Fracture risk in adult versus pediatric patients with linear morphea of the extremity: A single center retrospective study

Linear morphea of the extremity (LME) is a rare sclerosing disease of skin and underlying tissues, associated with musculoskeletal complications.[1] However, there are limited studies investigating fracture risk and bone density.[2] We hypothesize that LME-affected limbs may be at an increased risk of localized fracture, which may be compounded over time by factors including age and cumulative steroid exposure. This retrospective single center study compares fracture risk in LME-affected limbs to unaffected limbs in adult and pediatric patients.

This Institutional Review Board-approved retrospective chart review included adult and pediatric LME patients seen at the University of California, San Francisco between January 2015 and December 2022. Medical records of LME patients were reviewed for limb fracture history. Limb fractures were labeled as "cases," with the unfractured contralateral limb (either upper or lower extremity) from the same patient, designated as the "control." The fractured (case) and nonfractured (control) limbs were evaluated for LME and secondary fracture causes. Fisher exact testing was performed ($P < .05$ considered significant).

Twenty-seven adults and 21 pediatric LME patient records were identified. Five adults and 2 pediatric cases of fracture were included. Among adult cases, 3 (60%) received systemic steroids with a median latency between LME onset and fracture of 14 years (Table I). Additionally, all 5 (100%) adult fractures involved a LME-affected limb, of which 3 (60%) were atraumatic (Table II). One DEXA scan was available, demonstrating lower bone density in the adult patient's LME-affected limb (Z-score: −1.2) than unaffected limb (Z-score: −0.2). All 3 cases of osteopenia, identified through plain film, involved an LME-affected limb. Bony vascular occlusion was identified on magnetic resonance imaging in the LME-affected limb of one patient.

Among pediatric patients, 2 (100%) received systemic steroids. Median latency between disease onset and fracture was 2 years (Table I). All limb

Table I. Patient demographics, as per chart review

Characteristics	Adults* No. (%)	Pediatric No. (%)
No.	5	2
Patient-reported race†		
Native Hawaiian or Pacific Islander	1 (20)	0 (0)
White	2 (40)	1 (50)
Other‡	2 (40)	1 (50)
Patient-reported ethnicity		
Non-Hispanic or Latino	3 (60)	2 (100)
Hispanic or Latino	2 (40)	0 (0)
Patient-reported gender		
Female	5 (100)	2 (100)
Male	0 (100)	0 (0)
Age of LME onset, median, y	14.0	8.5
Age at time of fracture, median, y	36.0	10.5
Latency between LME onset and fracture, median, y§	14.0	2.0
History of LME crossing joints	3 (60)	2 (100)
Personal history of osteopenia‖	3 (60)	0 (0.0)
Personal history of bony vascular occlusion¶	1 (20)#	0 (0)
Personal history of other metabolic bone disease**	0 (0)	0 (0)
History of physical therapy	2 (40)	1 (50)
Functional impairments related to LME		
History of impaired joint mobility/ contracture	3 (60)	1 (50)
History of global limb atrophy	3 (60)	1 (50)
History of gait disturbance††	2 (40)	1 (50)
History of systemic steroid use	3 (60)	2 (100)
Consecutive duration of systemic steroid use in months, median	1	5
History of other medications that alter bone metabolism‡‡	0 (0)	0 (0)
History of DEXA scan	1 (20)	0 (0)

LME, Linear morphea of the extremity.
*Adult patients included patients (>18 years) and patients with pediatric onset LME transitioning to adult care.
†Only categories relevant to the study were included.
‡The "other" category is not further specified in chart review.
§The median values were calculated using the difference between the age of morphea onset and fracture in year for each patient.
‖In patients who were found to have osteopenia, method of identification was recorded. All 3 patients were noted to have osteopenia via plain films.
¶History of bony vascular occlusion includes history of avascular necrosis, osteonecrosis, and fibrosis.
#Limb was affected by osteonecrosis and avascular necrosis.
**History of other metabolic bone diseases includes osteoporosis, parathyroid disease, chronic kidney disease, and/or celiac disease.
††Cases of gait disturbance were recorded in patients with linear morphea involving the lower extremity only.
‡‡Medications that alter bone metabolism include bisphosphonates, RANK-L inhibitors, parathyroid hormone analog, selective estrogen receptor modulators (SERM), and sclerostin inhibitors.

