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## Essential thrombocythemia as underlying cause of malabsorption syndrome

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**Abstract:** Patients with malabsorption syndrome frequently exhibit thrombocytosis associated with iron-deficiency anemia. Indeed, in these patients, the presence of underlying Ph-negative myeloproliferative neoplasms (MPNs) is rarely suspected. Here is reported the case of a young woman with persistent thrombocytosis and with malabsorption syndrome of unknown origin. In the hypothesis of a reactive thrombocytosis, patient received iron and folic acid supplementation without platelet count normalization. When patient developed a protein losing enteropathy was admitted to the hospital and specific haematological investigations allowed the diagnosis of underlying Essential Thrombocythemia. Considering the high thrombotic risk associated with MPNs, splanchnic vessel thrombosis was investigated by CT angiography, revealing the chronic occlusion of superior mesenteric artery. Interestingly, although MPNs are one of the most frequent causes of hepatic, portal or mesenteric vein thrombosis, they rarely involve mesenteric arteries. Indeed, this case underscores that, in patients with malabsorption syndrome of unknown origin, it is important to accurately exclude the primitive origin of haematological abnormalities before considering them as reactive manifestations.

## Essential thrombocythemia as underlying cause of malabsorption syndrome.

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Dear Editor,

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2 In October 2008 we observed a 31-year old woman with 2 year-history of postprandial abdominal  
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4 pain, diarrhea and peripheral oedema. Patient had no evidence of cardiac or renal failure, liver and  
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6 spleen size were normal and there was not palpable lymphadenomegalies. Liver, pancreatic and  
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8 thyroid function tests and acute phase reactants were normal, antigliadin, antiendomysial and anti  
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10 DNA antibodies were absent and antibodies to HCV, HBV and HIV were negative. Repeated  
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12 controls of hematological parameters evidenced slight thrombocytosis and leukocytosis, in the  
13  
14 presence of low serum ferritin. Moreover low serum protein and cholesterol, marked  
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16 hypoalbuminaemia and hypogammaglobulinaemia were present. No proteinuria was found and it  
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18 was hypothesized an enteric loss of protein. The stool specimens were negative for occult blood and  
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20 parasitic contamination. A total body CT scan evidenced no lymphadenomegalies, while a  
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22 generalized thickening of ileal wall with mesenteric edema was present. Furthermore, no alterations  
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24 of hepatic and portal veins were found. Gastrointestinal endoscopy showed jejunal mucosa covered  
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26 by congested, stocky and whitish villi and biopsies of duodenal and jejunal tract documented a  
27  
28 protein-losing enteropathy. Thrombocytosis was considered secondary to the iron deficiency and  
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30 patient received two week intravenous supplementation of iron and folic acid with recovery of  
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32 ferritin value. Because of thrombocytosis persisted, we decided to investigate the presence of  
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34 myeloproliferative neoplasms (MPNs). The patient was proved negative for *JAK2*<sup>V617F</sup> and  
35  
36 *MPL*<sup>W515K/L</sup> mutations and for *BCR/ABL* rearrangement, whilst endogenous erythroid colonies were  
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38 found. In addition, the HUMARA assay demonstrated monoclonal expansion of hematopoiesis.  
39  
40 Bone marrow biopsy evidenced megakaryocytic hyperplasia and the diagnosis of essential  
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42 thrombocythemia (ET) was made [1]. The high incidence of splanchnic thrombosis in patients with  
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44 MPNs prompted us to a more accurate investigation of abdominal vessels [2,3]. Actually, a CT  
45  
46 angiography revealed a significant stenosis of the superior mesenteric artery (Figure 1). The patient  
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48 underwent selective mesenteric angiography and percutaneous endovascular angioplasty, with  
49  
50 adequate revascularization. The presence of acquired or hereditary thrombophilic defects was ruled  
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1 out and therapy with low dose aspirin and hydroxycarbamide was undertaken. During the following  
2 months, the nutritive state and the clinical conditions of patient progressively improved.  
3

4 The most relevant feature affecting the clinical course of Ph-negative MPNs is the prothrombotic  
5 state causing both arterial and venous thromboses and microvessel disturbances [2,3]. In particular,  
6 splanchnic vein thromboses (SVT), including both portal and mesenteric vein thromboses, are  
7 frequent presenting complications of MPNs, whilst thrombosis involving the arterial mesenteric  
8 district are rarely reported [2,3]. In these cases, malabsorption syndrome might represent the  
9 unique clinic manifestation of a mesenteric arterial flow disturbance [4]. Patients are usually oligo-  
10 or asymptomatic for a long time, although the chronic intestinal ischemia causes a progressive  
11 malabsorption syndrome resulting in weight loss in almost all affected patients [4]. Since  
12 thrombocytosis is frequently observed during the course of malabsorption syndromes [5], an  
13 underlying MPN could remain misdiagnosed even in the presence of an overt hematological picture.  
14 Importantly, in contrast to SVT, which can be sensitively diagnosed by conventional CT scan or  
15 ultrasonography, the diagnosis of mesenteric arterial occlusion requires the CT angiography with  
16 evaluation of the arterial phase, or, as a gold standard, the angiography [4].  
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**Figure legend.**

**Figure 1.** CT volume rendering oblique reconstruction showing occlusion severe stenosis of proximal superior mesenteric artery (arrow) of the celiac trunk, with a compensatory circulation throughout the hypertrophic gastroduodenal artery (asterisks).

Figure  
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