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Case Report

Hemoptysis Revealing Microscopic Polyangiitis: A Case Report

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Abstract

Microscopic polyangiitis is a rare systemic necrotizing vasculitis of unknown origin that affects small-caliber vessels. We report the case of a 42 years old némale patient who presented for 4 months several episodes of gradual onset, medium-abundance haemoptysis, associated with worsening dyspnoea to mMRC stage II and burning-type mid-thoracic pain, in a context of apyrexia and weight loss without quantification. The clinical examination was essentially normal, apart from some mucocutaneous pallor. Chest radiography revealed diffuse bilateral reticulo-micronodular infiltrates. Chest CT scan showed bilateral ground-glass opacities associated with reticulations giving a "crazy paving"appearance. Biological work-up revealed regenerative normocytic normochromic anemia. Flexible bronchoscopy revealed diffuse 3rd-degree inflammation of the entire bronchial tract, with hyperhemic mucosa and no visible bleeding. On bronchoalveolar lavage (BAL), the alveolar fluid was unrepresentative, showing significant siderophagy. Immunological tests revealed positive anti MPO pANCA. Given that our patient had a five factor score (FFS) prognostic score for vasculitis of 0, she received Prednisolone : 1 mg/kg/d for 1 month, followed by a gradual decrease associated to Aziathioprine to prevent the progression of the disease. The evolution was marked by the cessation of haemoptysis and regression of anaemia.

INTRODUCTION

Microscopic polyangiitis is a rare systemic necrotizing vasculitis of unknown origin that affects small-caliber vessels. We report the case of a patient presenting hemoptysis revealing microscopic polyangiitis.

CASE PRESENTATION

Mrs F.H aged 42, dyspneic on effort since 6 years, poorly followed for anemia with a history of blood transfusion of 2 globular pellet 6 years ago without documentation and without notion of externalized haemorrhage.

For the past 4 months, she had presented several episodes of gradual onset, medium-abundance haemoptysis, associated with worsening dyspnoea to stage II mMRC and burning-type mid-thoracic pain, in a context of apyrexia and weight loss without quantification.

The clinical examination was essentially normal, apart from some mucocutaneous pallor.

Chest radiography revealed diffuse bilateral reticulo-micronodular infiltrates (Figure 1).

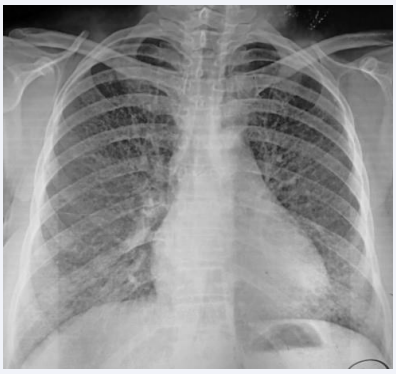

Figure 1 Diffuse bilateral reticulo-micronodular infiltrates.

Chest CT scan showed bilateral ground-glass opacities associated with reticulations giving a "crazy paving"appearance (Figures 2). Biological work-up revealed regenerative normocytic normochromic anemia. In view of the intra-alveolar haemorrhage, cardiac (ECG and cardiac ultrasound) and renal work-ups were carried out and were normal, with a preserved glomerular filtration rate (GFR).

Flexible bronchoscopy revealed diffuse 3rd-degree

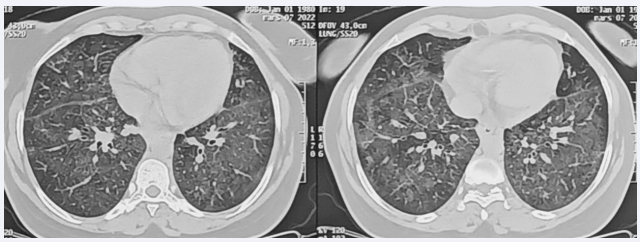

Figure 2 Bilateral ground-glass opacities associated with reticulations giving a crazy paving aspect.

inflammation of the entire bronchial tract, with hyperhemic mucous membrane and no visible bleeding; bronchial biopsies were unremarkable. On bronchoalveolar lavage (BAL), the alveolar fluid was unrepresentative, showing significant siderophagy; consequently, the Gold score could not be achieved. Immunological tests revealed positive anti MPO pANCA. Nasofibroscopy and nasal biopsy were without abnormalities. In view of these clinical and paraclinical findings, and after a multidisciplinary consultation, the diagnosis of microscopic polyangiitis was retained.