Table II. Profiles of fractures* and secondary outcomes in adults and pediatric patients with linear morphea of the extremity

Characteristics	Fractures in adult patients *N* (%)	Fractures in pediatric patients *N* (%)	*P* value
No.	5	2	
Disease status			.05
Morphea-affected	5 (100)	0 (0)	
Unaffected	0 (0)	2 (100)	
Age at time of fracture, median, y	36	10.5	NA
Anatomic location			>.99
Upper extremity	1 (20)	1 (50)	
Lower extremity	4 (80)	1 (50)	
Personal history of osteopenia	3 (60)	0 (0)	.16
Personal history of bony vascular occlusion	1 (20)	0 (0)	.35
Fracture type			.095
Atraumatic	3 (60)	0 (0)	
Traumatic	0 (0)	2 (100)	
Not specified	2 (40)	0 (0)	
Systemic steroid exposure	3 (60)	2 (100)	.57
Pulse steroid exposure	1 (20)	2 (100)	.15

*For patients with multiple fractures, the first fracture was recorded.

fractures occurred in unaffected limbs and were traumatic [2 (100%)] (Table II). There were no pediatric patients with known osteopenia, bony vascular occlusion, or available DEXA scans.

To our knowledge, this is the first study to compare extremity fracture risk in adult and pediatric patients with LME. Overall, our study observed a trend towards (1) atraumatic fracture, lower bone density, and vascular occlusion in LME-affected limbs in adults, (2) traumatic fractures in pediatric patients, and (3) increased disease latency in adults. These trends of ipsilateral atraumatic fracture and bone injury observed in LME-affected limbs in adults may suggest an underlying morphea-related risk to bone structure and integrity that evolves longitudinally over time. Furthermore, functional impairments may predispose LME-affected limbs to disuse atrophy which, when combined with systemic steroid exposure, may exacerbate overall risk of localized fracture and bone loss.[3-5] Altogether, morphea-related structural changes, functional asymmetry, and medication exposures may disrupt the bone remodeling cycle, leading to accrued injury over time and ultimately culminating in localized bone loss. Our sample size remains small, and laterality associations between fracture and LME did not attain statistical significance. While we observed distinctive trends in our pediatric patients, larger studies are needed, particularly to determine if early intervention should be considered to reduce risk of adult complications.

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Patient consent: In accordance with the institutional review board, the authors obtained written consent from patients for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available. Patient consent forms were not provided to the journal but are retained by the authors.

IRB approval status: This study was conducted with IRB approval.

Key words: adults; bone; bone density; fracture; linear morphea; linear morphea of limb; linear morphea of the extremity; morphea; orthopedic; pediatrics; skeletal.

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Conflicts of interest

Dr Haemel is a consultant to CSL Behring and an investigator for Priovant and AstraZeneca. Drs Fan, Cordoro, and Authors Kazmi, Obiakor, and Jacobson have no conflicts of interest to declare.

REFERENCES

1. Schoch JJ, Schoch BS, Werthel JD, McIntosh AL, Davis DMR. Orthopedic complications of linear morphea: implications for early interdisciplinary care. *Pediatr Dermatol*. 2018;35(1):43-46. https://doi.org/10.1111/pde.13336
2. Kazmi M, Shaw K, Fan W, Obiakor B, Vleugels RA, Haemel A. Fracture risk in linear morphea-affected limbs: a pilot retrospective study of adult patients. *JAAD Int*. 2024;18:27-29. https://doi.org/10.1016/j.jdin.2024.08.019
3. Epstein S, Inzerillo AM, Caminis J, Zaidi M. Disorders associated with acute rapid and severe bone loss. *J Bone Miner Res*. 2003;18(12):2083-2094. https://doi.org/10.1359/jbmr.2003.18.12.2083
4. Weinstein RS, Jilka RL, Parfitt AM, Manolagas SC. Inhibition of osteoblastogenesis and promotion of apoptosis of osteoblasts and osteocytes by glucocorticoids. Potential mechanisms of their deleterious effects on bone. *J Clin Investig*. 1998;102(2):274-282. https://doi.org/10.1172/JCI2799
5. Takata S, Yasui N. Disuse osteoporosis. *J Med Investig*. 2001;48(3-4):147-156.

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