Original Research Paper

Volume-8 | Issue-10 | October-2018 | PRINT ISSN No 2249-555X

RADIO-DIAGNOSIS.

CASE REPORT OF NEUROSCHISIS OF THE CERVICAL SPINAL CORD WITH KLIPPEL-FEIL SYNDROME.

Dr.Nilesh Gupta*	Assistant Professor, PTJN Medical College and BRAM Hospital, Raipur, Chhattisgarh, India. *Corresponding Author
Dr Manish Meshram	Radiology Resident, PTJN Medical College and BRAM Hospital, Raipur, Chhattisgarh, India
Dr. Vishal Jain	Assistant Professor, PTJN Medical College and BRAM Hospital, Raipur, Chhattisgarh, India.

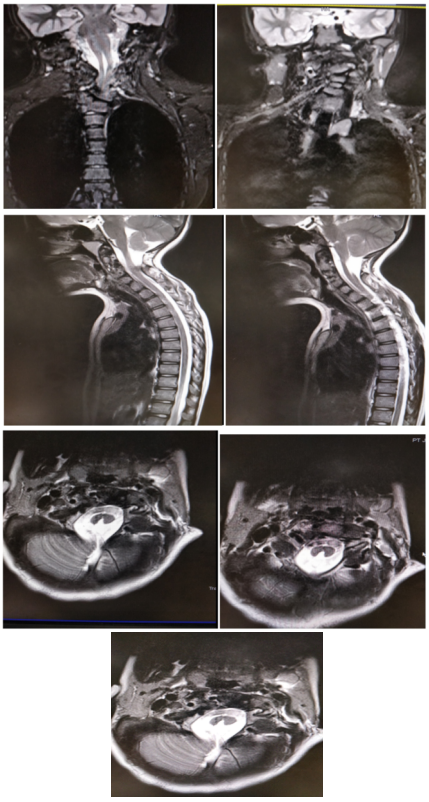
KEYWORDS : Neuroschisis, Klippel Feil, cord, MRI

INTRODUCTION:

Klippel Feil anomaly is a rare high cervical split spinal Cord associated with extensive vertebral fusions. Two distinctly separate mechanisms are suggested for the development of split cords observed: a midline lesion bisecting the neuroepithelium and the notochordal plate could be responsible for complete splitting of the cervical cord with vertebral fusion anomalies. Our purpose was to investigate the association between cervicomedullary neuroschisis and mirror movements in patients with Klippel-Feil syndrome (KFS).

CASE:

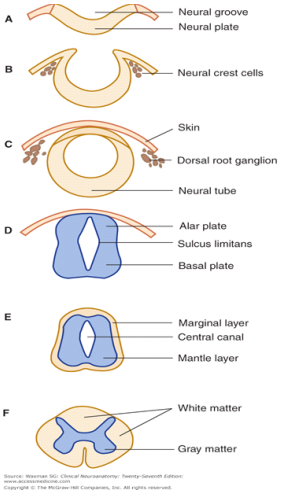
We present a case of high cervical split cord associated with extensive vertebral fusions (Klippel-Feil anomaly). Patient had mirror movements of eyes. The patient had no restriction of movement of neck. On MRI, we found fusion of atlanto-occipital joint (with non visualisation of posterior arch of C1 vertebral body), fusion of C1-C2, C2-C3, C3-C4 and D1-D2 vertebral bodies: block vertebras. Butterfly shaped of C3 and C4 vertebral bodies and few cervical hemi vertebrae noted. There is small slit noted extending from C2 to C6 cervical cord filled by T2/FLAIR hyperintense CSF, suggesting the neuroschisis of upper cervical spinal cord. Scoliosis of the cervico-dorsal spine with concavity to the right side noted.



