



Temporal and spatial correlation between choroidal thickness and visual function in POEMS syndrome.

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Title page:

Manuscript title: **Temporal and spatial correlation between choroidal thickness and visual function in POEMS syndrome.**

Running title: **Choroidal thickness in POEMS syndrome.**

Keywords: POEMS syndrome, choroidal thickness, optic coherence tomography, disc swelling, multiple myeloma, vascular endothelial growth factor, VEGF

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Purpose

To report the spatial and temporal correlation of visual function with subfoveal choroidal thickness (SCT) in one patient with POEMS syndrome.

Methods:

Case report of one 53-year-old male patient diagnosed with POEMS syndrome.

Results:

Best corrected visual acuity was at the first visit 0.5 in his RE and 0.7 in his LE, and a mild defect was present in his RE visual field. CST was initially 356 microns in his RE and 263 microns in his LE. In the last examination, SCT was 284 in his RE and 222 in his LE, BCVA was 1,2 in both eyes and no defects were present in visual field.

Conclusion

SCT was inversely correlated in our patient with visual function in space and time. Due to the high sensitivity of choroidal tissue to vascular endothelial growth factor (VEGF), SCT is an excellent biometric marker to monitor the activity of POEMS syndrome.

INTRODUCTION

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome is a rare, multisystem disorder, typically presenting in the fifth or sixth decade. It usually occurs secondary to a plasma cell dyscrasia. Ocular manifestations are common in POEMS syndrome. Bilateral optic nerve swelling is the most common ocular

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3 feature of POEMS syndrome. In one of the major series it affected 52% of the patients,¹ and it
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5 is probably an independent prognostic factor.² Although frequently called papilloedema, it is
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7 not clear if it is a manifestation of increased intracranial pressure, nerve infiltration, ischemia
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9 or vascular hyperpermeability. Indeed the presence of edema elsewhere¹ and the response of
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11 some cases to antiVEGF agents,³ supports hyperpermeability as the main mechanism. Other
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13 reported ocular manifestations are cystoid macular edema,³ serous macular detachment,
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15 infiltrative orbitopathy,⁴ venous sinus thrombosis¹ and peripapillary choroidal
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17 neovascularization.^{5,6} More recently a few articles have reported increased choroidal thickness
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19 as a common manifestation of POEMS syndrome. This fact is probably the consequence of the
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21 high vascular endothelial growth factor (VEGF) levels present in these patients and the
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23 sensitivity of choroidal tissue to VEGF.
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28 MATERIALS AND METHODS

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30 In this article we report a patient with POEMS syndrome in which visual acuity and visual field
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32 were correlated with SCT.
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37 RESULTS

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39 A 53 year-old man, with unremarkable ocular history and previously diagnosed with POEMS
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41 syndrome was referred to our Department because of blurred vision on his RE. He had been
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43 diagnosed of this uncommon syndrome after an 8 month history of hepatomegalia,
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45 splenomegalia, polyneuropathy eritrodermia, astenia and loss of weight
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49 On the first examination, best corrected visual acuity (BCVA) was 0.5 in his right eye (RE) and
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51 0.7 in his left eye (LE). IOP was 11 mmHg in both eyes. Fundus demonstrated severely elevated
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53 and blurred optic discs(Figure 1). Visual fields showed an enlargement of the blind spot and
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55 inferior bundle defects in both eyes (Figure 2), which were more severe in his RE. Cranial
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57 magnetic resonance imaging and cranial computerized tomography were normal. Lumbar
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3 puncture demonstrated increased proteins with normal opening pressure. VEGF plasma levels
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5 at this stage were higher than 1000 ng/ml (normal<128.9 nm/ml).
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8 Optic coherence tomography (OCT), using Topcon 3D- 2000 revealed the presence of severe
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10 bilateral optic disc edema with hypoechoic cavitations. Quantification of the retinal nerve fiber
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12 layer (RNFL) thickness was very difficult due to the high volume of the optic disc head.(Figure
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14 3). Subfoveal choroidal thickness (SCT), measured with Spectralis OCT, using “Enhanced deep
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16 imaging” was 356 microns in his RE and 263 microns in his LE without increased macular
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18 thickness (Figure 3).
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21 Intravenous immunoglobulines and Melphalan-prednisone treatment were ineffective, so
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23 autologous peripheral blood stem cell transplantation was performed (APBSCT). The patient
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25 response to APBSCT was very good. Visual function recovery took place after chemotherapy.
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27 BCVA was 1,2 in both eyes 16 months after the initial visual examination. OCT demonstrated
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29 improvement in optic disc edema and significant reduction of choroidal thickness (290/222
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31 microns). (Figure 3) and VEGF was 131.4ng/ml (normal<128.9 nm/ml).
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38 DISCUSSION

