SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

CHILDHOOD GRANULOMATOSIS WITH POLYANGIITIS: EXPERIENCE OF A TERTIARY REFERENCE CENTER IN CEARA, BRAZIL

Camila Emídio Bastos (Hospital Geral de Fortaleza, Fortaleza, CE, Brasil), Carlos Nobre Rabelo-Júnior (Hospital Geral de Fortaleza, Fortaleza, CE, Brasil), Míra Paula Vieira Cavalcante (Hospital Geral de Fortaleza, Fortaleza, CE, Brasil), Larissa Elias Pinho (Hospital Geral de Fortaleza, Fortaleza, CE, Brasil), Marco Felipe Castro Silva (Hospital Geral de Fortaleza, Fortaleza, CE, Brasil)

BACKGROUND

Granulomatosis with polyangiitis (GPA) is a form of necrotizing vasculitis that affects small and medium-sized vessels. It is a multisystem disease that may affect respiratory (upper, middle or lower) and urinary tract. Despite its rarity in the pediatric population, GPA can have devastating outcomes.

MATERIALS AND METHODS

Retrospective, descriptive study of patients aged <18 years diagnosed with GPA according to EULAR/PRINTO/PReS classification criteria during a five-year period (2014-2019) and regularly followed in a tertiary hospital of Ceará. Data were collected through medical charts review. Results are showed as frequency(%) or median(range).

RESULTS

Five patients fulfilled GPA criteria, four (80%) were female. The median age at diagnosis, follow-up duration and interval between first symptoms until diagnosis was 11.1 (9-12.3) years, 2.7 (0.7-4.4) years and 9 (1-22) months, respectively. At disease onset, all patients reported sinusitis and had abnormal urinalysis, with hematuria and nephrotic proteinuria. Other clinical features were arthalgia/arthritis (80%), epistaxis (60%), chronic otitis (40%) and hearing loss (20%). All patients required hospitalization at presentation and three required intensive care unit, mainly due to renal or respiratory failure. Two patients evolved with alveolar hemorrhage and one required orotracheal intubation and mechanical ventilation. There were no patients with subglottic, tracheal or bronchial stenosis. cANCA was positive in only 2 of them (40%). Computed tomography findings were fixed pulmonary infiltrates in three patients, cavitation in one and nodules in one. Renal biopsy showed minimal change in one patient, focal segmental necrotizing glomerulonephritis in two and rapidly progressive glomerulonephritis with crescent in two. Indirect immunofluorescence showed few or no immune deposits in all cases. Renal replacement therapy was necessary in two patients and one has already undergone transplantation. Methylprednisolone pulse and intravenous cyclophosphamide were given to all patients, but no one received plasmapheresis. Moreover, intravenous immunoglobulin and rituximab was required in three and two patients, respectively, due to renal activity persistence or worsening. Only two patients achieved remission.

CONCLUSION

Our findings are consistent with the literature, showing a high prevalence of the most common triad found in GPA (upper and lower respiratory tract associated with renal disease). The low positivity of the ANCA was probably due to our difficulty performing the test through ELISA. Methylprednisolone pulse and cyclophosphamide combined are in accordance with recently published guidelines. Low rates of remission highlights the severity of this disease, reinforcing the need for early recognition, aggressive initial treatment and long-term follow-up of these patients.