DISCUSSION:

Klippel Feil anomaly is a rare high cervical split spinal Cord associated with extensive vertebral fusions. In 1912, Klippel and Feil (1) first described what is known today as KFS. In 1919, Feil (2) reported 13 additional cases, which he classified into three groups. KFS is associated with the classic triad of a short neck, a low posterior hairline and a limited range of motion of the neck; it is caused by congenital fusion of two or more cervical vertebrae (4, 5). **Organogenesis:** All neurons and supporting cells are derived from ectoderm. Cells of the early embryo's midline dorsal ectoderm are induced by the underlying notochord to form a thickened neural plate. **Neural Tube Formation:** The plate's lateral border thickens and the center invaginates, forming a troughlike neural groove**.** As the groove deepens, the lateral borders contact each other to close the groove and form the neural tube**.** Cells lining the tube elongate to form a mitotically active pseudostratified columnar epithelium (neuroepithelium), and they eventually develop into various types of neurons and supporting glial cells.

As the neural groove closes, cells at its lateral borders proliferate to form two columnar masses that come to lie dorsal to the neural tube and form the neural crest**.** Neural crest cells migrate away from the neural tube and form the PNS, including the sensory neurons of the craniospinal ganglia, the postganglionic neurons of the ANS, the Schwann cells of peripheral nerves, and the satellite cells of ganglia. Neural crest cells also form the meninges and the craniofacial mesenchyme. **Differentiation:** At about the third week of prenatal development, the ectoderm of the embryonic disk forms the neural plate, which folds at the edges into the neural tube (neuraxis**)**. It forms at the embryo's dorsal midline which runs for most of the length of the embryo. The cellular elements of the tube appear undifferentiated at first, but they later develop into various types of neurons and supporting glial cells. A group of cells migrates to form the neural crest, which gives rise to dorsal and autonomic ganglia, the adrenal medulla, and other structures. The middle portion of the neural tube closes first; the openings at each end close later.



KFS can be classified into three subtypes (6)—type I, II or III:
Type I is defined as having multiple cervical or upper thoracic vertebral fusions;

Type II is defined as having isolated fusions at 1 or 2 cervical interspaces;

Type III is defined as having fusions in the cervical spine combined with lower thoracic or lumbar fusion.

This classification has remained useful during the years, and the term KFS is generally regarded to include any form of congenital cervical vertebral fusion.

Grading system for cervical cord neuroschisis
Grade 0 Normal cord
Grade 1 Deformity without Cleft
Grade 2 Posterior cleft alone
Grade 3 Anterior cleft with or without posterior cleft
Grade 4 Bow tie configuration
Grade 5 Complete neuroschisis.

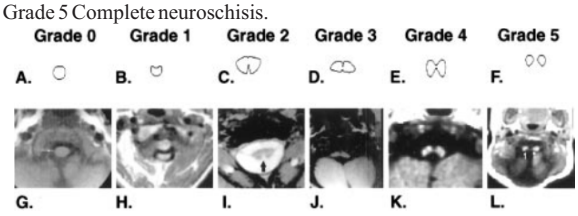


FIG . Illustration of grading system for neuroschisis.
A-F, Line drawings of the grading system of neuroschisis.
G-L, Axial MR image examples of each grade chosen from patients in this study.

According to above mention classification, our case was classified as type I of KFS. In recent years, numerous studies of KFS have been reported, particularly regarding the genetic etiology, such as a paracentric inversion on 8q (6), mutations of MEOX1 gene (7, 8), Notch signaling pathway (33, 36), Pax 1 and Pax 9 (5), or HOX gene (21). However, most of these conditions have only been confirmed in animal studies, and further studies are warranted to investigate the exact genetic origins of KFS.

CONCLUSION:
Unusual presentation and radiological findings in a patient with Klippel-Feil syndrome prompted this report. Our case of type I of KFS with is delineating a strong association exists between cervicomedullary neuroschisis and mirror movements in cases of KFS. Screening of patients with mirror movements may help identify clinically unsuspected KFS and may also help stratify risk within this patient population, identifying patients who might benefit from early neurosurgical intervention.

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