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40 OCT imaging is highly reproducible. Some authors have proposed the use of macular or
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42 peripapillar retinal nerve fiber layer measurements to monitor the disease.⁷ Few publications
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44 have reported increased choroidal thickness in this syndrome. One series demonstrated that
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46 choroidal thickness was highly correlated with VEGF levels, and one recent case report,
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48 communicated significant reduction of both choroidal thickness and optic disc edema after
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50 treatment with thalidomide.^{8,9} The authors of this case report, suggested that this dramatic
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52 change in choroidal thickness was related to the reduction of VEGF. Indeed improvement in
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54 ocular manifestations of POEMS syndrome have been described after intravitreal antiVEGF
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56 administration.^{10,11}
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3 In our patient SCT was asymmetric (higher in the eye with worse visual function, Figure 3),
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5 suggesting that SCT is correlated with the severity of the syndrome and could be useful to
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7 monitor the course of the disease (Figure 4).
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10 It has been demonstrated that VEGF levels are highly correlated with choroidal thickness in
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12 POEMS syndrome.⁸ This correlation is easy to understand because VEGF secreted by the retinal
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14 pigment epithelium (RPE) has an important role in the maintenance of the choriocapillaris¹².
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16 Additionally, due to its spongy configuration, choroid volume is probably a very good index of
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18 the degree of organomegaly. Indeed correlation between choroidal thickness and VEGF levels
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20 have been demonstrated.⁸ In our patient, choroidal thickness measurements were easier to
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22 perform than RNFL measurements because, the enlargement of the optic disc made very
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24 difficult to determine the disc boundaries. In addition choroidal thickness is not affected by
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26 optic disc atrophy.
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30 Choroidal thickening has been previously described in patients with POEMS syndrome with
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32 ocular involvement. The spongy configuration of this vascular tissue and its sensitivity to VEGF
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34 levels, makes it a good biomarker of POEMS activity. In our patient it was highly correlated
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36 with the severity of ocular manifestations both in space and time and with the severity of
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38 systemic manifestations (it was greater in the most affected eye and it was reduced when the
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40 patient visual and general state improved).**state es el patrimonio, no se si queda un poco
41
42 raro** In summary in our patient SCT was correlated with ocular and systemic manifestations
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44 of POEMS syndrome and were more convenient than RNFL measurements.
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49 ACKNOWLEDGEMENTS

