ABSTRACT

Diastematomyelia is a rare dysraphic lesion of spinal cord and the defect occurs as a result of an incorrect development of notochord between 15th and 18th day of pregnancy. In this defect, the spinal cord splits into two hemicords in sagittal plane. We hereby present a case of 30 years old female who had been experiencing spine pain without any neurological deficit for over 10 years. Despite the confirmed osseous anomalies of the lumbar spine, the correct diagnosis was established only after performing the MRI. She was treated surgically and was asked to come for regular neurological examinations after discharge.

CASE REPORT

A 30-year-old female patient came to Department of Orthopaedics with a complaint of back pain for many years. Before proceeding for MRI study, detail history and local examination of the patient was done. On local examination there was no obvious dimpling or hypertrichosis seen in the lumbar region.

For the last two years, the patient has been experiencing muscle weakness in both lower limbs and progressive difficulty in walking. The patient had no other neurological deficit. No other gait abnormalities were revealed; only the disturbances of articular mobility. The symptoms were aggravated since last 6 months. Patient had intermittent back pain since two years, for which she consulted a general physician in a primary health centre of her village many times. She was being prescribed analgesics for back pain from which she used to get temporary relief. But since she experienced severe back pain and lower limb weakness she came to our hospital. No radiological investigation was done earlier, since she had no such complaint when she was young.

The MRI lumbar spine was performed in Department of Radio-diagnosis, AVBRH Hospital, Wardha. The examination was carried out with 1.5 Tesla MRI Scanner, all sequences including T1, T2, PD was taken in sagittal, axial and coronal planes. On the basis of performed sequences lumbar lordosis was found.

There was congenital fusion of L1-L2 vertebral bodies and partial fusion of posterior elements at L1-L2 [Table/Fig-1], widening of dural space and splitting with thickening of cord noted at L1-L2 level [Table/Fig-2]. Two hemicords contained within individual neural tubes separated by a bony septum that extends from vertebral body to posterior arches at L1-L2 vertebral level [Table/Fig-3&4]. Diagnosis of diastematomyelia with bony septa was made. Decompression (surgery) of neural elements and removal of bony spur was done. The patient was asked to come for regular neurological examinations after discharge.

DISCUSSION

Diastematomyelia is an uncommon condition which is usually more commonly seen in children and it is quite rare in adults.
In most of adults, it is most commonly seen in lumbar and lower two third of thoracic region [1].

It can be associated with other spinal dysraphism such as meningocele, myelomeningocele, spinal lipoma and congenital defects of spine including hemivertebrae, butterfly vertebrae and scoliosis. In this disorder there is separation of spinal cord into two parts in sagittal plane (hemi-cords). It may coexist with other spinal dysraphism, such as meningomyelocele, myelocoele, spinal lipoma, neuroenteric cyst or dermal sinuses and vertebral anomalies such as hemi-vertebrae, butterfly vertebrae or scoliosis [2].

Diagnosis of diastematomyelia is usually first made in the neonatal period, and sometimes in late stages. On the basis of the location of the mass lesion with respect to the surface of the back, these lesions can be categorised into three groups, the open dysraphism in which there is dorsal protrusion of soft tissues of the spinal canal which is not covered by skin and there are close dysraphisms with a protrusion which is covered by skin, and occult spinal dysraphisms in which there is no dorsal protrusions of soft tissues of the spinal canal [3,4].

MRI is most important diagnostic imaging modality of choice for dysraphisms of the spinal cord which plays a very important role for making a final diagnosis and for planning a further intervention [4-6].

In, diastematomyelia there is vertical splitting of the spinal cord. Pang et al., has classified diastematomyelia into various categories. According to them, in type I Diastematomyelia there are two hemicords, each of which is contained in a separate dural tube and separated by a bony septum. Type II Diastematomyelia consists of both the hemicords are contained within single dural sac and separated by a non-rigid fibrous septum and the fibrous septum is rarely visualised in imaging studies [7]. This differentiation which is made through MRI has surgical importance as type I Diastematomyelia are technically more difficult to correct and are associated with poor prognosis and outcome than type II, especially if there is an oblique septum which divides the cord asymmetrically [8].

In the presented case, the 30-year-old woman had been experiencing spine pain without any neurological deficit for over 10 years. Despite the confirmed osseous anomalies of the lumbar spine, the correct diagnosis was established only after performing the MRI.

According to the classification and in our case, the presence of the bony septum dividing the spinal canal into two parts with two hemicords in two separate dural tubes is considered as Diastematomyelia of type I, although in our case, above and below the septum, the hemicords were fused in one cord lying in the one common dural sac again [5,9-11].

It is interesting to note that in our case, the female patient did not have any significant neurological or orthopaedic abnormalities at younger age, apart from long-term back pain.

Folic acid deficiency is important nutritional factors connected with spinal dysraphisms. Other important factors among nutritional factors are zinc deficiency, high consumption of nitrates, vitamin A deficiency or hypervitaminosis A [5]. Though genetic factors have been mentioned to play a role, the aetiology is complex due to the involvement of physiologic and psychological factors [6].

Diplomyelia, is a condition in which there is isolated accessory spinal cord which presents in the form of, absence of roots at the ventral lumbosacral level. The use of term diplomyelia should be restricted to cases of an isolated accessory spinal cord, ventral or dorsal to the normal cord [12].

CONCLUSION

Diastematomyelia is a rare dyraphic defect of the spinal cord which is mostly diagnosed in the prenatal period or in children and much less often in adults. It is interesting to note that in our case, the female patient did not have any significant neurological or orthopaedic abnormalities at younger age, apart from long-term back pain.
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