An Infantile Large and Extensive Intramedullary Mature Spinal Teratoma
—Case Report—

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Abstract

A 5-month-old boy presented with a rare case of intramedullary teratoma extending from T1 to S5 and associated with a lumbosacral lipoma. Magnetic resonance imaging showed the tumor extended over the lipoma. Since the tumor could not be clearly demarcated from the spinal cord, surgery was performed under a preoperative diagnosis of spinal glioma. The lack of demarcation made safe resection of the tumor difficult to perform, so that the tumor was only partially removed by decompressing the spinal cord. The histological diagnosis was mature teratoma. This case was likely the result of some dysembryogenetic mechanism and the tumor was not neoplastic. The long-term outcome cannot yet be determined, but no recurrence of the symptoms has been observed for 3 years. Spinal intramedullary teratoma is rare and tends to be located in the lumbosacral region. Surgical intervention is crucial and total removal may be achievable, but is likely to be partial with tight adhesion to the spinal cord, so that aggressive approaches should be avoided in such cases.

Key words: spinal cord teratoma, spinal lipoma, spina bifida, magnetic resonance imaging, infant

Introduction

Teratoma of the intraspinal canal is uncommon and accounts for only 2% of all cases of teratoma in the central nervous system.8,14) Spinal intramedullary teratomas are even less common and are usually small and located in the conus medullaris.12) The majority of such tumors are benign and frequently associated with congenital dysraphic defects, indicating that some dysembryonic mechanisms may be involved.5–6,13) Preservation of both the normal spinal anatomy and the spinal cord function is the primary concern in the treatment of infantile intramedullary spinal teratoma, so the optimal role of surgical intervention is difficult to establish. We treated an infant with an intramedullary mature teratoma extending from T1 to L5 and associated with lumbosacral lipoma.

Case Report

A 5-month-old boy was admitted to our hospital with symptoms of slowly progressive paraparesis which his parents had first become aware of when he was 4 months of age. A dimple in the sacrococcygeal region was noted at birth and simple observation detected no signs of either cerebrospinal fluid leakage or infection. When the patient was 2 months old, he suffered from a urinary tract infection and was treated with antibiotics.

On admission, his developmental status was normal...
with body weight of 8.4 kg and height of 69 cm. No cranial nerve signs or deformities were detected, and no pigmentation or hair was observed around the dimple in the sacrococcygeal region. Muscle tonus and deep tendon reflex were normal in the upper extremities, but paraplegic and hypotonic in the lower extremities. The patient was suffering from urinary retention and constipation.

Spinal computed tomography showed spina bifida occulta without spinous processes below the L5 level (Fig. 1A). The bilateral intervertebral foramens were enlarged at T12–L4, indicating that a huge mass had been present in the spinal canal for a long time. Magnetic resonance (MR) imaging of the whole spine showed a large intramedullary tumor extending from T1 to the lumbosacral region (Fig. 1B–D). The lower end of the tumor extended to a lipoma associated with multiple cysts in the lumbosacral region but not to the subcutaneous fat. The major part of the thoracolumbar intramedullary tumor was not enhanced by gadolinium-diethylenetriaminepenta-acetic acid, with only faint enhancement in the axial view of the center of the tumor around the T3–T6 levels (Fig. 1E). The tumor extended through the enlarged intervertebral foramens, thus forming a retroperitoneal paravertebral mass (Fig. 1F).

The MR imaging findings of the thoracolumbar intramedullary tumor resembled those of astrocytoma. The spinal canal was entirely filled by the tumor, so the spinal cord was difficult to demarcate from the tumor. We planned surgical intervention to decompress the spinal cord and establish the histological diagnosis.

Intraoperative inspection found hypoplasia of the spinous process of L5 as well as absence of lamina below the S1 level. Reconstructive laminotomy was performed from T1 to L5 and the hypoplastic S1–S3 laminas were opened. Opening of the dura revealed that the spinal canal was occupied by a yellowish-white, elastic but hard, homogeneous solid tumor. The normal spinal cord could not be identified. The lipoma and cysts were located in the sacrococcygeal region, whereas the dysplastic conus medullaris extended to the extradural space at the S3 level. The nerve roots were identified at the L5–S2 levels, but were encased by the lipoma. The conus medullaris was highly dysgenetic and not clearly demarcated from the rostrally located intramedullary tumor or the spinal cord. The tumor was elastic, hard, and hemorrhagic without cystic components. Since the plane between the tumor and the spinal cord could not be clearly identified, the surgery was terminated after the spinal canal had been thoroughly decompressed. The postoperative course was un-
Fig. 2  T₁-weighted magnetic resonance images with gadolinium obtained 3 years after surgery showing adequate decompression of the spinal canal but the normal spinal cord could not be satisfactorily visualized (A), and the tumor has formed a large lumbosacral cyst (B).

