

Tissue, Time, and Teamwork: Rethinking Perioperative Care in Cutis Laxa

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Background and Purpose

- Cutis laxa is a rare connective tissue disorder marked by loose, sagging skin with reduced elasticity due to abnormalities in elastic fiber formation. Fewer than 200 cases have been documented in the literature, underscoring its clinical rarity. Beyond cutaneous findings, the disorder may involve multiple internal organ systems, creating complex perioperative considerations. This case highlights the need for coordinated, interdisciplinary, patient-centered planning to safely manage individuals with rare connective tissue disorders.

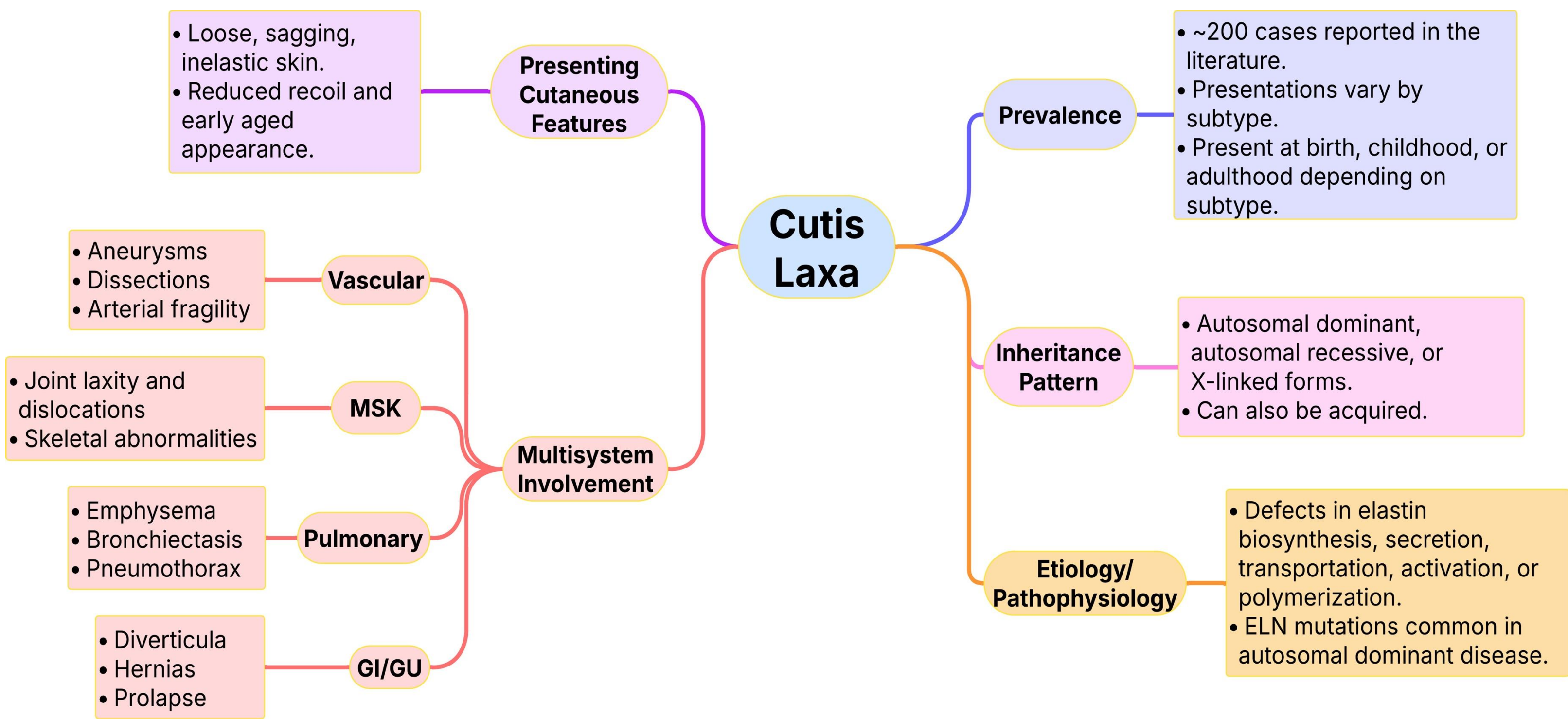
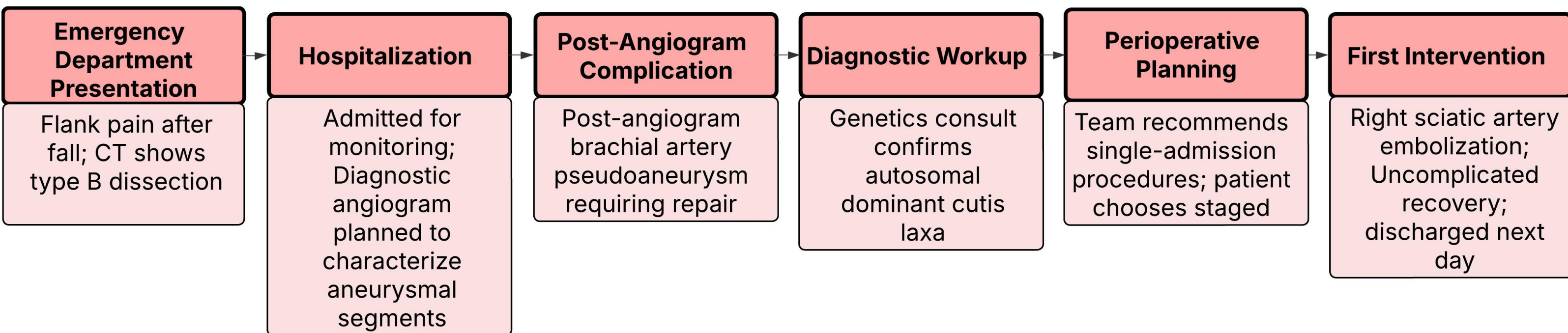


