

## Asteroid Hyalosis in POEMS Syndrome: A Case Report

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### Abstract

### Case Report

Asteroid hyalosis is a rare degenerative vitreous disease and it is generally asymptomatic. Its etiology is still unknown. We report a case of asteroid hyalosis in a 59-year-old diabetic patient with peripheral neuropathy related to POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M component protein, skin changes). The aim of this work is to report the unusual association of asteroid hyalopathy with POEMS syndrome.

**Keyword:** Asteroid hyalosis, etiology, organomegaly, endocrinopathy.

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## INTRODUCTION

Asteroid hyalopathy is a rare degenerative vitreous disease. Its prevalence is between 0.2 and 0.9% [1, 2]. It is most often unilateral, and it is more commonly seen in patients with diabetes. The pathophysiology of asteroid hyalopathy is still not fully understood [1, 3]. It is considered as a secondary vitreous degeneration [1].

POEMS syndrome is a multisystemic condition characterized with constantly progressive and disabling neuropathy associated with monoclonal gammopathy in 82% of cases and numerous other systemic clinical manifestations [4]. Its prognosis is reserved, with a median overall survival of 35 months in absence of treatment [4].

The aim of this work is to report the unusual association of asteroid hyalopathy with POEMS syndrome.

## OBSERVATION

We report the case of a 59-years-old patient, known to be diabetic under treatment for 10 years, systemic hypertension under monotherapy for 2 years, and followed in neurology and internal medicine departments for a POEMS syndrome.

He presented myodesopsias in the right eye during his hospitalization in neurology service.

The ophthalmological examination found corrected visual acuity at 8/10 in the right eye and 10/10 in the left eye. Direct and consensual photomotor reflexes were present and symmetrical.

Biomicroscopic examination of the right eye revealed the presence of multiple spherical vitreous particles, shiny, whitish, and mobile with the movements of the vitreous fibers, leading to the diagnosis of an asteroid hyalopathy (Figure 1).

Funduscopy examination revealed slight papillary hyperaemia in the right eye and the presence of a few punctiform peripheral retinal hemorrhages in both eyes.

The neurological examination found sensory-motor neuropathy affecting all 4 limbs, making walking difficult with diminished osteo-tendinous reflexes. ENMG confirmed the presence of demyelinating sensorimotor polyneuropathy.

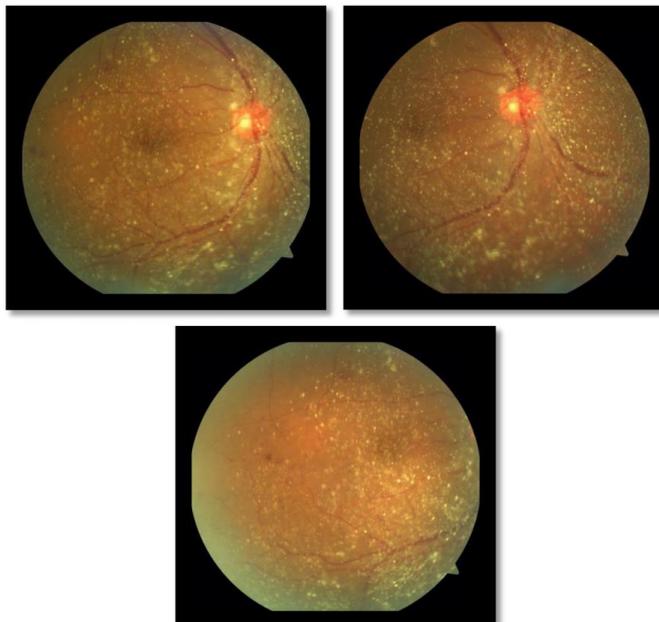
The general examination found anasarca (pleurisy, ascites, edema of the lower limbs), right atrioventricular hypertrophy with right bundle branch block objectified on the electrocardiogram and scleroderma-like skin (Figure 2).

