Six year old male with abnormality noticed by parents

Donald Imwalle
Congenital Elevation of the Scapula

• AKA Sprengel’s deformity
  – Described by Sprengel way back in 1891
• Congenital elevation and deformity of the scapula, almost always with other congenital anomalies
• Most common congenital shoulder anomaly
Features

- Scapular winging
- Scapular elevation (kinda sorta)
- Hypoplasia of the scapula
- Omovertebral connection
  - Bony and/or fibrous connection between the cervical spine and superomedial angle of scapula
Embryology

• Scapula appears normally at 5\textsuperscript{th} week as a mesenchymal mass level with C4 and C5
• From the 6\textsuperscript{th} through the 12\textsuperscript{th} week, it migrates inferiorly
  – Normally inferior angle between T6 and T8
• During the same time, morphology changes
  – Becomes taller than wide
Pathophysiology

- Clearly interruption of the descent is the problem
- But the why is not well known
  - Possibly interruption of vascular supply?
  - Genetic mutation? (Some forms are familial)
- Main thing: Scapula is closely associated with the C-spine, so SD is often associated with vertebral malformations
Associated Malformations

• Klippel-Feil Syndrome
• Spina bifida
• Kyphoscoliosis
• Torticollis
• Diastematomyelia
• Underdevelopment of clavicle or humerus
Clinical Presentation

- Usually present at birth or very early
- More commonly girls than boys (3:1)
- Patients may have noticeable disfigurement and functional deficits
  - High riding medially rotated scapula
  - Loss of shoulder abduction
  - Loss of forward flexion
  - Scapula is misshapen, being as wide as it is tall
- Usually unilateral
Radiology Evaluation

• X-ray
  – Initial screening
  – Evaluate degree of elevation
  – Evaluate for a bony omovertebral bar

Niknejad, M. Radiopaedia.org, rID: 20941
Radiology Evaluation

- **CT**
  - Exact bony morphology
    - Omovertebral bar
    - Evaluation of scapular shape
  - 3D for preoperative planning
Radiology Evaluation

- MRI
  - Evaluate omovertebral bar (non-osseous)
  - Evaluate for associations

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Radiology Evaluation

• Ultrasound
  – Can show both osseous and fibrous connections
  – MRI is difficult in this young population, so US may be most helpful

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Radiology Evaluation

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Treatment

• Observation if deformity is mild
• Surgical:
  – Best between age 3 and 8 (after 8 → nerve injury)
  – Technique:
    • Resection of omovertebral bar (if present)
    • Detachment and reattachment of parascapular muscles
      – Origin of the spinous processes (Woodward)
      – Scapular insertion (Schrock and Green)
    • Scapular osteotomy
    • Sometimes with a clavicular osteotomy

References