Eosinophilic Fasciitis with Visceral Involvement in the Pediatric Age

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Port J Pediatr 2022;53:620-1
DOI: https://doi.org/10.25754/pjp.2022.25589

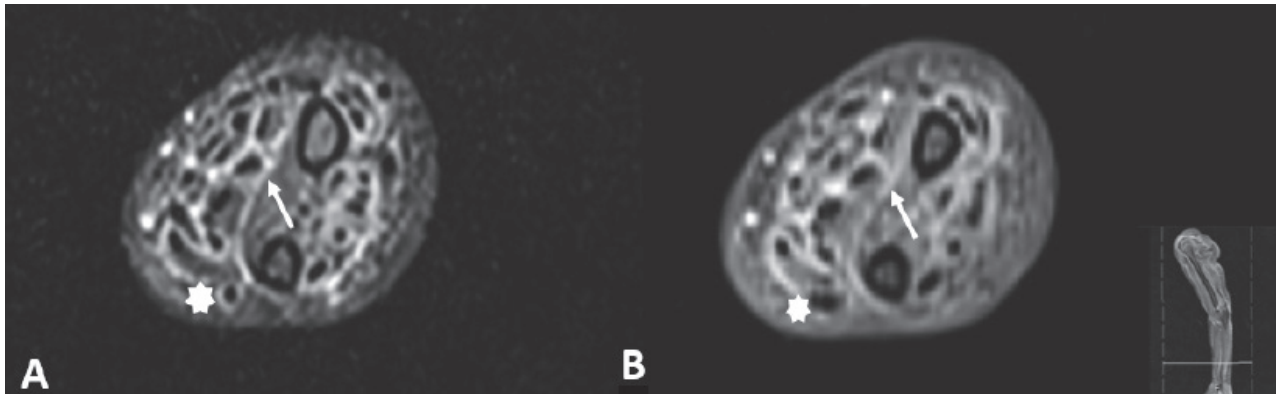
Eosinophilic fasciitis is a rare disease from the group of scleroderma-like connective tissue diseases with unclear etiopathogenesis.[1] The onset of eosinophilic fasciitis is often sudden, developing over a few days or weeks.[2] The main symptoms are symmetrical, full-circumference swelling, and plate-like hardness of the distal limbs which have also been observed extending to the proximal limbs. Systemic symptoms, such as fever, occur in many cases.[3] *En bloc* biopsies from the skin to the fascia show marked fascial thickening and inflammatory cell infiltrates.[3-5]

Most patients are in their fourth or fifth decade of life. However, pediatric cases have also been reported.[5] There have been isolated reports of systemic or visceral involvement.[6]

We report the case of a previously healthy 3-year-old girl who presented to her local hospital due to generalized pitting edema which had started in the lower limbs and progressed centripetally over one week, characterized by pain, stiffness, and hepatosplenomegaly. Her mother and maternal grandfather had vitiligo.

Laboratory workup showed hypereosinophilia (maximum 7200 cells/µL), microcytic anemia, hypofibrinogenemia, mildly increased troponins, and B-type natriuretic peptide, normal values of serum transaminases and muscle enzymes, hypoalbuminemia, and non-nephrotic proteinuria. Erythrocyte sedimentation rate was normal and C-reactive protein was mildly elevated. Immunoglobulin (Ig) G was increased, with positive antinuclear antibodies (1/640 homogeneous, DFS70 positive), negative myositis-specific antibodies, and normal complement levels. There was no evidence of hemolysis or abnormal cells in the blood film. The patient was transferred to a tertiary hospital for further investigations. Serologies showed IgM positivity for Epstein-Barr virus, confirmed by polymerase chain reaction, and all other microbiological examinations were negative. Results of echocardiography, electrocardiogram, cardiac magnetic resonance imaging, and thoraco-abdominopelvic computed tomography were normal. Bone marrow aspirate and liver biopsy showed only increased eosinophils. Left upper limb magnetic resonance imaging findings showed fasciitis, involving the superficial and deep fascial planes and to a lesser extent the muscle planes, which were more prominent in the forearm, consistent with the diagnosis of eosinophilic fasciitis (Fig. 1).

Changes were also evident along the chest and abdominal wall. Pending biopsy, oral corticosteroids were started, 1.5 mg/kg/day, with immediate laboratory improvement. *En bloc* forearm biopsy, performed a week later, showed mild



A - axial short-tau inversion recovery (STIR) image; B - T1 axial T1-weighted with fat-suppression post-contrast at the forearm level.

Figure 1. Magnetic resonance imaging demonstrates the involvement of the superficial and deep peripheral fasciae (asterisks) and diffuses deep intermuscular fasciae (arrows), which are thickened, with high signal intensity in A, enhancing after contrast administration in B. To a lesser extent, there is also signal change in the muscles and the subcutaneous fat.

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Received: 01/10/2021 | Accepted: 20/03/2022 | Published online: 01/07/2022 | Published: 01/07/2022

dermis fibrosis, subcutaneous tissue hyaline sclerosis with lymphohistiocytic infiltrate, and skeletal muscle with mild inflammatory infiltrates, and no eosinophils were seen.

Methotrexate was added (15 mg/m^2/week, subcutaneously). During early follow-up, limb swelling and pain subsided quickly, and skin thickening over the hands, forearms, forehead, feet, and trunk became increasingly apparent in the first months, with mild hand contracture. These features have been gradually improving since then. Raynaud phenomenon has been absent, and muscle strength is normal. Laboratorial changes and systemic manifestations have subsided. Vitiligo became apparent four months later.

After 15 months, the patient is still on the same methotrexate dose and low-dose steroids (0.14 mg/kg/day) and has normal growth and development. Only mild skin thickening on the forehead and hands is still present.

Therefore, eosinophilic fasciitis is a rare disease that presents diagnostic challenges and requires multidisciplinary management. The clinical presentation is very similar to other autoimmune diseases, namely scleroderma disorders, since they are manifested essentially by skin induration or fibrosis, mainly of the extremities.[7] Although fascial biopsy has classically been considered the gold standard for making a diagnosis of eosinophilic fasciitis, magnetic resonance imaging has been increasingly used for diagnosis, biopsy planning, and monitoring treatment responses. Systemic corticosteroids have remained the first-line treatment for eosinophilic fasciitis, and although it was not performed in our clinical case, a bolus of methylprednisolone, 0.5-1 g/day for three days, before the oral treatment with corticosteroids was recommended.[8] Methotrexate has emerged as the leading corticosteroid-sparing agent for eosinophilic fasciitis,[4,9] and more recently, therapeutic alternatives, such as rituximab and infliximab, are based on intravenous monoclonal antibody drugs.[10]

Keywords: Child, Preschool**;** Eosinophilia; Fasciitis/diagnosis; Fasciitis/diagnostic imaging; Fasciitis/drug therapy; Magnetic Resonance Imaging; Methotrexate/therapeutic use

Author Contribuitions

IC participated in acquisition of data. IC, MR and SD participated in the analysis or interpretation of data. IC participated in the drafting of the manuscript. MR, SD and IB participated in the critical revision of the manuscript. All authors approved the final manuscript and are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

Funding Sources

There were no external funding sources for the realization of this paper.

Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki 2013).

Provenance and peer review

Not commissioned; externally peer reviewed

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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