Blood vascular endothelial growth factor (VEGF) was elevated and protein electrophoresis and

immuno-electrophoresis showed monoclonal IgA lambda gammopathy with 5% dyscrasic plasma cells.

All the clinical and paraclinical signs strongly suggested a POEMS syndrome. The patient received prednisone over a period of 9 months. The evolution

under treatment was marked by the recovery of walking observed from the 3rd month, the improvement of ENMG parameters with a reduction in the blood VEGF level. The aspect of asteroid hyalopathy did not change after treatment.



**Figure 1: Fundus photographs of asteroid hyalopathy in the right eye**



**Figure 2: Skin changes in POEMS syndrome**

## DISCUSSION

Asteroid hyalopathy is a rare, and it is considered to be a secondary, benign vitreous degeneration [1]. It is generally asymptomatic, unilateral and is often seen after the age of 60 and in diabetic patients [5]. Histologically, the crystalline particles are less than 0.1 mm in size, bound to collagen fibers [5], and are composed of calcium complexes rich in phospholipids [1, 6]. According to Streeten, these phospholipids diffuse from altered vessels, which can be found in systemic diseases such as diabetes, atherosclerosis and arterial hypertension [6]. Clinically, there is a divergence between the density of the deposits

and the functional impairment felt by the patient [1]. Visual acuity is often preserved and the diagnosis is most often incidental [1]. Biomicroscopic examination of the vitreous reveals the presence of whitish particles, shiny, mobile with the movements of the vitreous fibers unlike the sparkling synschisis characterized by mobile crystals independently of the movements of the vitreous [1, 5]. B-mode ultrasound shows significant hyperreflective elements, very mobile, with a hypochoic aspect of the peripheral vitreous (posterior shadow cone), which can be confused with a complete PVD [1]. Since asteroid hyalopathy is asymptomatic, it should not be treated [5]. However, in few cases, particularly in cases of associated macular pathology, vitrectomy may be indicated, but it remains tricky, because the vitreous is very rarely detached [1, 5].

Our patient had, in addition to diabetes and arterial hypertension, a multi-systemic condition that is the POEMS syndrome. POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M component protein, Skin changes) is a rare disease that was first described by Scheinker in 1938 [7]. It has also been known as Crow-Fukase syndrome, Takatsuki syndrome or osteosclerotic myeloma [7, 8]. Although the pathophysiology of POEMS syndrome is not fully understood, a chronic increase in pro-inflammatory cytokines has been demonstrated, including IL-1, IL-6, IL-12, TNF and growth factor vascular endothelial (VEGF), inducing angiogenesis and increased vascular

permeability, which could explain the diversity of clinical manifestations of the disease [7, 9]

Clinically, it is a progressive and very debilitating sensory-motor polyneuropathy that is life-threatening, most often associated with monoclonal lambda light chain gammopathy and numerous systemic manifestations: endocrine, cutaneous, organomegaly, anasarca, and others [4].

There are several ophthalmologic manifestations of POEMS syndrome. Papillary edema being the most frequent with an incidence between 29 and 64% [7, 10, 11]. It is usually bilateral, chronic and there is a risk for the vision to be compromised at an advanced stage but without reaching optic atrophy [7]. Ocular manifestations can also include central or peripheral visual field loss, diplopia, macular edema, serous retinal detachments, and neovascularization with increased choroidal thickness, which directly correlates with elevated VEGF levels [7, 10, 11].

Diagnostic criteria of POEMS syndrome were first described in 2003 by Dispenzieri and were recently updated in 2018 by Suichi [7, 12] who cataloged VEGF elevation as a major criteria thus improving sensitivity and specificity of the disease. On the other hand, the papilledema which was part of the minor criteria was recently removed (Table 1) [7].

Local ophthalmological management is based on intravitreal injections of anti-VEGF, triamcinolone or even vitrectomy [7]. The use of acetazolamide has also been reported to reduce vascular permeability. At the systemic level, monthly courses of chemotherapy with melphalan-prednisone should be initiated as soon as possible, given the high degree of progression and malignancy of the disease, and should be maintained over a prolonged period of 20 to 56 months [4]. Other treatments include azathioprine, interferon alpha, plasmapheresis and some authors even recommend autologous hematopoietic stem cell transplantation [7, 13].

