History: 2 year old girl referred from outside institution for evaluation. Above: Scoliosis series.
AP and lateral views of the cervical spine
A - C: Multiple axial CT images extending from superior to inferior
Axial T2 image of cervical spine

Axial Fat Sat MPGR images of C-spine

What's the Diagnosis – Case 47
Coronal reformats of CT cervical spine
Findings

Multiple modalities demonstrate elevation of the left scapula with an abnormal bone emanating from the cervical spine and extending towards the left scapula. Fusion anomalies are seen of the cervical spine. There is a non osseous connection seen between the bone emanating from the cervical spine towards the left scapula. This is demonstrated on the CT by the absence of bone and on the MRI by high signal extending between the additional bone and the native scapula.
Elevation of the left scapula
Additional bone emanating from cervical spine
Fusion anomalies of C-spine

Additional bone extending from C-spine to scapula

Elevation of left scapula

Scapula
B: Additional Bone
S: Scapula

Non-osseous connection
** Additional bone extending to scapula

** Note on the right side no scapula is seen as the left scapula is elevated
Diagnosis: Sprengel’s Deformity

Sprengel’s deformity is a congenital deformity yielding an elevation and medial rotation of the scapula related to a failure of the normal caudal migration of the scapula. In approximately 30% of the deformities, an omovertebral bone is present that extends from the posterior elements of the cervical spine to the native scapula. This may be connected directly to the scapula by osseous bridging or by non-osseous (cartilage or fibrous tissue) bridging as in this case. The deformity is most typically seen in the setting of a Klippel Feil (KF) syndrome where there is fusion of two or more cervical vertebrae. KF often has associated other vertebral anomalies, a webbed neck, cervical ribs, and cardiac/pulmonary/renal/ and GI anomalies.
Resources
