Situs inversus totalis

A rare, genetic, developmental defect during embryogenesis characterized by total mirror-image transposition of both thoracic and abdominal viscera across the left-right axis of the body.

Abdali et al. reported a child with an extremely rare association of Klippel-Feil syndrome with situs inversus totalis (SIT). Both KFS and SIT are genetically heterogeneous and their co-occurrence suggests a high possibility of sharing the same underlying causative agent. They reviewed the genetic background that is known for these two conditions in the literature. [1].

Few aberrations involving the neuraxis, and rarely intracranial aneurysms, have been reported in association with situs inversus totalis.

Chakravarthi et al. described the radiological findings and endovascular management of one of the first of its kind: the association of bilateral carotid cavernous aneurysms (one large and the other small-sized) with situs inversus totalis [2].

The association of situs inversus totalis (SIT) and VACTERL syndrome an extremely rare coincidence.

Patients: The patient was first diagnosed as simple SIT with lumbosacral neoplasms according to the prenatal magnetic resonance imaging (MRI) examination; however, the local hospital ignored the important to physical examination so that missed anal atresia with fistula. The patient was presented to our hospital owing to constipation for 1 week. And then, she was diagnosed as VACTER syndrome with situs inversus totalis.

Results: Anorectoplasty was performed to treat constipation, one month later, we performed intramedullary tumor resection and pathological diagnosis of ependymal cyst. Postoperative recovery was uneventful and the baby was doing well at 5-months follow up.

Conclusion: It is extremely necessary for careful physical examination and detailed auxiliary examination to each system (including echocardiography, MRI, and so on) when diagnosing SIT. Also, recognizing and understanding the spectrum of situs anomalies is important, which aids in the diagnosis of disease and accordingly plan the therapeutic interventions [3].

A case report of myelopathic hypoplasia of the atlas with situs inversus totalis.

Objectives: To describe a case of cervical myelopathy caused by hypoplasia of the atlas with situs inversus totalis, and to briefly review the pertinent literature.

Setting: Department of Orthopaedic Surgery, Kagoshima, Japan.

Methods: The history, results of examination, and findings of radiographic imaging studies for a 56-

year-old man with a 10-year history of progressive myelopathy who presented to our hospital are described.

Results: Imaging studies revealed congenital hypoplasia of the atlas, ossification of the posterior longitudinal ligament at the levels of C3-C4, and situs inversus totalis. He underwent laminectomy of the atlas and laminoplasty of C3-C7 for decompression of the spinal cord. Operative intervention resulted in significant neurological improvement and relief of occipital neuralgia.

Conclusion: To our knowledge, no case of myelopathic hypoplasia of the atlas with situs inversus totalis has previously been described. When encountering inherited disorders such as situs inversus totalis, a thorough search must be made for anomalies of the craniovertebral junction [4].

First published report of an adult complex spinal dysraphism with situs inversus.

Objectives: To describe a previously asymptomatic adult patient of multiple vertebral anomalies with cervical split cord malformation type II, tethering of the spinal cord (cervical and lumbar), and intraspinal arachnoid cyst along with dextrocardia and situs inversus.

Summary of background data: Only 5 cases (fetus, 1; neonates, 3; child, 1) of spinal dysraphism with dextrocardia or situs inversus have been reported. All these cases have had associated multiorgan developmental anomalies usually incompatible with survival and requiring multidisciplinary care.

Methods: The case has been described and relevant literature reviewed.

Results: The patient was operated for cervical and lumbar levels in the same sitting. A C4-C5 laminectomy was performed, 2 hemicords enclosed in the same dural sac were visualized, dorsal paramedian nerve roots and the tethering arachnoid bands were cut, and the arachnoid cyst wall was partially excised. This was followed by L4-L5 laminectomy and detethering by sectioning of the thickened filum terminale. The patient showed significant neurologic improvement after surgery.

Conclusions: The present case is a rare instance in which there has been an association of adult onset occult spinal dysraphism along with situs inversus totalis. Successful management requires appropriate understanding of embryology, anatomy, and imaging and has implications in neurosurgical and perioperative anesthetic care [5].

Tubbs et al. examined the gross anatomy of an elderly cadaveric female for a possible “situs inversus” of the intracranial contents. This study has found that many structures commonly dominant on one side in the intracranial compartment were reversed in this specimen. These findings support the concept that a reversal of more commonly found intracranial anatomy may occur in situs inversus totalis, and this should alert the clinician performing invasive procedures in this population. These data will also hopefully provide further insight into possible mechanisms that contribute to situs inversus totalis [6].

1)
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6)

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