left eye showed an abnormally thick retina with significant elevation of the macula and with hemorrhages as well as areas of hypoperfusion were identified (**Fig. 1D**).

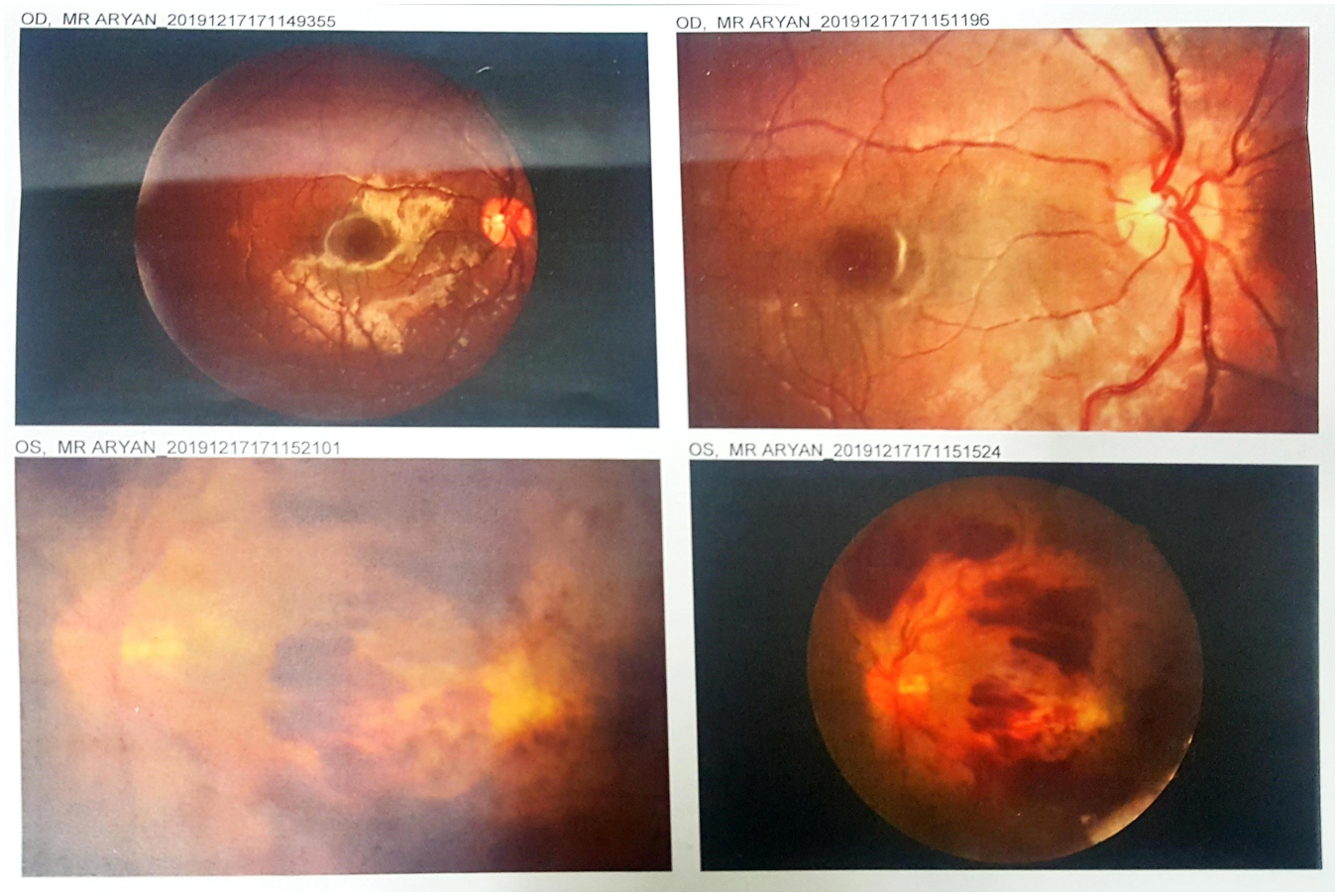


Fig. (1A): Fundus images showing a normal pattern in Right eye (OD). Fundus of Left eye (OS) depicting widespread Intra- Retinal and Pre-Retinal Hemorrhages. OS= Left Eye, OD= Right Eye. Patient reviewed at Department of Ophthalmology, Dow University of Health Sciences, Ojha Campus, Karachi, Pakistan.

