Juvenile Eosinophilic Fasciitis: A Single Center Cohort

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Background

- Eosinophilic fasciitis (EF) is a rare fibrosing disease that presents as painful swelling & progressive skin induration.
- Laboratory findings include elevated inflammatory markers, aldolase, eosinophils, and immunoglobulin G.
- EF diagnosis requires biopsy or magnetic resonance imaging (MRI) indicating fasciitis.
- Since first described in 1975, less than 30 pediatric cases have been described.

Purpose

- This juvenile EF case series describes the demographics, clinical findings, treatment, and outcomes for five patients.

Methods

- A retrospective chart review was performed for all patients diagnosed with EF at TCH between November 2011- 2021. Inclusion criteria included age at diagnosis < 18 years and EF confirmation by histology and MRI.

Conclusions

- Juvenile EF can present with swelling and progressive induration without skin abnormalities.
- Unlike adult cohorts, there were no underlying malignancies, significant hematologic abnormalities, or association with trauma.
- Previous juvenile EF cohorts have described systemic involvement (hepatosplenomegaly, lymphadenopathy), which was not present within this cohort.
- Although non-specific, the prayer sign could be a helpful finding to identify juvenile EF leading to early recognition and preventing long-term disabling outcomes.

Results

Table 1. Patient demographic, laboratory, and clinical characteristics.

Characteristics	Patient 1 (GW)	Patient 2 (DM)	Patient 3 (VV)	Patient 4 (HK)	Patient 5 (AS)
Sex	M	F	F	F	F
Race	White	White	White	White	White
Ethnicity	Non-Hispanic	Hispanic	Hispanic	Non-Hispanic	Non-Hispanic
Age at diagnosis (diagnosis year)	14 (2009)	4 (2012)	10 (2018)	12 (2021)	16 (2021)
Current age	27	13	13	13	17
Other medical conditions	Localized scleroderma	Localized scleroderma	Localized scleroderma	22q11.23 deletion	Hashimoto thyroiditis, atopy
Symptom onset to diagnosis	~ 1 year	~1 year	~6 mo	~1 year	< 1 mo
Groove sign	No	No	No	No	No
Peau d'orange	Yes	No	No	Yes	No
Prayer sign	Yes	Yes	Yes	Yes	Yes
Absolute eosinophil count (cells/uL)	**700**	**8206**	**1390**	**865**	**2130**
IgG (mg/dL)	**1990**	**1888**	**2100**	**2221**	1330
Aldolase (U/L) ref range: 3.3 - 9.7	7.1	**12**	**11.4**	**17.3**	**15.6**
ESR (mm/hr)	14	***25***	***75***	6	**21**
Corticosteroids	Yes	Yes	Yes	Yes	Yes
Methotrexate	Yes	Yes	Yes	Yes	Yes
Hydroxychloroquine	Yes	Yes	Yes	No	Yes
IVIG	No	Yes	Yes	No	Yes
Other	N/a	N/a	Rituximab Tocilizumab	Mycophenolate	N/a
Relapse	Morphea but not EF (2013, issues with non-compliance)	Last flare 2018	Yes, requiring increase medication	n/a	N/a
Remission	Yes, 2014	Yes, 2019	Yes, 5/2020 (MRI confirmed)	Yes, pending repeat MRI	10/2021

Figure 1. Positive prayer sign at initial presentation in juvenile EF. Examples of patients 3, 4, and 5 demonstrating a positive prayer sign at diagnosis. The prayer sign is due to skin induration and fascial fibrosis leading to joint contractures and tendon retraction.

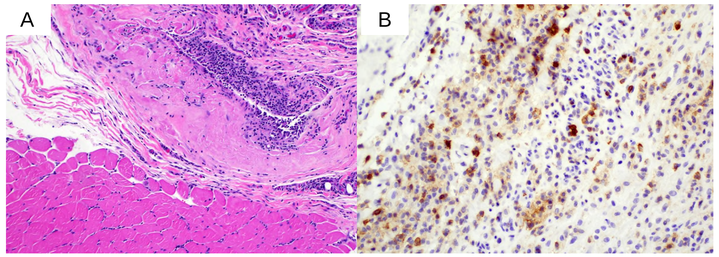

Figure 2. Patient 4 lower leg muscle biopsy. (A) Hematoxylin and eosin (H&E) section (magnification, x100) showing fascia with underlying muscle. The fascia shows thickening and inflammatory cell infiltration. (B) CD45 (leukocyte common antigen, LCA) immunohistochemical stain (magnification, x200) highlighting the inflammatory cells.

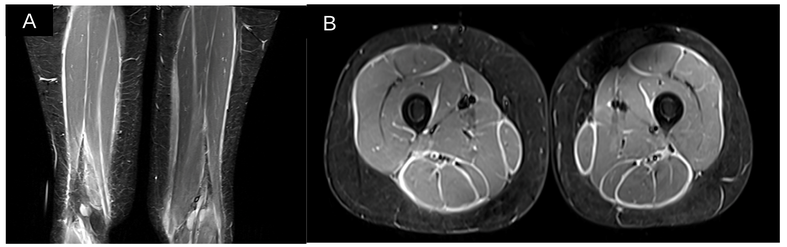

Figure 3. Patient 4 MRI thighs without contrast with extensive, symmetric fasciitis. (A) STIR coronal. (B) STIR axial.