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POLYPOIDAL CHOROIDAL VASCULOPATHY MANIFESTATION IN EXTRA-AXIAL CAVERNOUS TUMOR: A RARE FINDINGS IN CHRONIC PAPILLEDEMA

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Abstract

Introduction: Age-related macular degeneration (AMD) is a leading cause of blindness worldwide. Polypoidal choroidal vasculopathy (PCV), a subtype of neovascular AMD, is characterized by an abnormal branching network of vessels with aneurysmal dilations (polyps). Choroidal neovascularization can also result from chronic disc edema. This case presents a rare occurrence of PCV in a patient with an extra-axial cavernous tumor and explores the best management approach.

Case Report: A 43-year-old man presented with an 8-month history of blurry vision in his right eye, double vision on left gaze, and left eye protrusion, accompanied by headaches and occasional nausea. Visual acuity was 2/60 in the right eye and 5/10 in the left improving with pinhole become 5/6.5. RAPD was found in left eye. Funduscopy revealed peripapillary atrophy in both eyes, with exudates in the right macula. OCT showed dome-shaped polyps in both eyes, larger in the right. MRI revealed a left sphenoid meningioma compressing the orbital cavity. The patient was diagnosed with both eyes PCV and compressive optic neuropathy, left eye multiple cranial nerve palsy and dyslipidemia. Intravitreal anti-VEGF injection was planned.

Discussion PCV is a subtype of AMD characterized by recurrent serosanguineous detachments. Chronic papilledema, possibly due to intracranial tumors, may lead to visual loss from retinal nerve fiber damage or neovascularization. Chronic posterior globe flattening and choroidal vessel abnormalities likely contributed to PCV development.

Conclusion: PCV may result from chronic papilledema due to intracranial tumors. Anti-VEGF therapy offers a viable treatment option, balancing polyp regression and visual acuity stabilization.

Keywords: Polypoidal Choroidal Vasculopathy, Choroidal neovascularization, Papilledema, Meningioma **Cite This Article:** ASTI, Annisa Kinanti; AGUSTINI, Lukisiari; FIRMANSJAH, Muhammad. POLYPOIDAL CHOROIDAL VASCULOPATHY MANIFESTATION IN EXTRA-AXIAL CAVERNOUS TUMOR: A RARE FINDINGS IN CHRONIC PAPILLEDEMA. **International Journal of Retina**, [S.l.], v. 8, n. 1, p. 57, mar. 2025. ISSN 2614-8536. Available at: <https://www.ijretina.com/index.php/ijretina/article/view/306>. Date accessed: 05 mar. 2025. doi: https://doi.org/10.35479/ijretina.2025.vol008.iss001.306....

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INTRODUCTION

Worldwide, 170 million people will be blind due to age-related macular degeneration (AMD) by 2040, with 110 million instances predicted in Asia alone.[1] An irregular branching network of blood vessels with aneurysmal dilations called polyps characterises polypoidal choroidal vasculopathy (PCV), a variant of neovascular AMD. PCV is also known as type 1 neovascularisation. In extreme cases, persistent disc oedema might lead to choroidal neovascularisation.[2,3] Diagnosing papilledema can be challenging, particularly when it is accompanied by visual loss. The narrowing of visual fields (VF) caused by the gradual atrophy of retinal nerve fibres is the usual cause of this visual impairment. Occasionally, it can develop as a result of choroidal folds, neovascularisation, macular exudates, or haemorrhages; the latter is more common in long-term instances.[5]

The goal of this case study is to discuss the optimal course of treatment for a patient who presented with the unusual symptoms of polypoidal choroidal vasculopathy in addition to their extra-axial cavernous tumour.

CASE REPORT

A 43-year-old man presented to the outpatient clinic with complaints of blurry vision in his right eye, ongoing for the past 8 months. The complaint also accompanied by double vision when looking to the left. Over the same period, the left eye had become protruded, occur with headaches, nausea, and occasional vomiting. The patient was referred by the neurosurgery department due to the detection of a tumor. No history of systemic disease was found, but had undergone strabismus surgery a year earlier. The patient family history included cervical cancer in the patient's mother.

