Pulmonary hemosiderosis secondary to rheumatic heart disease – diagnostic challenges for a treatable condition

Introduction

Pulmonary hemosiderosis (PH) is defined as the clinical and functional consequence of iron overload of the lungs, which usually occurs due to recurrent intra-alveolar bleeding. It is a rare but life-threatening condition, that has been reported in up to 16% of adult patients with severe mitral stenosis (MS). Although the leading etiology of MS is rheumatic fever, PH secondary to MS occurs very rarely in pediatric patients with rheumatic heart disease. In addition, patients with PH may not present with the full triad of hemoptysis, iron deficiency anemia and diffuse pulmonary infiltrates. We report a case that illustrates the diagnostic challenges in PH secondary to rheumatic heart disease, which prompt recognition is essential for appropriate treatment.

Case Report

16 years old African origin

Past medical record
Rheumatic mitral valvulopathy anuloplasty at the age of 12

Asymptomatic until 5 months before presenting again (2 years after the first presentation)...

Second visit (18 years old)
Hemoptysis (5 months)
Progressive dyspnea (3 months)
Low-grade fever (1 month)
Pale and tachycardic (100bpm)
Systolic and diastolic murmurs (grade III/VI)

First visit to our Emergency Unit (16 years old):
Symptomatic iron-deficiency anemia (hemoglobin=6.6g/dL; ferritin=19μg/dL).
Treatment: red blood cell transfusion followed by iron supplementation with complete response.

Last reevaluation at our outpatient clinic 1 year later:
Asymptomatic. Hb=13.8g/dL; ferritin=120μg/dL.
Iron supplementation was withheld.
A reevaluation appointment was scheduled.
The patient was lost to follow-up...

2. Gastrintestinal etiology investigation
• Upper endoscopy: no signs of bleeding
• Meckel's diverticulum scintigraphy: negative
• t-transglutaminase antibodies: negative
• Fecal calprotectin ✓
• A course of albendazole and metronidazole

3. Infectious etiology investigation
• Hemocultures(3); Stool culture + O&P: negative
• Echocardiograms: no signs of endocarditis
• VDRL; Taso & Dnase; rose bengal test: negative
• Plasmodium: negative
• CMV/EBV/HAV/HBV/HCV/HIV+2: negative
• Tuberculosis
Mantoux: 20x23mm
Quantiferon®-TB: positive
Cultural tests (BAL): negative

1. Initial laboratory workup
• Hemoglobin = 10.3g/dL ↓
• Ferritin 30 μg/L ↓
• sTFR
• No signs of hemolysis
• Vitamin B12 and folate ✓

4. Cardiopulmonary etiology investigation
• Fiberoptic bronchoscopy: >95% hemosiderophages.
• Transthoracic echocardiography: severe mitral stenosis (valve area: 0.9cm²) and severe aortic regurgitation, with secondary pulmonary hypertension.

TREATMENT
• Oral corticosteroids were empirically started for pulmonary hemosiderosis.
• Surgical mitral and aortic valve replacement with prosthetic valves.
• At 8 months of follow-up after surgery, the patient remains without hemoptysis or anemia, free of glucocorticoids.

Conclusion

• PH secondary to mitral stenosis is usually reported in adults as a lifelong complication, rarely seen in Pediatrics.
• It may be successfully managed with valvuloplasty and/or valve replacement obviating the need for steroid therapy.
• A high index of suspicion is required for the early diagnosis of PH secondary to RHD. The diagnosis of secondary PH may be missed in patients with RHD who have an incomplete clinical presentation, namely iron-deficiency anemia and/or miliary nodular opacities on chest radiographs without overt hemoptysis.

References: