IMAGES IN RHEUMATOLOGY

Reversed halo sign: an uncommon presentation of pediatric eosinophilic granulomatosis with polyangiitis

Silva L[1], Jesus AR[2], Gata L[3], Ferreira S[4], Basso S[5], Estanqueiro P[6], Nascimento J[7], Salgado M[8]

CASE REPORT

A 12-year-old girl presented at the Emergency Department with dyspnea and a two-day pleuritic left chest stabbing pain, without fever. She reported weight loss (6% of total body weight) and recurrent back pain over the previous two months. Personal and familiar health history and physical examination were unremarkable. Blood analysis, aside eosinophilia (1060x10^9/liter), was normal (erythrocyte sedimentation rate - 10 mm/1st hour and C-reactive protein - 0,12 mg/dl). Chest radiograph showed a cavitary lung opacity in the left lung (Figure 1). A chest computed tomography (chest-CT) performed in the following day, confirmed the presence of two cavitary lung opacities, also known as reversed halo sign, in the left lung (Figure 2, 3 and 4).

Initial etiologic investigation was negative. Serologies for *Aspergillus fumigatus, Chlamydophila psitacci, Echinococcus* species and Human Immunodeficiency Virus were negative as were Interferon Gamma Release Assay, tuberculin test, SARS-CoV-2 protein chain reaction, antinuclear antibodies, anti-neutrophil cytoplasmic antibody (ANCA)-(anti-myeloperoxidase, anti-proteinase 3) and serum angiotensin-converting enzyme. She had an isolated elevation of immunoglobulin E (5509 UI/ml), with

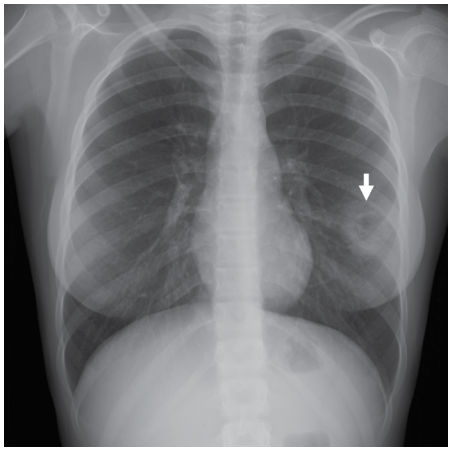

Figure 1. Chest radiograph (anterior posterior) showing reversed halo sign (arrow) in the left lung

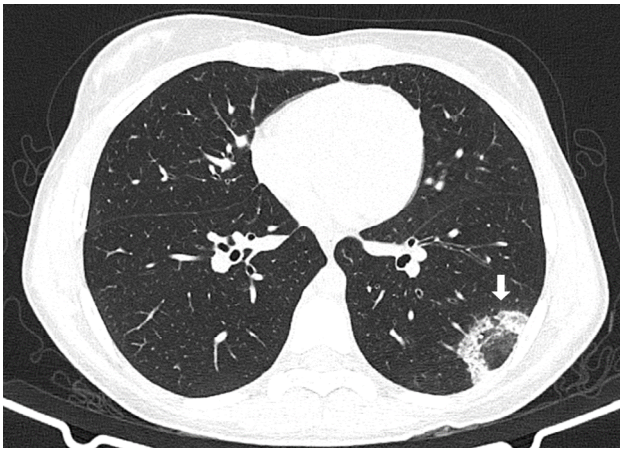

Figure 2. CT-scan (axial plane) showing a subpleural consolidation with a reversed halo sign image (ground glass center with a surrounding consolidation) in the left lower lobe

normal complement and lymphocyte subpopulation. Urinalysis was normal. An unrevealing abdominal ultrasound and bone scintigraphy were performed. Otorhinolaryngology evaluation revealed seasonal rhinitis. Non-steroidal anti-inflammatory drugs and paracetamol were initiated for pain management. No other treatment was administered.

One month after presentation, a chest-CT guided pulmonary biopsy was performed, showing micro-

[1] Pediatric Rheumatology Unit, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra - ORCID: https://orcid.org/0000-0002-0316-6167
[2] Emergency Department, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra - ORCID: https://orcid.org/0000-0002-2649-5430
[3] Emergency Department, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra
[4] Pneumology Unit, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra
[5] Imagiology Department, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra
[6] Pediatric Rheumatology Unit, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra - ORCID: https://orcid.org/0000-0003-2778-7643
[7] Pediatric Rheumatology Unit, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra - ORCID: https://orcid.org/0000-0002-8503-4466
8 Pediatric Rheumatology Unit, Hospital Pediátrico - Centro Hospitalar Universitário de Coimbra

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Correspondence to: Luana Silva
E-mail: luanamarisa@gmail.com

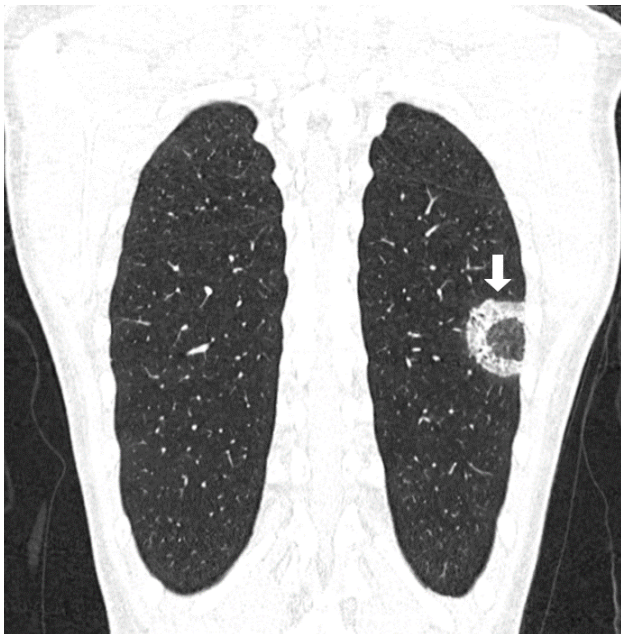

Figure 3. CT-scan (coronal plane) showing a subpleural consolidation with a reversed halo sign image (ground glass center with a surrounding consolidation) in the left lower lobe

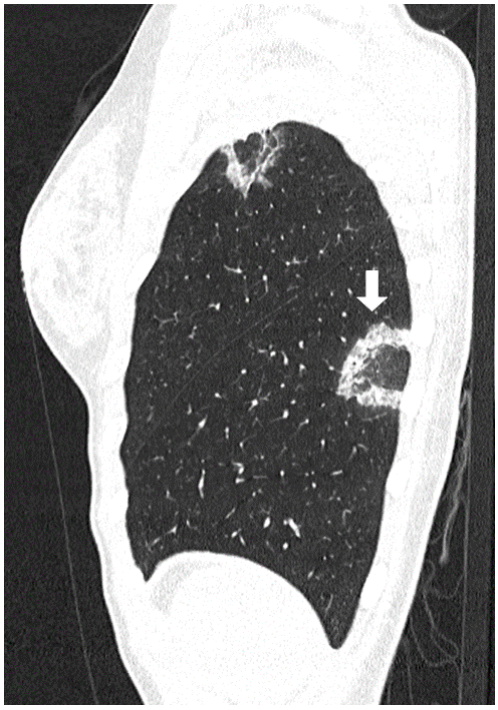

Figure 4. CT-scan (sagittal plane) showing two subpleural consolidation with a reversed halo sign image: 1 in the left superior lobe (31x20 mm) and 1 in the left lower lobe (45x26 mm)

scopic nodular lesions in capillaries and arterioles and intra-alveolar desquamation, with a predominance of macrophages and eosinophils, consistent with eosinophilic granulomatosis with polyangiitis (EGPA).

Cardiac evaluation and pulmonary function tests were normal. Oral prednisolone (0,5 mg/kg/day) and azathioprine (1 mg/kg/day) were initiated. She had transient chest pain at month two of treatment with worsening of eosinophilia (1.4x10^9/liter). At month six of treatment, she was asymptomatic, under low dose prednisolone (5 mg/48 hours) and azathioprine (1 mg/kg/day) and had a normal chest radiograph.

DISCUSSION

EGPA, also known as Churg-Strauss Syndrome, is an ANCA-associated vasculitis extremely rare in childhood. It presents with constitutional symptoms, asthma, peripheral eosinophilia and peripheral neuropathy[1]. Nodules, ground glass and reticular opacities have been described on chest-CT in children with pediatric EGPA[2]. ANCA are positive in 35-40%. Pediatric EGPA has an increased mortality rate, probably associated with higher incidence of cardiomyopathy.[1]

The 2022 American College of Rheumatology/Europe Alliance of Associations for Rheumatology classification criteria for EGPA (for patients with a previous diagnosis of small or medium vasculitis) includes clinical (obstructive airway disease, nasal polyps and mononeuritis multiplex) and laboratory and biopsy criteria (blood eosinophil count ≥1x10^9/liter, extravascular predominant inflammation on biopsy, positive ANCA antibodies and hematuria). A score of ≥6 is necessary for classification.[3] Our patient had ≥1x10^9/liter eosinophilia and extravascular inflammation, with a score of 7. Although evidence for management of pediatric EGPA is lacking, glucocorticoids plus azathioprine should be considered as initial therapy for active non-severe EGPA.[4]

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