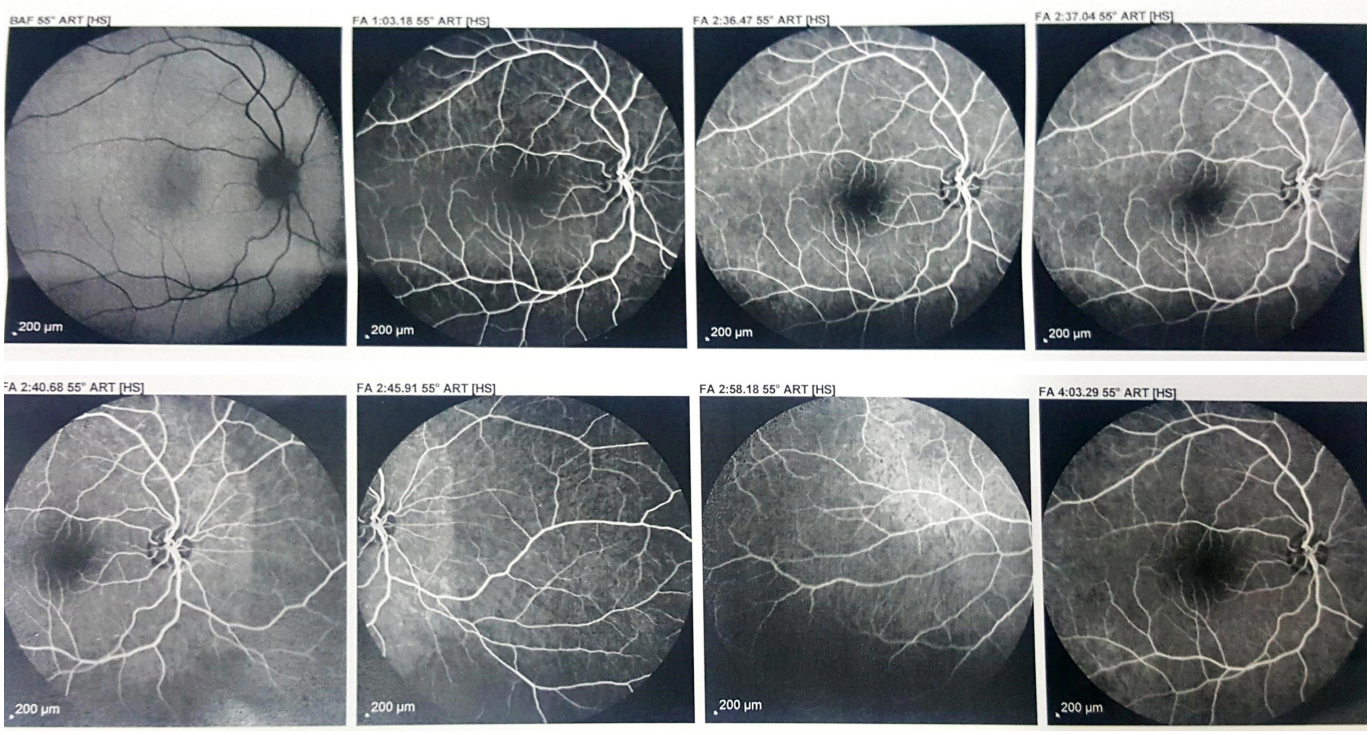


Fig. (1B): FFA images of the Right Eye.

