

**Title:**  
**POEMS syndrome and idiopathic portal hypertension: a possible association**

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DOI: 10.17235/reed.2017.4623/2016

Link: [PubMed \(Epub ahead of print\)](#)

Please cite this article as:  
Campos Sara, Agostinho Cláudia, Cipriano Maria Augusta. POEMS syndrome and idiopathic portal hypertension: a possible association. Rev Esp Enferm Dig 2017. doi: 10.17235/reed.2017.4623/2016.

Enero 2017	Volumen 109	Número 1	Páginas 1-86	CODE: REEDBN	ISSN: 1033-0108
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<b>Revista Española de Enfermedades Digestivas</b> <b>THE SPANISH JOURNAL OF GASTROENTEROLOGY</b> Acceso al texto completo en: <a href="http://www.reed.es">www.reed.es</a> o <a href="http://www.sedp.es">www.sedp.es</a> Factor de Impacto 12016: JCR: 1.455-4248 SCIR: 0.34-4203	
ORGANISMO OFICIAL DE: SOCIEDAD ESPAÑOLA DE PATOLOGÍA DIGESTIVA, SOCIEDAD ESPAÑOLA DE ENDOSCOPIA DIGESTIVA Y ASOCIACIÓN ESPAÑOLA DE ECOGRAFÍA DIGESTIVA	
	

  

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**POEMS syndrome and idiopathic portal hypertension: a possible association**

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**Key words:** Portal hypertension. Non-cirrhotic portal hypertension. Pro-inflammatory factors. Pro-thrombotic factors.

*Dear Editor,*

A 48-year-old female patient was admitted to the Emergency Department with upper gastrointestinal bleeding. Endoscopy showed large esophageal varices that were treated with band ligation. She had been treated with cyclophosphamide, melphalan, lenalidomide and corticosteroids for POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy or edema, M protein, skin changes). She had no other risk factors for chronic liver disease. Laboratory and radiological examinations could not confirm the etiology of portal hypertension. The liver biopsy suggested hepatportal sclerosis, compatible with idiopathic portal hypertension (IPH). Under a band ligation program, with beta-blocker, diuretics and prophylactic anticoagulation, the patient remains stable.

**Discussion**

In POEMS syndrome, a rare multi-systemic disease, the most frequent liver manifestation is hepatomegaly (1). To the best of the authors' knowledge, IPH has been reported only three times in POEMS syndrome (2-4), and this is the second case reporting the occurrence of histological findings compatible with IPH. As the precise etiopathogenesis of IPH is not fully

elucidated, and as it is an extremely uncommon manifestation in POEMS, it is not yet certain whether IPH is part of the syndrome or it develops independently. While some investigators believe that there is a congenital vascular anomaly involving the changes in the portal tract, the majority support an acquired vascular defect hypothesis (5). The acquired IPH hypothesis emphasizes a possible role of various pathogenic determinants: infections, prothrombotic states, prolonged exposure to several medications and toxins, immunological basis and progressive fibrosis of the portal veins (5). In this hypothesis, a hepatic circulation defect secondary to POEMS syndrome, which includes pro-inflammatory (VEGF, interleukin [IL]-6, IL-1 $\beta$ , tumor necrosis factor- $\alpha$ , tumor growth factor- $\beta$ ) and pro-thrombotic factors (aberrations in the thrombin-antithrombin complex) (1) which have been documented in the IPH and POEMS syndrome, may contribute to a possible association between these entities.

Although the association between these two entities is yet to be confirmed, the authors present this case in order to contribute to its elucidation.

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