On examination, the patient visual acuity was 2/60 in the right eye and 5/10 in the left, improving to 5/6.5 with pinhole correction. Intraocular pressure (IOP) was normal in both eyes. Color vision was impaired in the right eye (1/38) but normal in the left (38/38). Ocular motility was restricted in the left eye, with -3 limitation in the superotemporal, temporal, and inferotemporal directions, though without pain during movement. Confrontation testing was normal, and the Hertel test measured 20-121-22. The Hirschberg test showed orthophoria. A relative afferent pupillary deficit (RAPD) measuring 4 mm was detected in the left eye, but the anterior portion of the right eye seemed to be functioning normally.

Peripapillary atrophy and inferotemporal perimacula exudates were seen during funduscopy in the right eye, whereas the left eye also showed signs of this condition. Ocular Coherence Tomography (OCT) scans of the macula in both eyes showed a dome-shaped or thumb-like elevation polyp with hyperreflective rings and a hyporeflective and hyperreflective lumen within,

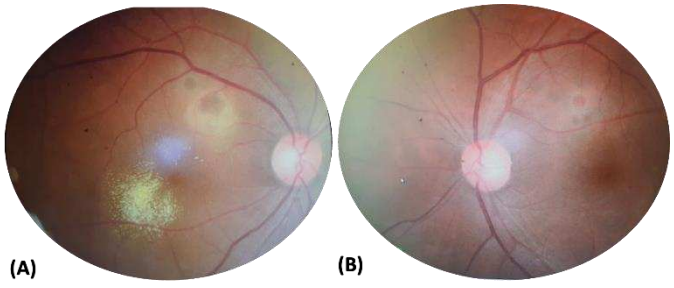


Figure 1. (A) Right eye funduscopy show peripapillary atrophy around the optic nerve head and exudate in inferotemporal perimacula. (B) The peripapillary atrophy findings in left eye. **Pictures taken with patient's consent. Courtesy: Poli Mata RSUD Dr. Soetomo.*

with the polyp being more prominent and elevated in the right eye. Between the Bruch's membrane and another hyperreflective tissue is the double-layer sign, which is depicted in Figure 2, which is a pigment epithelial separation notch. There were no noticeable issues with the optic nerve head (ONH) or retinal nerve fibre layer (RNFL) complex in either eye's macula. The layers are staying the same thickness. Retinal pigment epithelium and internal limiting membrane swelling is observed in the nasal, lateral, and superior quadrants of the right eye. In the left eye, the upper quadrant is thicker while the lower and nasal quadrants are thinner.

The Humphrey Visual Field (HVF) test for both eyes was deemed unreliable due to excessive fixation losses and a high rate of negative errors (Figure 3). Laboratory tests revealed elevated lipid levels, and the patient's HbA1C was in the prediabetic range. All IgG tests, including Toxoplasma, Rubella, and Cytomegalovirus (CMV), returned positive. A contrast-enhanced brain MRI showed a left sphenoid meningioma causing slight compression of the orbital cavity, along with left ethmoidal sinusitis, bilateral mastoiditis (more prominent on the left), nasal septum deviation, and chronic rhinitis.