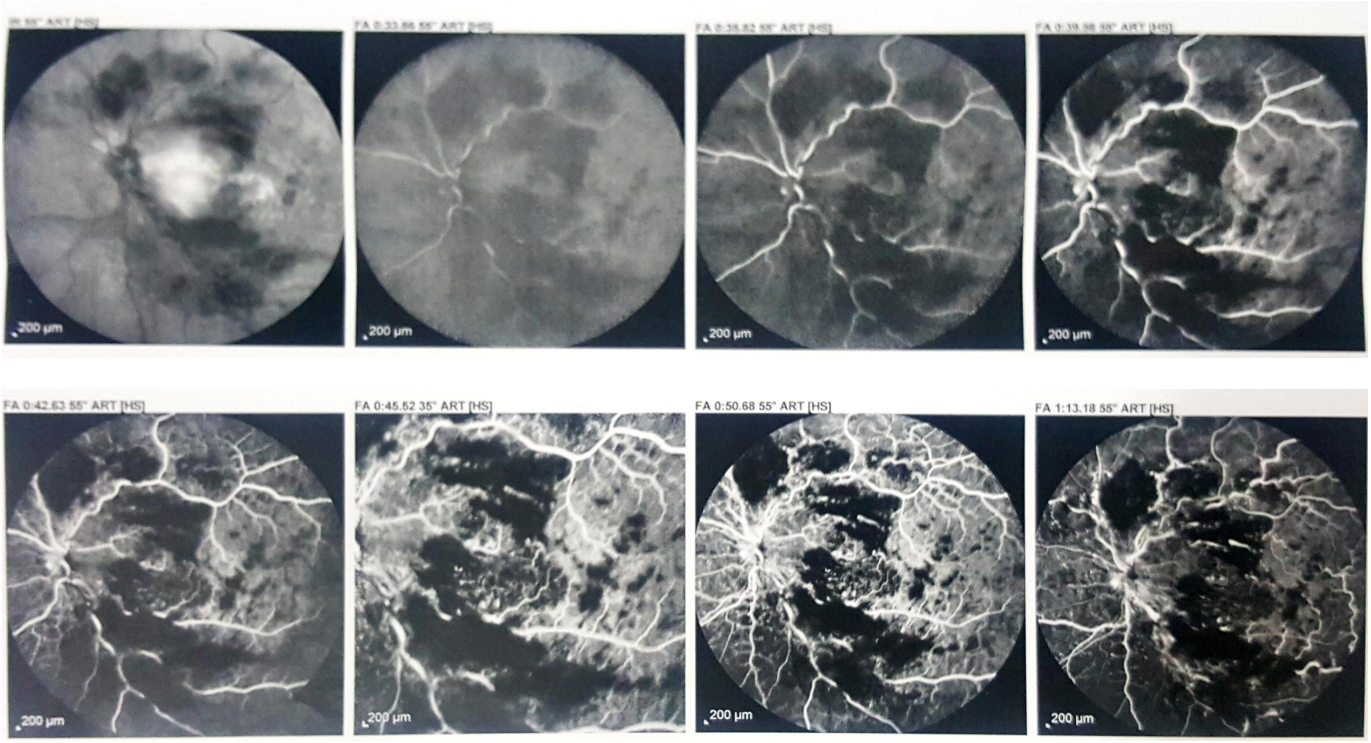


Fig. (1C): FFA images of the Left Eye depicting Intra and Pre- Retinal Hemorrhages and areas of hypoperfusion in all quadrants.

