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GRANULOMATOSIS WITH POLYANGEITIS: A DIAGNOSTIC CHALLENGE IN THE PEDIATRIC AGE GROUP

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BACKGROUND

Granulomatosis with polyangiitis (GPA) is a severe systemic vasculitis, rare in childhood, that affects small to medium-size arteries in multiple organs, mainly upper and lower airways, kidneys and sinuses.

CASE REPORT

GFF, 13 years old, male, started at age of seven with left ocular proptosis, associated with ocular pain and hyperemia and diplopia. He underwent ocular biopsy that showed chronic inflammation with fibrosis, lymphoid hyperplasia and eosinophilia. Anti-neutrophil cytoplasmic antibodies (ANCAs) were negative. He remained without any systemic involvement and was followed up with the hypothesis of Immunoglobulin G4 related disease for six years. He has been treated initially with pulse therapy of methylprednisolone and azathioprine as maintenance therapy. Rituximab was indicated but it was not available. At age 13, he was admitted to the emergency room with massive epistaxis and hemoptysis, maintaining left ocular proptosis (Figure 1). Exams showed acute renal failure, with dysmorphic hematuria and nephrotic proteinuria, and hemodialysis was needed. He received pulse therapy with methylprednisolone followed by IV cyclophosphamide. Chest computed tomography showed ground glass opacities associated with diffuse smooth septal thickening tending to form consolidations in some regions, notably in the right lung (figure 2). Sinus computed tomography was unaltered. Renal biopsy was performed showing necrotizing crescentic glomerulonephritis, fibrocellular crescents in all glomeruli, multifocal interstitial nephritis and signs of moderate chronicity. Immunofluorescence showed pauci-immune pattern. ANCA performed by immunofluorescence and MPO and PR3 performed by Elisa were negative. Patient remained on hemodialysis, initiated plasmapheresis (seven sessions on alternate days), and oral cyclophosphamide (dosis of 1 mg/kg/day) with a good improvement of renal function.

CONCLUSION

We describe a case of GPA with a delayed diagnosis due to the wrong diagnosis of Immunoglobulin G4 related disease. The patient presented a severe course of the disease that had to be treated aggressively. Although the ANCA and MPO and PR3 antibodies were negative their presence is not a mandatory criterion, and clinical and radiological manifestations compatible with diagnosis were considered. Immunoglobulin G4 related disease is a very important differential diagnosis of GPA.