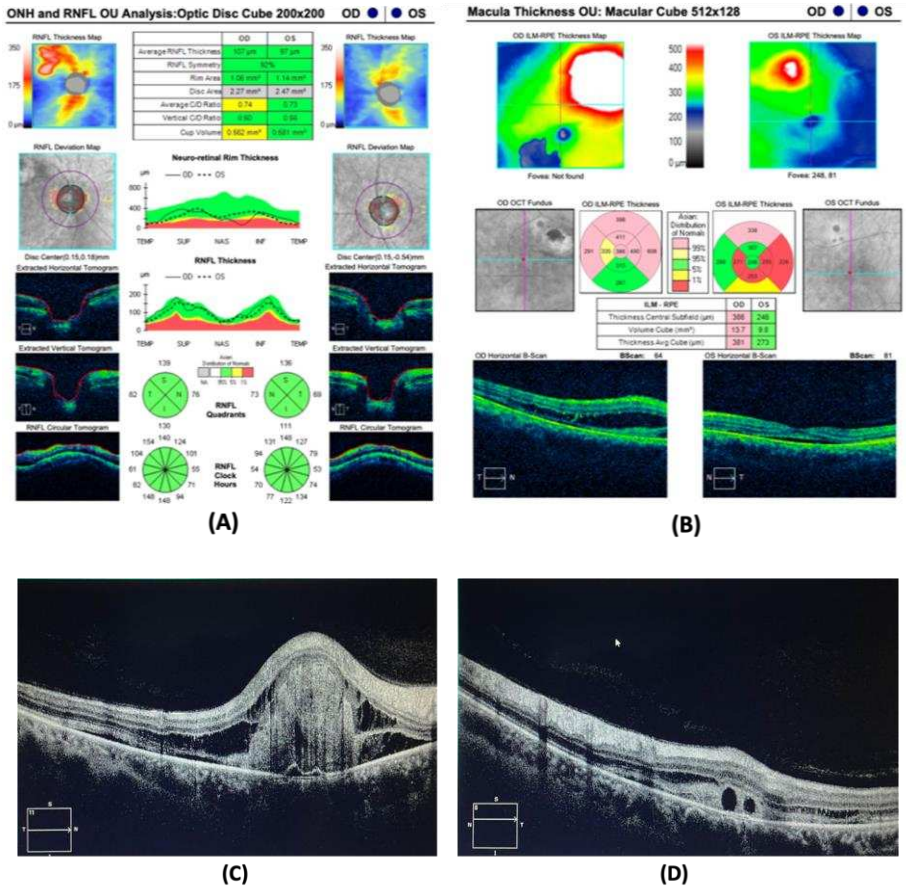
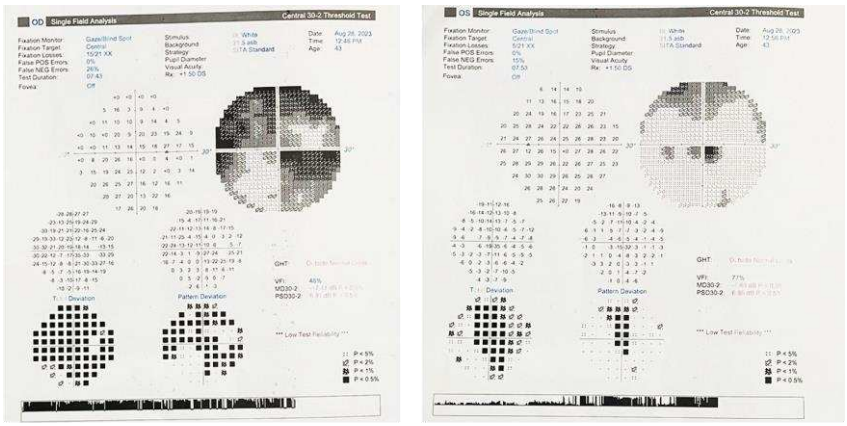


Figure 2. (A) ONH and RFNL OCT in both eyes are within normal limit. (B) Macula Thickness OCT represent edema in some quadrants. (C) Thumb-like elevation polyp is shown in HD-21 OCT, and shown in the left eye but smaller in size (D) **Pictures taken with patient's consent. Courtesy: Poli Mata RSUD Dr. Soetomo*.*



(A) (B)

Figure 3. Humphrey Visual Field examination in right eye (A) and left eye (B) show unreliable result. *Courtesy: Poli Mata RSUD Dr. Soetomo.*

The patient was found to have symptoms such as compressive optic neuropathy in both eyes, dyslipidaemia, a suspected left-sided extra-axial cavernous meningioma, and left-eye cranial nerve III and VI palsy with pupillary involvement, according to the results of the examination. The treatment plan includes an intravitreal anti-VEGF injection for the left eye, while conservative management is recommended for the right eye. Routine follow-up visits are advised prior to tumor excision via craniotomy to monitor for any unexpected ocular changes. Unfortunately, this patient had return to the patient place in Gorontalo and no further follow up.