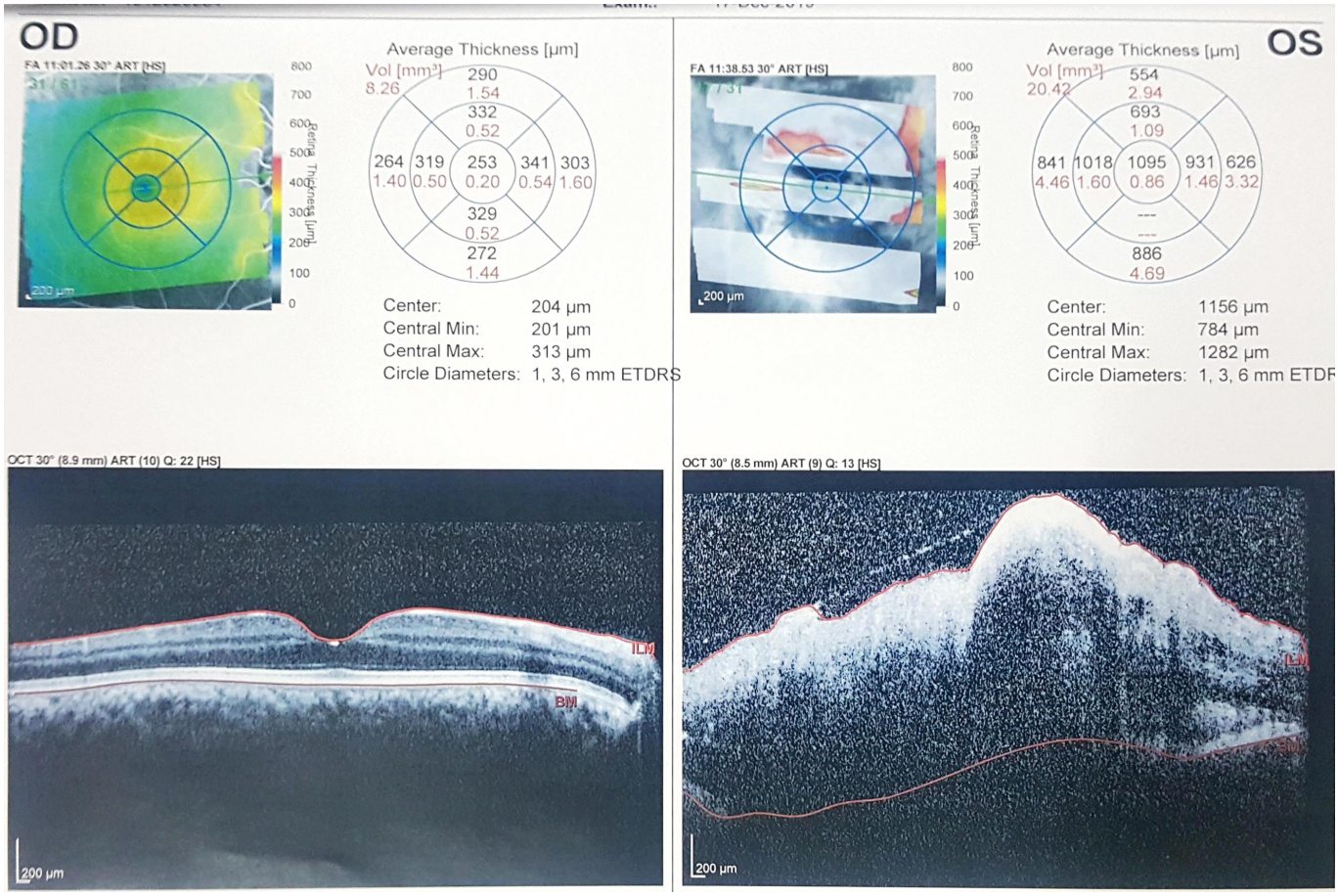
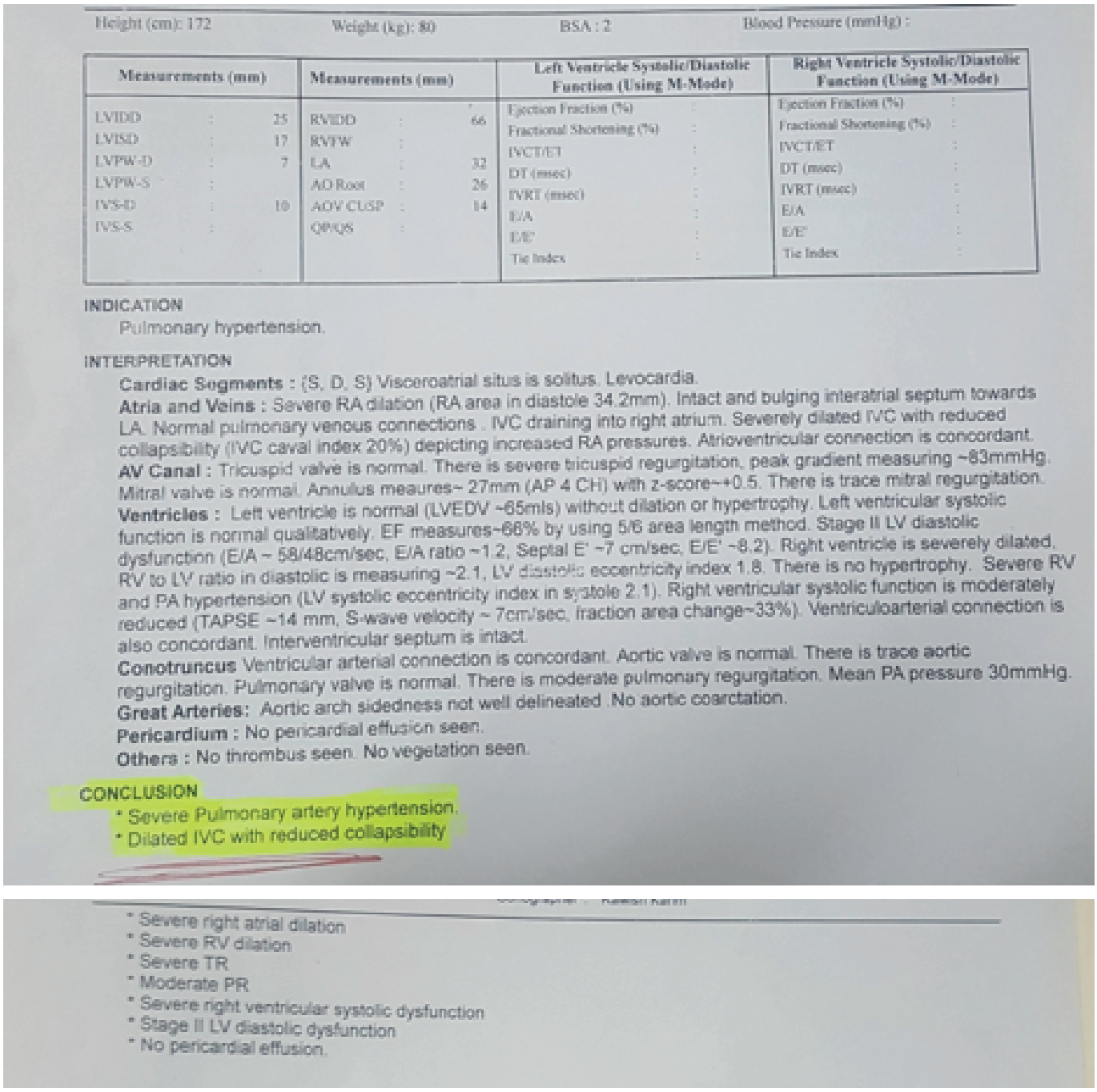