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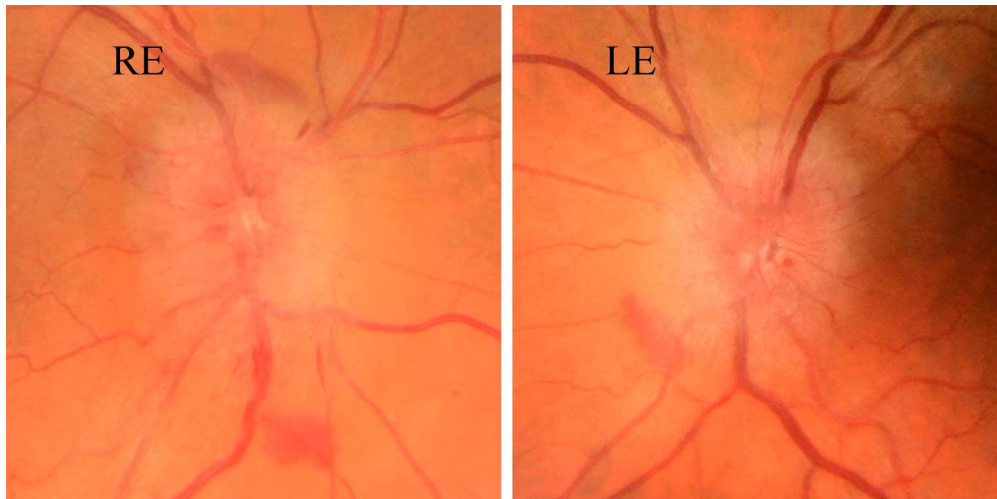
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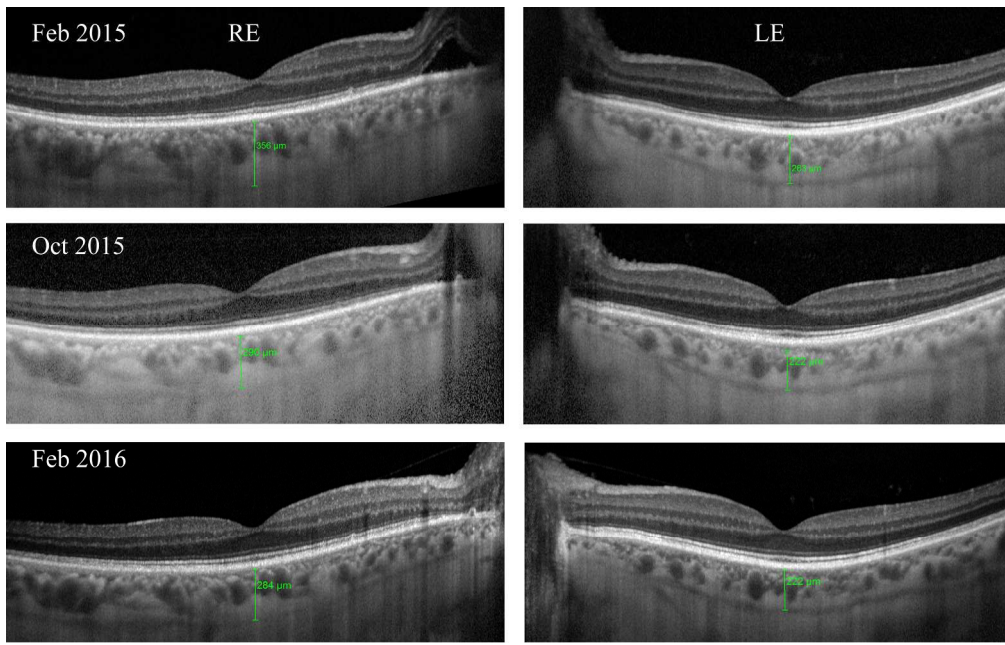
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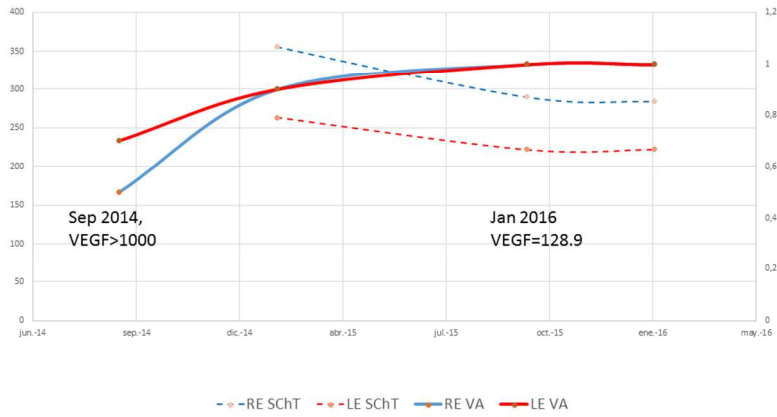


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Temporal correlation between subfoveal choroidal thickness and visual acuity



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