Figure 1: Diagram summarizing key characteristics of cutis laxa - including prevalence, inheritance patterns, etiology/pathophysiology, cutaneous features, and multisystem involvement.

Presentation

- A 56-year-old man with a known past medical history of asthma presented to the emergency department with a 3-day history of right flank pain and new-onset hematuria following a fall from a horse. His physical examination was notable only for focal right lumbar tenderness without cardiopulmonary or neurologic abnormalities. CT abdomen and pelvis with IV contrast revealed a type B aortic dissection originating below the renal arteries with extension into the bilateral common, external, and internal iliac arteries, along with bilateral persistent sciatic artery aneurysmal dilation measuring 15 mm. These findings prompted ICU admission for close hemodynamic monitoring, impulse control therapy, and further vascular evaluation.

Clinical Course



- A diagnostic angiogram was performed via left brachial access in preparation to exclude aneurysmal arterial segments to reduce the risk of aneurysmal rupture.
 - Postoperatively, a left brachial artery pseudoaneurysm developed, which required emergent surgical intervention.
- Genetic consult was obtained, and evaluation confirmed an ELN c.484G>A (p.Gly162Ser), heterozygous gene mutation associated with cutis laxa.
- Given the patient's connective tissue disorder and elevated procedural risks, a multidisciplinary team - including vascular surgery, vascular anesthesiology, perioperative medicine and surgical ICU - recommended consolidating interventions in one admission to address their aneurysmal arterial segments.
 - However, incorporating patient shared decision making, the patient preferred a staged approach due to treatment-related concerns.
- A subsequent right sciatic artery aneurysm repair was performed using coil embolization and Amplatzer plug placement.
 - Procedure was uncomplicated and had a satisfactory outcome.
 - Patient was discharged the following day on ASA, statin, beta blocker, and anticoagulant, with vascular surgery and primary care follow-up.

