Diastematomyelia and spinal teratoma in an adult

Case report

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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. The authors describe a 42-year-old man 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. He underwent surgery and was followed for 1 year. This is the fourth case of an adult who simultaneously presented with diastematomyelia and an intradural teratoma.

KEY WORDS • diastematomyelia • intradural teratoma • occult spinal dysraphism • split cord malformation

Diastematomyelia, or SCM, is a form of OSD that appears with duplication of the spinal canal.^{4,8,11} It is usually accompanied by a number of other malformations such as cutaneous changes, skeletal and vertebral anomalies, meningocele or myelomeningocele, hydromyelia, Klippel-Feil syndrome, hydrocephalus, and Arnold-Chiari malformation, as well as intradural spinal teratoma in rare cases.^{4,11,16} Intradural spinal teratomas are also extremely rare dysembryogenetic tumors.² Moreover, a spinal teratoma is less common in adults.^{2,7} There are several reports in the literature of patients presenting with diastematomyelia and intradural spinal teratoma simultaneously.^{2,7} We describe the case of an adult patient who presented with diastematomyelia and an intradural spinal teratoma.

CASE REPORT

Presentation and Examination. This 42-year-old man had experienced increasing pain and distal left-leg weakness as well as neurogenic claudication for 1 month. Neurological examination revealed 2/5 strength in the left anterior tibialis and extensor hallucis longus muscles. Hy-

pesthesia was found from the left leg distally to the knee region. The Achilles reflex was depressed in the left leg and there were no pathological reflexes. The radiological examination consisted of plain radiography, 3D CT scanning, and MR imaging. Two-plane radiography revealed scoliosis of the lumbar spine and the lumbar vertebral body anomalies including hemivertebrae, butterfly vertebrae, block vertebrae, and decreased intervertebral disc height (Fig. 1). Spina bifida was also present. A central bone spur was seen at the level of L-5. These radiographic findings were consistent with a diagnosis of diastematomyelia. The MR imaging study of the lumbar spine revealed a large spinal canal divided into two sections by a bone spur that originated from the L-5 vertebral body, and there was development of the subcutaneous fat within the bone spur. At the level of the spur, two dural sacs were contained within separate hemicanals. Caudal to the spur, the two separate dural sacs fused into a single-wide dural sac. Both of the hemicanals were large. At the level of the bone spur, T₁- and T₂-weighted MR imaging demonstrated a large mass in the left hemicanal containing heterogeneous signal intensities compatible with a mixture of calcification, soft tissue, fat, and a cyst. Following intravenous administration of contrast material, the mass showed inhomogeneous enhancement (Fig. 2). The findings were consistent with the diagnosis of a teratoma. The axial reference, sagittal reformats, and 3D reconstructions

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance; OSD = occult spinal dysraphism; SCM = split cord malformation; 3D = three-dimensional.

Fig. 1. Plain radiograph of the spine revealing scoliosis, hemi-vertebrae, and block vertebrae. A central bone spur was seen at L-5 (arrow).

of the 3D CT scans confirmed and better delineated the vertebral anomalies and bone spur detected by plain radiography and MR imaging (Fig. 3). Fat was observed within the spur. The mass showed heterogeneous density containing calcium, cyst, fat, and soft tissue. The MR imaging and CT findings were consistent with diastematomyelia and intraspinal teratoma. Magnetic resonance imaging examinations of the thoracic and cervical spine demonstrated normal findings.

Operation. The patient underwent a lumbar laminectomy, removal of the bone spur and cystic mass, and sectioning of the terminal filum. The postoperative period was uneventful.

Follow-Up Period. One year after surgery, the neurogenic claudication had completely resolved, but strength in the left anterior tibialis and extensor hallucis longus muscles was still 2/5.

Histological Examination. The tissue was processed in the conventional manner with formalin fixation followed by paraffin embedding. Histopathologically, the lesion was composed of stratified squamous epithelium with underlying sebaceous glands and other skin adnexal structures intermingling with glial tissue (Fig. 4). Histopathological diagnosis was a mature cystic teratoma.

