JUVENILE EOSINOPHILIC FASCIITIS: A SINGLE CENTER COHORT

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Background: Eosinophilic fasciitis (EF) is a rare fibrosing disease. Since described in 1975, less than 30 pediatric cases have been reported. EF presents with painful swelling and progressive skin induration causing peau d' orange appearance and the groove sign. Laboratory findings include elevated inflammatory markers, aldolase, eosinophils, and immunoglobulin G. EF diagnosis requires biopsy or MRI indicating fasciitis. Risk factors for treatment resistance include concurrent morphea and pediatric age of onset. As triggers, presentation, treatment, and course often differ between pediatric and adult patients, it is important to further characterize juvenile EF cohorts. We report the demographics, pathology, MRIs, and clinical course for five patients with juvenile EF.

Materials/Methods: A retrospective chart review was performed for all patients diagnosed with EF at our institution between November 2011- 2021. Inclusion criteria included age at diagnosis < 18 years as well as EF confirmation by histology and MRI.

Results: For our juvenile EF cohort, the majority were female (80%) and non-Hispanic (60%) (Table 1). Age at diagnosis ranged from 4-16 years (median: 12) with associated follow-up ranging from 6 months to 12 years. Duration of symptoms prior to diagnosis ranged from 1 month to 1 year (median: 1 year). Associated medical conditions included localized scleroderma (60%), Hashimoto's thyroiditis, and 22q11 deletion. These patients presented with bilateral, progressive painful extremity swelling and severe joint limitation mostly without any positive skin findings. All patients presented with a positive prayer sign. Initial treatment included steroids and methotrexate. Other medications included hydroxychloroquine, immunoglobulin, mycophenolate mofetil, rituximab, and tocilizumab. Two patients had relapse of both morphea and fasciitis. Now, 80% are in remission with the remaining patient only 6 months into therapy with mycophenolate recently added.

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 clinical finding to identify juvenile EF leading to early recognition and preventing long-term disabling outcomes.

Images / Graph / Table

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Sex	M	F	F	F	F
Race	White	White	White	White	White
Ethnicity	Non-Hispanic	Hispanic	Hispanic	Non-Hispanic	Non-Hispanic
Age at diagnosis (year)	14 (2009)	4 (2012)	10 (2018)	12 (2021)	16 (2021)
Current age	26	13	13	13	16
Other medical conditions	Localized scleroderma	Localized scleroderma	Localized scleroderma	22q11.23 deletion	Hashimoto thyroiditis
Duration of symptoms prior to diagnosis	~ 1 year	~ 1 year	~6 months	~1 year	< 1 month
Diagnosis Labs					
Absolute eosinophil count (cells/uL)	**700**	**8206**	**1390**	**865**	**2130**
IgG (mg/dL)	**1990**	**1888**	**2100**	**2221**	1330
Aldolase (U/L)	7.1	**12**	**11.4**	**17.3**	**15.6**
ESR (mm/hr)	14	**25**	**75**	6	**21**