Fig. (1D): Spectral Domain- OCT; Right Eye (OD): Normal thickness Macula with intact Retina. Left Eye (OS): Increased thickness of retina with significant elevation of at the Macula.

Laboratory investigations revealed normal routine lab results with a Hemoglobin level of 14.3g/dl and hematocrit of 44%. Pediatric Transthoracic Echocardiography (**Fig. 2**) showed severe Pulmonary Hypertension, Dilated IVC, Tricuspid Regurgitation, Right Ventricular Dilatation, and Enlarged Right Atrium, Moderate Pulmonary Regurgitation.

Given the presence of hemorrhages and areas of hypoperfusion in the left eye confirmed on Fundus Fluorescein Angiography and SD-OCT (**Fig. 1A-1D**), a diagnosis of Central Retinal Vein Occlusion was made, and subsequent treatment was planned.

The patient is currently taking Sildenafil (PDE5 inhibitor), Bosentan (ET-1 Antagonist) and Spironolactone (Potassium-sparing diuretic). After the induction of medications, his episodes of dyspnea improved significantly. Treatment for the left eye was begun with Intra-Vitreal Avastin Injections. Three Intra-Vitreal Avastin Injections were given at regular intervals of one week apart. The patient was again examined after giving Intra-Vitreal Injections, but it did not improve the overall visual acuity as well as the hemorrhages. Treatment with Acetazolamide 250mg BD was begun but it did not resolve his ocular symptoms either.



Height (cm): 172 Weight (kg): 80 BSA : 2 Blood Pressure (mmHg) :

Measurements (mm)		Measurements (mm)		Left Ventricle Systolic/Diastolic Function (Using M-Mode)	Right Ventricle Systolic/Diastolic Function (Using M-Mode)
LVIDD : 25		RVIDD : 66		Ejection Fraction (%) :	Ejection Fraction (%) :
LVISD : 17		RVFW :		Fractional Shortening (%) :	Fractional Shortening (%) :
LVPW-D : 7		LA : 32		IVCT/ET :	IVCT/ET :
LVPW-S :		AO Root : 26		DT (msec) :	DT (msec) :
IVS-D : 10		AOV CuSP : 14		IVRT (msec) :	IVRT (msec) :
IVS-S :		QP/QS :		E/A :	E/A :
				E/E' :	E/E' :
				Tie Index :	Tie Index :

INDICATION

Pulmonary hypertension.