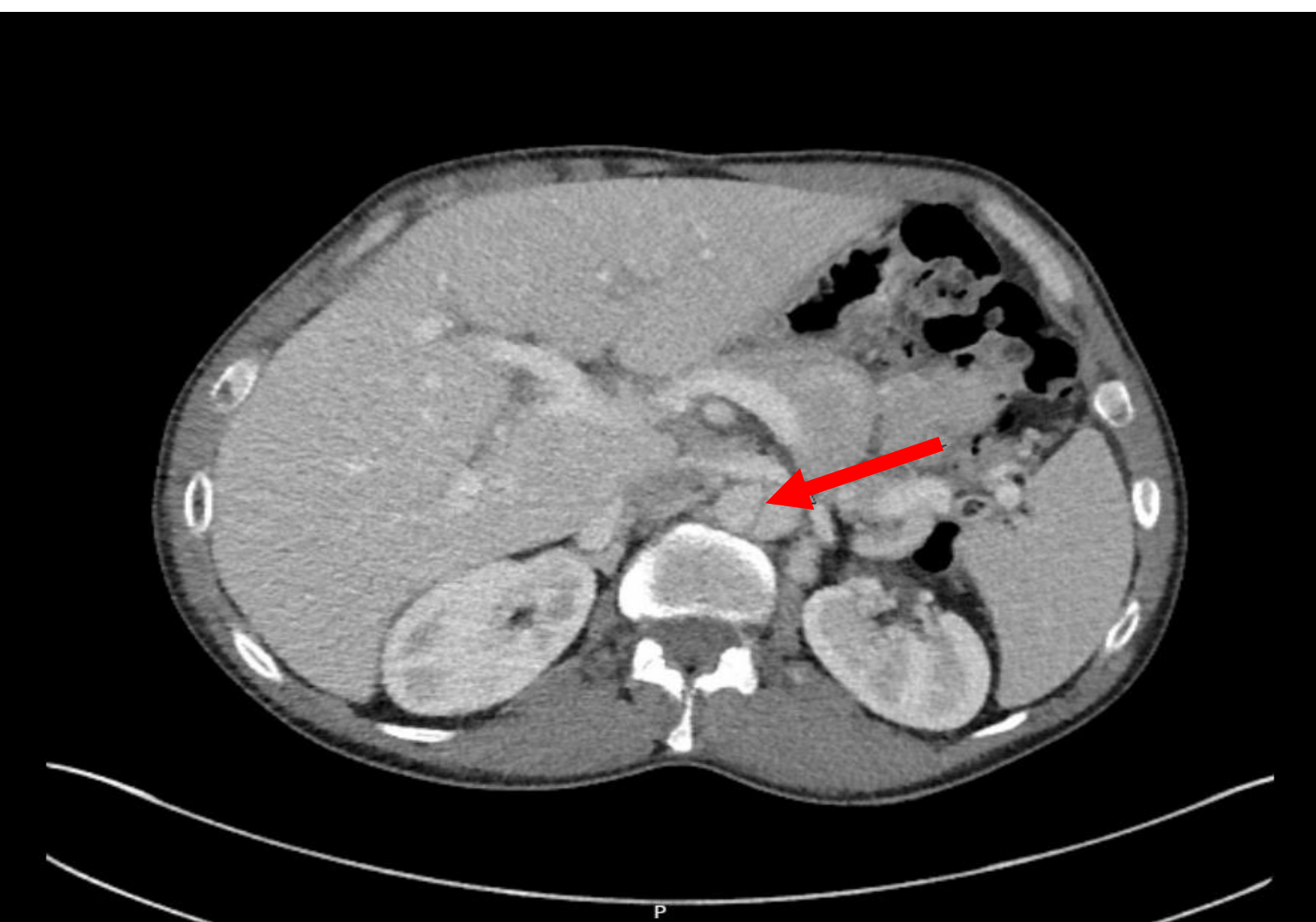


Figure 2: CT Abdomen and Pelvis with IV Contrast (axial view) demonstrating the dissection (arrow) beginning at the level of the left main renal artery.

