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Letter to Editor

# Common Variable Immunodeficiency and Autoimmune Cytopenia

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## Letter to the Editor

The clinical expression of Primary Immuno Deficiencies (PID) is dominated by infectious complications. The frequency, severity or opportunistic nature of these infections most often alerts the clinician.

In fact, autoimmune manifestations are part of the clinical spectrum of many of these diseases and are dominated by autoimmune cytopenias, in particular, Immunologic Thrombocytopenic Purpura (ITP) and Auto Immune Hemolytic Anemias (AIHA).

We report the case of a young woman with Immunologic Thrombocytopenic Purpura (ITP) in a context of Common Variable Immuno Deficiency (CVID).

A 26-year-old woman, followed for CVID since the age of 7 years. She used to present recurrent infections. The biological assessment showed a decrease in two isotypes of IgG and IgA immunoglobulins. The diagnosis was made after rouling out other primary and secondary immunodeficiencies responsible for hypogammaglobinemia. The treatment consisted of monthly courses of intravenous immunoglobulins.

She was admitted to our department for cutaneo-mucous haemorrhagic syndrome. The anamnesis and clinical examination found an isolated purpura without anemic or tumoral or infectious syndrome. The blood count showed isolated thrombocytopenia, whose etiological assessment, allowed us to retain the diagnosis of ITP. Therapeutic management was based on corticosteroids for three months, and after two years, the patient did not experience a relapse of thrombocytopenia.

Autoimmune manifestations are now recognized as an important manifestation of several PIDs [1].Approximately one fifth of patients with CVID develop autoimmune cytopenias, dominated by ITP (7-20%), AIHA (2.8-6.4%), and autoimmune neutropenia [2,3]. These cytopenias occur in most cases before or at the time of CVID diagnosis.Generally, this association is seen in patients with "a particular phenotype" with fewer infectious manifestations and more autoimmune manifestations [1]. Mouillot et al. recently showed that patients with autoimmune cytopenia had more marked deficit in memory B cells and naive T4 cells, and a decrease in regulatory T cells [4]. These modifications are potentially responsible for a breakdown of mechanisms responsible for inducing and maintaining peripheral and central tolerance [5].

Therapeutic management is close to that of idiopathic autoimmune cytopenias, it is mainly based on corticosteroids, and immuno-modulating dose of intravenous immunoglobulins. However, the lack of specific data makes it impossible to recommend a formal diagram. Immunosuppresses are generally contraindicated in patients at risk of developing lymphoma and at high risk of infections. However, Azathioprine seems to have the best therapeutic index during CVID [1]. The place of anti-CD20 antibodies used during refractory cytopenia remains to be defined in such situations. Because of the increased risk of pneumococcal infections, splenectomy, should be avoided and probably reserved, as for other immunosuppressant's, at the fourth line of treatment. Thrombopoietin agonists, which have been shown to be effective in the management of idiopathic ITPs- although not specifically evaluated during the CVID- should be used early [1].

This case highlighted the isolated nature of ITPs in CVIDs. Blood count monitoring is necessary in case of any PID; and as autoimmune cytopenias frequently precede the diagnosis of CVID, the practice of electrophoresis of blood proteins with a weighted immunoglobulin assay must be performed at diagnosis.

## Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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