Our patient presents 3 major criteria which are polyneuropathy; monoclonal IgA lambda gammopathy and VEGF elevation and 2 minor criteria namely anasarca and skin changes, thus confirming the diagnosis of POEMS syndrome.

## CONCLUSION

Literature review confirms the usual association of asteroid hyalopathy with diabetes, arterial hypertension and atherosclerosis, hence the need to measure fasting blood glucose and take blood pressure in these patients. To the best of our knowledge, this is the first case of association between asteroid hyalopathy and POEMS syndrome.

## REFERENCES

1. Genevois, O., & Lemoigne, O. (2015). Pathologie dégénérative du vitré. *EMC-Ophthalmologie*, 12(1), 1-7.
2. Feist, R. M., Morris, R. E., Witherspoon, C. D., Blair, N. P., Ticho, B. H., & White Jr, M. F. (1990). Vitrectomy in asteroid hyalosis. *Retina (Philadelphia, Pa.)*, 10(3), 173-177.
3. Bergren, R. L., Brown, G. C., & Duker, J. S. (1991). Prevalence and association of asteroid hyalosis with systemic diseases. *American journal of ophthalmology*, 111(3), 289-293.
4. Adams, D., Lozeron, P., Theaudin, M., Ribrag, V., Bourhis, J. H., & Lacroix, C. (2011). Nouveautés dans le diagnostic et le traitement des neuropathies périphériques liées à l'amylose AL et au syndrome POEMS. *Revue neurologique*, 167(1), 57-63.
5. Basseur, G., & Si Ahmed, K. (2006). Pathologie dégénérative du vitré. EMC (Elsevier SAS, Paris), *Ophthalmologie*, 21-248-A-25.
6. Streeten, B. W. (1982). Vitreous asteroid bodies: ultrastructural characteristics and composition. *Archives of Ophthalmology*, 100(6), 969-975.
7. Zapata, A. O., Cárdenas, P. L., Acuña, M. F., Álvarez, M. P., Zapata, J. O., & Imaicela, B. P. (2019). Manifestaciones neurooftalmológicas del síndrome de POEMS. *Archivos de la Sociedad Española de Oftalmología*, 94(10), 495-499.
8. Li, H., Huang, Y., Li, Y., Zheng, B., Cui, J., & Liu, M. (2019). Endocrine Manifestations in POEMS Syndrome: a case report and literature review. *BMC endocrine disorders*, 19(1), 1-7.
9. Cerri, F., Falzone, Y. M., Riva, N., & Quattrini, A. (2019). An update on the diagnosis and management of the polyneuropathy of POEMS syndrome. *Journal of Neurology*, 266(1), 258-267.
10. Barnés, C., Sánchez-Dalmau, B. F., Vela, D., & Alarcón, S. (2008). Papilledema as an indicator of POEMS syndrome. *Archivos de la Sociedad Espanola de Oftalmologia*, 83(12), 709-711.
11. Zhang, X., Cai, Q. Q., Huang, X. F., Cao, X. X., Cai, H., Zhou, D. B., ... & Li, J. (2017). Ocular manifestations and treatment outcomes in Chinese patients with POEMS syndrome. *Retina*, 37(9), 1784-1791.
12. Suichi, T., Misawa, S., Sato, Y., Beppu, M., Sakaida, E., Sekiguchi, Y., ... & Kuwabara, S. (2019). Proposal of new clinical diagnostic criteria for POEMS syndrome. *Journal of Neurology, Neurosurgery & Psychiatry*, 90(2), 133-137.
13. Kourelis, T. V., Buadi, F. K., Kumar, S. K., Gertz, M. A., Lacy, M. Q., Dingli, D., ... & Dispenzieri, A. (2016). Long-term outcome of patients with POEMS syndrome: an update of the Mayo Clinic experience. *American journal of hematology*, 91(6), 585-589.