

## A rare case of diastematomyelia with hemivertebrae and intradural teratoma.

Jayakrishnan Nambiar<sup>1</sup>, Sukhen N Shetty<sup>2</sup>, Padmanabha S<sup>3</sup>

<sup>1</sup>Post graduate 2nd year, Department of anaesthesiology, Yenepoya medical college and hospital,

<sup>2</sup>Post graduate 3rd year, Department of anaesthesiology, Yenepoya medical college and hospital

<sup>3</sup>Professor and head of department, Department of anaesthesiology, Yenepoya medical college and hospital, Deralakatte, Mangalore

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**Abstract:** Diastematomyelia, or split cord malformation, a complete or incomplete sagittal division of the neural axis into halves, is seen in association with many other congenital anomalies. Among these anomalies, intradural spinal teratoma is extremely rare. Diastematomyelia is a well-recognized although unusual clinical syndrome in children, but it is rarely reported in the adult. We report a case of 19 year old girl who presented with pain and distal left-leg weakness as well as neurogenic claudication for 1 month. The patient underwent radiological examinations, and diastematomyelia and an intradural lumbar teratoma were diagnosed. she underwent surgery with definite anaesthesia management. This is the first case of an adult who simultaneously presented with diastematomyelia and an intradural teratoma with hemivertebrae. We hereby discuss the anaesthetic plan and management of the same.

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### I. Case report:

A 19-year-old lady weighing 40 kg and measuring 133 cm in height presented with uterine prolapse for Fothergills surgery with anteroposterior repair. There was no history suggestive of any major illness in the past. She had undergone tubal ligation under general anaesthesia uneventfully four months prior to this surgery. She had hair growth on her lumbar vertebrae region. There was no history suggestive of a neurological deficit, breathlessness or limitation of lower limbs or spine.

One month back had a h/o fall from a parapet, following which she developed lower limb weakness, inability to walk, motor activity was reduced.

### II. Investigations:

On examination, her vital parameters were within normal limits. Mouth opening was adequate and score on modified Mallampati test was grade 2. She had protruding upper teeth. There was no limitation of flexion or extension at the cervical spine, but the cervical spine appeared short. Bilaterally the scapulae were elevated. lumbar spine was not well palpable. Her respiratory system was normal and her breath holding time was 20 seconds.

All routine investigations including arterial blood gases, ECG, and renal parameters were normal, except the x-ray chest, which showed bilaterally high scapulae There was also evidence of hemivertebrae [Figure - 2]. There was crowding of upper ribs. The cardiac shadow and both the lung fields were normal. A MRI of spine revealed spina bifida with diastematomyelia an extrusion, and compression of spinal cord. An indirect laryngoscopy revealed an overhanging epiglottis, with part of the posterior cord and arytenoids seen.

Written informed consent was taken after explaining the risk of anaesthesia to the patient. Routine pre-anaesthesia precautions and procedures were followed. Difficult intubation cart was kept ready. Patient was sedated with injection Midazolam 1.5 mg and injection Pentazocine 24 mg. Patient was induced with injection Propofol 90 mg (mixed with injection Xylocard 10mg) intravenously. Patient was given prone position taking care of her shoulder joints and spine. Patient was maintained on Propofol infusion. The infusion rate was titrated between 9 mg/kg/hour and 4 mg/kg/hour to maintain adequate depth of anaesthesia. Surgery proceeded smoothly with the patient maintaining adequate depth of anaesthesia. Surgery lasted for 4 hour and 15 minutes. Patient was fully awake three minutes after stopping the infusion. Total dose of propofol required throughout surgery was 570 mg. There were no untoward post-operative complications.

### III. Discussion

Diastematomyelia is a rare entity in which some portion of the spinal cord is split into two by a midline septum. Most cases occur in childhood, but some develop in adulthood. A variety of concurrent spinal anomalies may be found in patients with diastematomyelia.

The authors describe a 38-year-old right-handed woman who presented with a 7-month history of lower-extremity pain and weakness on the right side. She denied recent trauma or illness. Sensorimotor deficits, hyperreflexia, and a positive Babinski reflex in the right lower extremity were demonstrated on examination.

Neuroimaging revealed diastematomyelia extending from T-1 to T-3, an expanded right hemicord from T-2 to T-4, and a C6–7 syrinx. The patient underwent T1–3 total laminectomies, resection of the septum, untethering of the cord, and excision of the hemicord lesion. The hemicord mass was determined to be an intramedullary epidermoid cyst; on microscopic evaluation the diastematomyelia cleft was shown to contain fibroadipose connective tissue with nerve twigs and ganglion cells. Postoperatively, the right lower-extremity pain, weakness, and sensory deficits improved.

Diastematomyelia can present after a long, relatively asymptomatic period and should be kept in the differential diagnosis for radiculopathy, myelopathy, tethered cord syndrome, or cauda equina syndrome. Numerous spinal lesions can be found in conjunction with diastematomyelia. To the authors' knowledge, this is the first case in which a thoracic epidermoid cyst and cervical syrinx occurred concurrently with an upper thoracic diastematomyelia. Thorough neuraxis radiographic evaluation and surgical treatment are usually indicated.

Adequate workup of a patient with congenital spinal deformities like hemivertebrae is necessary. Particular attention should be paid to cardiac, neurological and genitourinary systems as many patients have associated anomalies. 20-30 % of the patients may have associated genitourinary tract anomalies. 10% of the patients may have congenital heart disease and 5 -10 % of the patients may have spinal dysraphism including cervical Sprengels deformity, tethered chord and fibrous bands[2].

In view of the likely association of neurological anomalies in these patients, it was decided to avoid regional anaesthesia and to conduct the case on total intravenous anaesthesia supplemented with a local block. This would avoid the trauma and morbidity associated with a difficult intubation, the likelihood of which could not be ruled out in such cases.

Propofol is a rapidly acting hypnotic which is used for intravenous anaesthesia. It has a brief duration of action due to redistribution and metabolism. It is not cumulative after repeated injections. Total intravenous anaesthesia with propofol is smooth, with a reduced incidence of coughing and hiccups, as compared with barbiturates and etomidate. It has fewer incidence of postoperative nausea and vomiting. Thus, propofol is a good hypnotic for use as a continuous infusion to maintain hypnosis in total intravenous anaesthesia. The dose required is 6-12 mg/kg/hour after a bolus dose[3].

Because of the rapid return of respiration and awakening, lack of postoperative side effects including vomiting, total intravenous anaesthesia using propofol with good premedication and oxygen supplementation provides excellent anaesthesia for surgeries lasting for an hour or less.

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