DISCUSSION

One kind of age-related macular degeneration is polypoidal choroidal vasculopathy, or PCV. Recurrent serosanguineous detachments of the retinal pigment epithelium (RPE) characterise this variety of type 1 macular neovascularisation (MNV), which is also known as posterior uveal haemorrhage syndrome. A unique "string-of-pearls" pattern is formed when feeder arteries attach to the RPE layer of the fibrovascular pigment epithelial detachment (PED) and create a network of polypoidal lesions. This pattern is characterised by PCV.[6,7]

When new blood vessels sprout through Bruch's membrane into the sub-retinal pigment epithelium (RPE) region, it is called type 1 macular neovascularisation (MNV) or occult choroidal neovascularisation (CNV). A vascularised serous or fibrovascular pigment epithelial detachment (PED) with an uneven surface can develop as a consequence of bleeding and leakage caused by this process. Type 2 macular hole (MNV), also called classic CNV, is marked by the formation of new blood vessels that link the neurosensory retina to the retinal pigment epithelium (RPE). In such a scenario, an examination can show a gray-green or lacy lesion. A minimally classic CNV or mixed type 1 and type 2 MNV is a form of neovascularisation that extends beneath the RPE and the neurosensory retina. The genesis of type 3 MNV is aberrant capillaries in the retina's deep capillary plexus; these growths, formerly called retinal angiomatous proliferations, eventually reach the RPE. Before this subtype fully develops, intraretinal pigment migration may occur. On examination, type 3 MNV often appears as a small red area with associated retinal exudate or subretinal fluid accumulation.[6]

PCV is identified in women and men of all races. In Asian population, 20%–50% of cases of nAMD are PCV type. In white people less than 5% of cases of MNV are PCV type. [6] One study found that between 20% and 60% of ICGA cases had polypoidal choroidal vasculopathy (PCV). In contrast, the percentage of PCV is much lower, ranging from 8% to 13% in cases when ICGA was performed, [8-9] There is a hereditary component to PCV risk factors,

Table 1. Polypoidal Choroidal Vasculopathy Diagnosis Criteria. [7]

"Polypoidal Choroidal Vasculopathy Diagnostic Criteria	
Japanese Study Group Guidelines	Definite PCV: • Elevated orange-red lesions on fundus examination, and/or Polypoidal lesions on ICGA
	• Probable PCV: Only abnormal BVN seen on ICGA or recurrent hemorrhagic or serous • RPE detachments, or both, without features of definite PCV
EVEREST criteria (based on confocal scanning laser ophthalmoscopy)	Focal hyperfluorescent lesions appearing before 6 minutes on ICGA, plus at least 1 of the following: • BVN on ICGA • Pulsatility on dynamic ICGA • Nodular appearance when ICGA viewed stereoscopically • Hypofluorescent halo on ICGA • Orange subretinal nodule on color photograph • Associated massive submacular hemorrhage
BVN: branching vascular network; ICGA: indocyanine green angi- ography; PCV: polypoidal choroidal vasculopathy; RPE: retinal pigment epithelium".	

and both PCV and conventional AMD share systemic risk factors such cigarette smoking, [8], obesity, and elevated C-reactive, [9,11] protein and other inflammatory indicators in the blood. [10,12] Some research shows a higher prevalence of dyslipidemia in patients with PCV compared to other subtypes of age-related macular degeneration (AMD), and dyslipidemia has been linked to increased severity or recurrence of PCV due to its role in promoting vascular instability.[13]

In the pathophysiology of PCV, there is localized choroidal thickening at the site of the disease, caused by abnormally dilated veins in the Haller layer, known as pachyvesselsDue to the presence of these pachyvessels, the Sattler and choriocapillaris layers that are located above the Bruch's membrane-RPE complex become thinner and eventually bleed. The subfoveal area and overall choroidal thickness is thicker in PCV patients.[14] Different forms of polypoidal choroidal vasculopathy (PCV) manifest in different ways. The lack of subretinal or intraretinal fluid as well as haemorrhage is a hallmark of quiescent PCV. Lipid exudation, neurosensory retinal thickening, pigment epithelial detachment (PED), subretinal fluid, or a mix of these traits is found in exudative PCV, but haemorrhage is not. The subretinal or sub-RPE haemorrhage that characterises hemorrhagic PCV can happen with or without the presence of exudative alterations.[15] In this case, the presentation is likely an exudative PCV type.