INTERPRETATION

Cardiac Sogments : (S, D, S) Visceroatrial situs is solitus. Levocardia.
Atria and Veins : Severe RA dilation (RA area in diastole 34.2mm). Intact and bulging interatrial septum towards LA. Normal pulmonary venous connections . IVC draining into right atrium. Severely dilated IVC with reduced collapsibility (IVC caval index 20%) depicting increased RA pressures. Atrioventricular connection is concordant.
AV Canal : Tricuspid valve is normal. There is severe tricuspid regurgitation, peak gradient measuring ~83mmHg. Mitral valve is normal. Annulus meaures~ 27mm (AP 4 CH) with z-score~+0.5. There is trace mitral regurgitation.
Ventricles : Left ventricle is normal (LVEDV ~65mls) without dilation or hypertrophy. Left ventricular systolic function is normal qualitatively. EF measures~68% by using 5/6 area length method. Stage II LV diastolic dysfunction (E/A ~ 58/48cm/sec, E/A ratio ~1.2, Septal E' ~7 cm/sec, E/E' ~8.2). Right ventricle is severely dilated. RV to LV ratio in diastolic is measuring ~2.1, LV diastolic eccentricity index 1.8. There is no hypertrophy. Severe RV and PA hypertension (LV systolic eccentricity index in systole 2.1). Right ventricular systolic function is moderately reduced (TAPSE ~14 mm, S-wave velocity ~ 7cm/sec, fraction area change~33%). Ventriculoarterial connection is also concordant. Interventricular septum is intact.
Conotruncus Ventricular arterial connection is concordant. Aortic valve is normal. There is trace aortic regurgitation. Pulmonary valve is normal. There is moderate pulmonary regurgitation. Mean PA pressure 30mmHg.
Great Arteries: Aortic arch sidedness not well delineated .No aortic coarctation.
Pericardium : No pericardial effusion seen.
Others : No thrombus seen. No vegetation seen.

CONCLUSION

* Severe Pulmonary artery hypertension.
* Dilated IVC with reduced collapsibility

* Severe right atrial dilation
* Severe RV dilation
* Severe TR
* Moderate PR
* Severe right ventricular systolic dysfunction
* Stage II LV diastolic dysfunction
* No pericardial effusion.

Fig. (2): Pediatric Transthoracic Echocardiography.

DISCUSSION

Pulmonary Arterial Hypertension is a rare disease in infants and children with significant morbidity and mortality. In our case report, we have presented ocular findings that were found in a very young patient, affecting his left eye that significantly deteriorated his visual acuity and is now limited to the only perception of light. The symptoms experienced by patients are non-specific including decreased vision, metamorphopsia, and pain due to increased IOP as reported [2]. Causes of Pulmonary Hypertension include both familial as well as idiopathic type. Primary pulmonary hypertension accounts for about 6% of all cases of pulmonary hypertension, and most of the reported ocular findings are associated with this subtype [2]. Austin *et al.* described the role of a genetic mutation that occurs in BMPR2 gene [4]. The pathophysiology of the disease reported has been vascular remodeling, lack of endothelin further exacerbated with underlying congenital heart disease. The significance of considering a familial cause of it; is the presentation at a younger age with gene anticipation observed in successive generations [1, 4].

Central Retinal Vein Occlusion, one of the feared complications of Pulmonary Hypertension occurred unilaterally in our patient, a similar article highlighted the importance of managing it earlier on as it increased the chance of involving the fellow eye. Increased IOP results in compressing the central retinal vein significantly [5]. Apart from CRVO our patient's OCT exhibited unusual Macular elevation deteriorating its normal architecture.

As Hammond *et al.* reported using acetazolamide which subsided macular edema and improved vision [6]. Given the unique nature of the ocular finding in our patient, we did use acetazolamide but to no resolve.

Most certainly it can be concluded that earlier diagnosis and systemic treatment in such a patient with pulmonary hypertension could avoid drastic complications.

CONCLUSION

Given the published evidence and thorugh experiences of different ophthalmologists around the world it is pertinent to say that such patients with pulmonary hypertension be referred to ophthalmologists early on to prevent loss of vision associated with it especially patients of pediatric age group, as our patient developed CRVO secondary to pulmonary hypertension.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

ACKNOWLEDGEMENTS

Declared none.

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