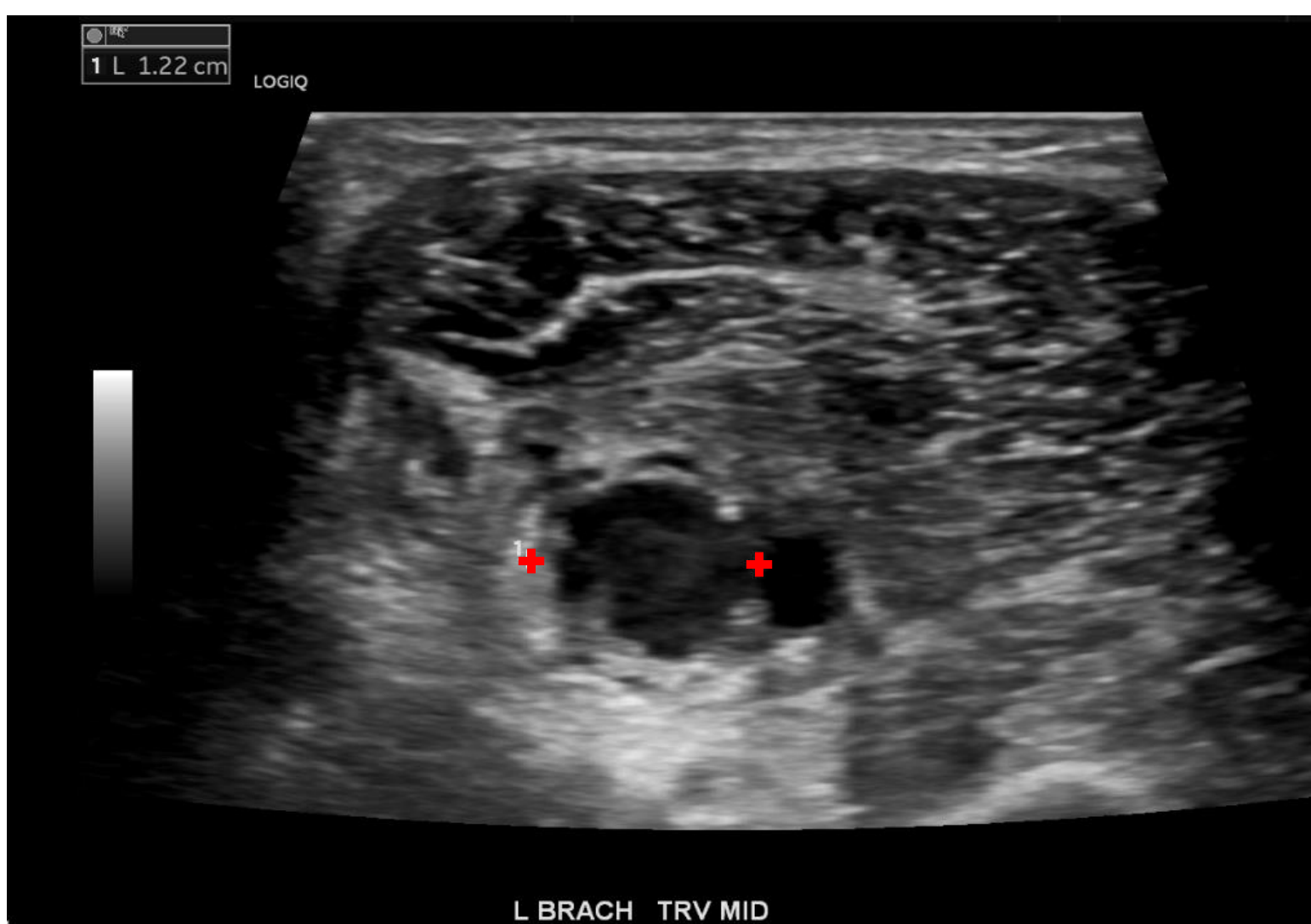


Figure 3: Cross-sectional ultrasound of the left brachial artery pseudoaneurysm; the plus signs represent the diameter of the pseudoaneurysm.

Discussion

- The coexistence of a connective tissue disorder and multiple aneurysms presented a complex perioperative risk profile requiring careful multidisciplinary coordination.
- Recognizing that this patient was at heightened risk for arterial complications, due to connective tissue fragility, the care team altered the perioperative approach for a subsequent procedure.
 - To avoid the recurrence of access-related vascular injury, arterial access was deliberately avoided, unless it became clinically indicated intraoperatively, during a right sciatic artery aneurysm repair.
- The patient's procedural plan was staged after a broad biopsychosocial, multidisciplinary assessment of risks and benefits, his support system, and social needs/responsibilities.
- This complex case highlights how individualized, risk-adapted perioperative strategies for patients with complex connective tissue disorders lead to success and patient satisfaction.
- **Cutis Laxa Management**
 - No specific treatments that prevent the progression or cure cutis laxa currently exist.
 - Skin manifestations can be improved via plastic surgery.
 - When internal organs are affected, a collaborative, interprofessional approach to comprehensive management of the condition is needed.

Acknowledgements

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References



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