

A case of cryptogenic organizing pneumonia in a patient with idiopathic thrombocytopenic purpura

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ABSTRACT

Introduction: Cryptogenic organized pneumonia (COP) or bronchiolitis obliterans-organizing pneumonia (BOOP) is clinical condition characterized by interstitial lung disease with loss of functioning parenchyma due to inflammatory damage and pulmonary fibrosis. We report a case of COP related to autoimmune condition in patients with idiopathic thrombocytopenic purpura (ITP) and diabetes mellitus type 1. **Case Report:** A 46-year-old deaf and mute male was admitted to our hospital for general sickness, severe dyspnea. He had a history of ITP started 20 years before, previous splenectomy, smoking, systemic hypertension, diabetes mellitus type 1, glaucoma, previous admission for pulmonary thromboembolism. High resolution computed tomography (HRCT) found diffuse interstitial thickening with a bilateral ground-glass opacification, emphysematous areas, left-lower-lobe consolidation (apparently due to passive atelectasis because of left elevation of diaphragm and hiatal hernia), bundle-like thickening areas, a micronodule and clear reversed atoll sign. He was treated

with corticosteroids obtaining progressive improvement of thrombocytopenia and pulmonary distress. **Conclusion:** Association between ITP and COP or BOOP could be ascribed to autoimmune derangement. Respiratory symptoms and imaging in patients with ITP could suggest association with COP or BOOP. However, both conditions might ameliorate with corticosteroid treatment.

Keywords: Atoll sign, Bronchiolitis obliterans-organizing pneumonia, Cryptogenic organized pneumonia, Idiopathic thrombocytopenic purpura

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INTRODUCTION

Idiopathic thrombocytopenic purpura (ITP) is an autoimmune disease characterized by acquired thrombocytopenia due to destruction of platelets in the reticulo-endothelial system of spleen [1]. Idiopathic thrombocytopenic purpura is generally a benign and self-limiting condition in childhood, and only 20% of patients progress to chronic disease. Idiopathic thrombocytopenic purpura prognosis is determined by risk of spontaneous hemorrhage due to immune thrombocytopenia, especially in older adults.

Pathogenesis of ITP is related to T cell receptor activation against platelet glycoprotein, but the primary mechanism for the loss of tolerance against platelet remains unknown [1]. Moreover inflammatory conditions related to platelets release cytokines, recruitment of white blood cells, and activation of complement could involve different organs such as lungs [2]. Cryptogenic organized pneumonia (COP), also known as bronchiolitis obliterans-organizing pneumonia (BOOP), is an interstitial lung disease characterized by loss of functioning parenchyma resulting from inflammatory damage causing pulmonary fibrosis. The clinical features of COP or BOOP are usually non-specific and include influenza-like symptoms, associated with a restrictive spirometric pattern [3].

We report a case of pulmonary fibrosis and COP in a patient with ITP.

CASE REPORT

A 46-year-old deaf and mute male was admitted to our hospital for general sickness, severe dyspnea, drug-resistant vomiting and epigastric pain. He had a history of ITP started 20 years before, previous splenectomy, smoking, systemic hypertension, diabetes mellitus type 1, glaucoma, previous admission for pulmonary thromboembolism and left recurrent bronchopneumonic focus. His recent CT scan of chest was suggestive for an interstitial lung disease with a bilateral ground-glass opacification. His complete therapy included Revolade® (eltrombopag olamina, started some weeks before the admission in replacement of systemic corticosteroids), long-acting insulin, pantoprazole, ramipril, timolol maleate ophthalmic solution, and fondaparinux.

At the time of admission, an electrocardiogram was performed and it was negative for myocardial ischemia. A chest X-ray showed left lower lobe consolidation, homolateral diaphragm elevation and hiatal hernia (Figure 1A). Blood tests revealed increased white blood cells count and progressive thrombocytopenia. Endoscopic evaluation identified mycotic esophagitis and chronic gastritis. Diabetic gastroparesis was suspected and vomiting improved after the administration of prokinetic agents. High resolution computed tomography (HRCT) found diffuse interstitial thickening with a bilateral ground-glass opacification, emphysematous areas, left-lower-lobe consolidation (apparently due to passive atelectasis because of left elevation of diaphragm and hiatal hernia), bundle-like thickening areas, a micronodule (24x16 mm) and clear reversed atoll sign (Figure 1B).

Pneumologist interpreted lung disease as COP, however biopsy was not performed because of low platelet count (41,000 cells/ μ l). During the hospitalization, we resumed the previous therapy with corticosteroids obtaining progressive improvement of thrombocytopenia and pulmonary distress. The patient was discharged on corticosteroid therapy. A subsequent hematologic visit restored the therapy with Revolade® without major problems.

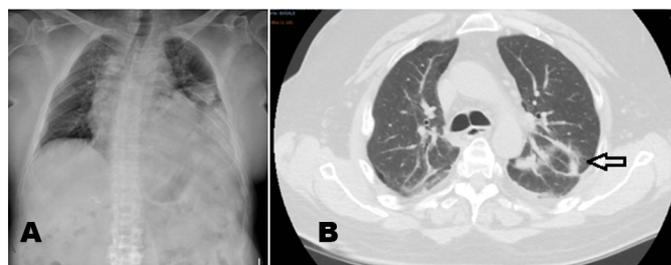


Figure 1: High resolution computed tomography of lung showing bilateral ground-glass opacification, a micronodule (24x16 mm), and reversed atoll sign.

DISCUSSION

In COP, chest radiograph could show bilateral patchy infiltrates in 68% of cases. On the other hand, CT scan reveals ring-shaped opacities surrounding an area of ground-glass opacification. Computed tomography abnormalities, defined as atoll sign, were first described by Voloudaki et al. in 1996 [4], and consisted in a ground-glass opacities with a circular consolidation pattern due to alveolar inflammation. Atoll sign has been considered a rare but highly suggestive sign of COP. Differential diagnosis takes into consideration different clinical conditions such as sarcoidosis, Wegener granulomatosis, infective pneumonias including tuberculosis and pulmonary paracoccidioidomycosis, collagen vascular diseases, bronchogenic neoplasms and non-specific interstitial pneumonia (NSIP) [5]. Moreover many of these clinical conditions have autoimmune pathogenesis that could be the cause of pulmonary lesions. The inflammatory pathogenesis of the COP or BOOP could be related to improved clinical conditions subsequent to corticosteroid treatment [6]. Moreover, a minority of patients need immunosuppressive therapy, such as rituximab or danazol that could be the cause of pulmonary lesions.

Association between ITP and COP or BOOP does not appear to be related to ITP treatment (Revolade®), but could be ascribed to autoimmune derangement suggested by association of ITP and diabetes mellitus type 1. Fontana et al. reported that ITP could be related to inflammation due to immune-mediated disorders and interstitial lung disease [2].

On the other hand, it should be underlined that COP or BOOP diagnosis is usually based on combination of HRCT findings and biopsy. Due to thrombocytopenia, biopsy was not performed in our case, therefore it could not be excluded that HRCT findings could also represent NSIP. However, both conditions respond to steroid treatment. Moreover, atoll sign, although highly specific, could be attributed to different clinical conditions as mentioned above, especially NSIP in this case.

CONCLUSION

Respiratory symptoms and imaging in patients with idiopathic thrombocytopenic purpura could suggest association with cryptogenic organized pneumonia or bronchiolitis obliterans-organizing pneumonia, however both conditions might ameliorate with corticosteroid treatment.

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Author Contributions

Alfredo De Giorgi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Marco Fiore – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Federico Moro – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Fabio Fabbian – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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