After concertation with the internists, given that our patient had a five factor score (FFS) prognostic score for vasculitis of 0, she was put on Prednisolone at the dosage of 1 mg/kg/d for 1 month, followed by a gradual decrease, associated to Azathioprine to prevent the progression of the disease.

The evolution was marked by the cessation of haemoptysis and regression of anaemia, and she is considered to have a good prognosis (FFS = 0).

DISCUSSION

Microscopic polyangiitis is a systemic necrotizing vasculitis primarily affecting small-calibre vessels. It is classified as an ANCA-associated vasculitis in the Chapell Hill Consensus Conference (1993) [1]. It affects Caucasians aged over 50 (85-100%), with a prevalence in France of 25 per million inhabitants, pulmonary involvement is found in 1/3 of cases [2]; there are no Moroccan epidemiological data.The pathophysiology is explained by inflammation mediated by anti MPO pANCAs, which activate neutrophils expressing MPO antigens, increase their membrane expression and lead to their degranulation, resulting in the release of free radicals and enzymes that act on endothelial cells, leading to necrotizing vasculitis lesions [3].

Pulmonary involvement is rarely inaugural, and manifests itself as the triad of intra-alveolar hemorrhage and hemoptysis:Hemoptysis, anemia and radiological pulmonary infiltrates [4]. Pneumo-renal syndrome is associated with extracapillary necrotizing glomerulonephritis of the kidneys [5].

Biologically, an inflammatory syndrome mA be observed, with increased C-reactive protein and sedimentation rate, or microcytic hypochromic anemia. ANCA assay is key, with pANCA found in 75-80% of patients, and anti-myeloperoxidase (anti-MPO) specificity in 60-75% of cases [4]. Flexible bronchoscopy reveals diffuse bleeding from distal sources, and bronchoalveolar lavage is performed, with cytology showing the presence of large numbers of red blood cells and siderophages, and calculation of the Golde score.

Good-prognosis forms (FFS = 0) are treated with corticosteroids alone, Immunosuppressants like Aziathioprine can be used to prevent progression of the disease, it is an effective, well-tolerated maintenance treatment. It induces fewer long-term side effects than cyclophosphamide [2].

Immunosuppressants are reserved for resistant forms, in case of relapse or worsening of the disease (FFS > 1). Today, however, some European protocols recommend the immediate combination of corticosteroids and immunosuppressants, whatever the type and severity of the disease [2,5]. Prognosis is assessed by the five-factor score (FFS) prognostic score for vasculitis detailed in Table 1 [6].

Table 1: Five Factors Score (FFS) 2009 revisited in 2011 [6]

1. Age > 65	+1 point
2. Renal failure (creatinine >150 mmol/l)	+1 point
3. Heart failure (clinical diagnosis)	+1 point
4. Severe digestive damage (hemorrhage, perforation, pancreatitis)	+1 point
5. ENT damage (clinical diagnosis confirmed on imaging)	–1 point

Overall survival is now 94% at 5 years, but relapses affect between a third and a half of patients [5].

The particularity of our observation is the fortuitous discovery of microscopic polyangiitis following the occurrence of hemoptysis.

CONCLUSION

Microscopic polyangiitis is a rare systemic necrotizing vasculitis of unknown origin, affecting small-calibre vessels. Management requires rigorous clinical, biological and imaging evaluation. Treatment depends on the prognostic score and relies mainly on corticosteroids and immunosuppressants.

Author's contributions

Hajar Arfaoui: Professor in charge and 1st author; Soukaina Hallouli: Attending physician and 2nd author; Salma Msika: Resident physician and 3rd author; Nabil Bougteb: Professor in charge ; Hasna Jabri: Professor in charge; Wiam El Khattabi: Professor in charge; Hicham Afif: Head teacher.

All authors contributed to this work. They have read and approved the final version of the manuscript.

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