DISCUSSION

Diastematomyelia (or SCM) is a form of OSD that appears with duplication of the spinal canal.^{4,8,11} It is usually accompanied by a number of other malformations, including cutaneous changes, especially skeletal vertebral anomalies, hydromyelia, Klippel–Feil syndrome, hydrocephalus, and Arnold–Chiari malformation and as well as intradural teratoma in rare cases.^{4,11,16}

Fig. 2. Sagittal (upper) and axial (lower) T₁-weighted MR images (TR = 450 msec, TE = 11 msec) of the lumbar spine demonstrating the mass and the hyperintense spur. The mass has solid, cystic, and fatty components.

Pang¹¹ has suggested alternative terms for SCM: types I and II. Type I SCMs consist of two hemicords that are separated by an osteocartilaginous septum; each hemicord is covered by a separated dural sheath. Type II SCMs consist of two hemicords within the same dural envelope and separated by a fibrous septum.¹¹ In our case, the patient was shown to harbor a type I lesion.

Teratomas are histologically characterized by the presence of tissues that are endomesodermal in origin and are thought to be from cells that did not differentiate during embryogenesis.^{15,18} Teratoma is also defined by Willis¹⁸ as a neoplasm composed of multiple tissues that are foreign to their present location. Teratomas are included in a group of tumors classified as inclusion tumors.⁸ Classically, teratomas are thought to originate from aberrant pluripotential cells of yolk sac migrating to the amnion during embryogenesis.^{2,10,14} The presence of teratomas in the spine is extremely rare. Two hundred fifty-six teratomas were reported by Tapper and Lack,¹⁵ and in four cases the tumors were located in the spinal canal. Sloof, et al.,¹⁴ reviewed 301 cases of teratoma and in two cases the tumor was found in the spinal canal. Caruso, et al.,² reviewed the intramedullary spinal teratoma in a total of

Fig. 3. Axial reference (*upper left*) and (*right*) and sagittal reformat (*lower left*) and craniocaudal view of 3D reconstruction (*lower right*) of a CT scan demonstrating a bone spur extending from the L-5 vertebral body to the lamina and dividing the spinal canal and the dural sac into two parts. Solid, cystic and fatty components, and calcification of the teratoma located within the left hemicanal can be seen.

33 cases, and two cases were associated with diastematomyelia.

In adults, simultaneous presentation of occult spinal dysraphic lesions and a spinal teratoma is rare. In many reports the authors describe the occurrence of SCM in association with some inclusion tumors.^{4-6,11} In contrast, some teratomas have been reported in the presence of spinal dysraphic lesions.^{1,12,17} Simultaneous presentation of SCM

extradural spinal teratoma has been reported in several cases.^{5,9,16} Hader, et al.,⁷ reported 12 cases of simultaneous presentation of SCM and intradural spinal teratoma. Only two of the 12 cases were observed in adults.^{3,10} Rosenbaum, et al.,¹³ have reported the case of an adult who presented with both SCM and intradural spinal teratoma. Our case is the fourth and describes the oldest adult patient who has presented with diastematomyelia and spinal intradural teratoma. In all four cases the lesions were type I SCM. In two cases, the SCM and intradural teratomas were located in the thoracic area^{3,13} and in one case the lesions were observed in the thoracolumbar region.¹⁰ The whole spinal canal must be evaluated radiologically in an adult patient who harbors a spinal teratoma because of the possibility that spinal dysraphic lesions may be present. Pain is the most frequent symptom of OSD in adults, and neurological deficits may accompany the pain.^{4,8,10,11} Teratoma may increase neurological problems that result from an SCM. This simultaneous presentation of SCM and teratoma only results in a more complex surgery. In our case a new neurological deficit was not observed in the postoperative period.

CONCLUSIONS

The association of diastematomyelia, as a form of OSD, and spinal intradural teratoma in adulthood is rare. In adult patients with an intradural spinal mass lesion resembling a teratoma, the whole spinal canal should be evaluated radiologically for additional occult spinal dysraphic lesions.

Fig. 4. Photomicrograph. The lesion is composed of differentiated organoid structures such as neural tissue, glandular elements, and clusters of squamous stratified epithelium. H & E, original magnification $\times 80$.

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