Polypoidal choroidal vasculopathy (PCV) can be diagnosed by funduscopy with reddish-orange subretinal nodules, serous retinal detachment, many hard exudates, or severe haemorrhage. On optical coherence tomography (OCT), the retinal pigment epithelium (RPE) appears as dome-shaped or thumb-like elevations, some of which may have internal polyps and others do not. The double-layer sign, consisting of hyperreflective tissue separating Bruch's membrane from hyperreflective RPE, is another discovery. Finally, pachyvessels or pachychoroid characteristics, which are enlarged blood vessels, may be present in Haller's layer.[6]

The presence of polypoidal dilatations of the choroidal circulation, along with or without feeder and draining veins, can be detected by Indocyanine green angiography (ICGA) as hyperfluorescent patches. Occult neovascularisation (NV) is the most common way that fluorescein angiography (FA) detects PCV, however a conventional NV pattern can be seen in few cases.

A worse visual prognosis may be indicated in cases of PCV that present as classic CNV on FA. The inability of FA to detect polyps and other sub-RPE structures limits its use in PCV diagnosis. In PCV, OCT-A gives a two-dimensional picture of aberrant vasculature, just like ICGA. In most cases, OCT-A can detect the branching vascular network (BVN), however it is far less effective than ICGA at identifying polyps.[7] ICGA, FA, and OCT-A are not performed in our case, but the diagnosis made based on the diagnosis criteria (Table 1).

When compared to other forms of macular neovascularisation (MNV), polypoidal choroidal vasculopathy (PCV) often responds less favourably to anti-VEFG treatment. After a year, the visual outcomes with anti-VEFG monotherapy or combination therapy with photodynamic therapy (PDT) are outstanding, on par with those from normal neovascular AMD, according to the EVEREST-II and PLANET trials. Therefore, for patients experiencing symptoms of PCV, both approaches are considered effective initial therapies.[6]

Signs of increased intracranial pressure (ICP) include papilledema, which manifests as fuzzy and raised optic disc borders. ICP can be caused by tumours within the brain, infections, haemorrhages, or blockage of the cranial ventricular drainage system. When papilledema is associated with visual loss—usually as a result of progressive damage to the retinal nerve fibres that narrows the visual fields—or, less frequently, with macular exudates, haemorrhages, choroidal folds, or neovascularization—common in long-term cases—the diagnosis becomes much more difficult.[16,17] It is believed that pressure-induced distortion at the border of Bruch's membrane near the optic disc causes choroidal neovascularisation (CNV) to occur in chronic papilledema. This anatomic weakening, combined with hypoxia from axonal swelling and subsequent vascular perfusion changes, promotes angiogenesis.[17]

Chronic papilledema may present with optic disc pallor, reduced swelling from axonal loss, gliosis (scarring of retinal fibers), optociliary shunts, and refractile bodies. In papilledema, the forward shift of the optic nerve head and surrounding peripapillary tissues can cause compression or ischemia. Chronic flattening of the posterior globe, along with abnormalities in the choroidal vessels, may have contributed to the onset of PCV in this case.[3,18,19] The patient, with left sphenoid meningioma and early papillary atrophy, shows signs of chronic papilledema, as explained by the presumed pathophysiology. The presence of an extra-axial cavernous tumor, specifically a left sphenoid meningioma, likely contributed to the development of PCV. The tumor-induced compression of the orbital cavity and optic nerve led to chronic posterior globe flattening and increased intracranial pressure, which could compromise the choroidal circulation and structural integrity of the posterior segment. This mechanical stress, combined with chronic ischemia and inflammation, may have triggered vascular remodeling and the formation of the polypoidal lesions characteristic of PCV. The co-occurrence of dyslipidemia in this patient further exacerbates vascular instability, increasing the risk of neovascularization and disease progression. The observed clinical features—dome-shaped polyps in both eyes and compressive optic neuropathy—underscore the interplay between systemic conditions (dyslipidemia), local tumor effects, and choroidal vascular pathology in PCV development.

CONCLUSION

Polypoidal choroidal vasculopathy (PCV) is a rare condition that describes a choroidal artery BVN and polypoidal dilations, both of which impact the choroidal vasculature. Possible contributors to the development of PCV in this case include intrinsic choroidal vascular abnormalities and chronic posterior globe flattening.

It is possible that an intracranial tumour produced the chronic papilledema. Either monotherapy or combo therapy with anti-VEFG can stabilise visual acuity and balance polyp regression.

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