

## IMAGES IN CLINICAL RADIOLOGY



### *Partial posterior split cervical spinal cord with Klippel-Feil syndrome*

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A 50 year-old woman was known with Klippel-Feil syndrome (KFS), consisting of a clinical triad of a short neck, low posterior hairline and restricted neck motion. She was complaining of new onset neck pain radiating down to mid-scapular region. Physical examination revealed moderate weakness of both biceps and plex muscles. She had diminished sensation of pinprick and light touch between C3-6 dermatoms. Radiographs revealed multiple contiguous fusion between C1 to C6 vertebrae (Fig. A). No sign of instability was detected. Magnetic resonance imaging (MRI) confirmed bony fusion of the vertebral bodies. There is complete fusion between first cervical vertebra to the fourth cervical vertebra and incomplete fusion between fifth and sixth cervical vertebrae. The seventh cervical vertebra is normal (Fig. B). Additionally posterior partial split cervical spinal cord from foramen magnum to third cervical vertebra was evident (Fig. C).

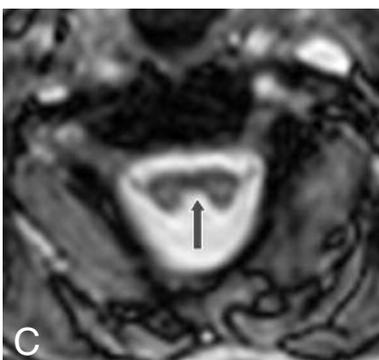
Based on the clinical presentation and typical imaging findings, the diagnosis of partial split cervical spinal cord and cervical upper vertebrae multiple non-contiguous fusions were made. MRI of the thoracolumbar spine was unremarkable. The patient has been neurologically stable for 2 years after the initial presentation. Her functional deficit remains mild and did not require any operation.

#### *Comment*

Split cord malformations (SCM) which relates to notochordal-dysraphic disorders, are rare congenital anomalies. The spinal cord splits totally or partially over a portion to form two hemicords. Symptoms varies according to the severity and length of the involvement. The lumbar and thoracolumbar regions were the most common sites involved. Cervical and cervicodorsal SCMs are very rare. Our case showed incomplete cervicodorsal SCM. Many of the patients are symptomatic at birth, but generally neurological deterioration occurs within the first 2 to 3 years of life mostly due to tight filum terminale. Numerous abnormalities are associated with split cord malformation. One of the most common is KFS. The incidence of the syndrome has been estimated to be one in 42,000 individuals. The most common abnormalities associated with KFS are skeletal anomalies.

The current classification is based on the fusion of the segments. Type I consists of a single congenitally fused cervical segment. There are multiple non-contiguous, congenitally fused segments in Type II, and in Type III patients had multiple contiguous, congenitally fused cervical segments. Our case represents Type III KFS.

Type III patients are mostly asymptomatic. Symptoms are predominantly associated with Type I patients. This explains our patient's late onset clinical findings. Symptomatology differs depending of the biomechanical features of the cervical vertebrae. Developmental basis of the SCM differs from the being complete or posterior partial. Complete SCM with anterior bony defects is mostly seen in thoraco-lumbar region, posterior partial SCM is encountered in the cervical region. Frequent clinical and radiological follow-up is recommended. Surgery may be required in the presence of neurological signs.



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