Fig. 3 Photomicrographs of the tumor showing pseudostratified ciliated columnar epitheliums and a seromucous gland (A), papillary proliferation of single-layer epithelium resembling choroid plexus cyst (B), cerebellar-like neuronal tissue (C), and mature fat tissue (D). Hematoxylin and eosin stain, original magnification A, B, D: × 100, C: × 200.

eventful, but the patient showed no neurological improvement. During the 3-year follow-up period, the neurological condition and tumor size remained unchanged, but the lumbosacral cyst became larger (Fig. 2).

Histological examination indicated that a major part of the tumor mimicked the characteristics of cerebrospinal tissue, and contained pseudostratified ciliated columnar epithelium and seromucous glands, papillary proliferation of the single-layer epithelium resembling choroid plexus cyst, cerebellar-like tissue containing granule cells with small somas, and mature fat tissue surrounded by sparse connective tissue (Fig. 3). No mitosis was observed. A large number of capillary vessels without endothelial proliferation were detected. The presence of pseudostratified ciliated columnar epithelia and seromucous glands mimicked the appearance of bronchus and only a small amount of mature fat tissue was observed. None of the specimens showed any malignant or immature elements. Finally, examination of the sacrococcygeal lipomatous tissue showed normal adipocytes. Based on the presence of three well-differentiated germinal matrix layers and a strictly morphological definition, the histological diagnosis was mature teratoma.

Discussion

The present case of teratoma was associated with lumbosacral spinal dysraphism extending up to the cervicothoracic junction level. Teratomas are occasionally associated with congenital dysraphic defects. Five of 33 intramedullary teratomas were associated with spinal dysraphic lesions, and 41.7% of teratomas were associated with a concomitant anomaly of the vertebral canal, most commonly a diastematomyelia. Therefore, lumbar and sacroccocygeal region can exhibit an unusual array of congenital tumors and malformations. The embryology of this part of the neural tube is complex and includes pluripotential cells capable of forming lesions with overlapping pathological patterns. Intraspinal teratomas have conventionally been thought to arise from primordial germ cells migrating from the primitive yolk sac. However, most central nervous system teratomas occur in midline structures and could be derived from pluripotent cells which remain at the sites of early neural tube closure, indicating a dysembryogenic origin for intraspinal teratoma.

The present case also presented with a lumbosacral lipoma which encased the nerve root and looked like a normal lipoma. Such lipomas are known to feature markedly diverse tissue in addition to fat. Tissues within such lipomas include the epidermal sinus tract, ependymal structures, neuroglial cells, fibrous tissue, skeletal muscle, cartilage, renal tissue, and the cerebellum. Histological examinations of congenital intraspinal lipomas have found complex forms including various unusual ectopic tissues of ectodermal, mesodermal, and/or endodermal origin in addition to the lipomatous component. The combination of these tissue elements suggests these lipomas might even be more accurately regarded as hamartomas.

Intramedullary teratomas are commonly small and localized around the conus, but a few cases of greatly extended teratomas have been reported. A truly intraspinal immature teratoma was located between C5 and T12 and contained cysts of various sizes, and a giant intramedullary teratoma occurred between the T2 and lumbosacral region, and extended to an intraabdominal cyst through the neural foramina. However, these two cases were not associated with spinal dysraphism as seen in our case. The dysembryogenic process may have been involved in our case. The two previously reported large teratomas without
dysraphism were easily dissected from the spinal cord, but the association of the tumor with dysraphism made discrimination from the normal structure of the spinal cord extremely difficult in our case. About half of all intramedullary teratomas feature intimal adhesion to the surrounding nervous roots so that complete removal has been achieved in only 61.6% of cases. However, subtotal removal has resulted in no subsequent recurrence of symptoms over many years, mainly due to the extremely slow growth of such tumors. The overall mortality rate of 28.3% for mature teratomas during a mean follow-up period of 38 months was better than that for either malignant or intramedullary teratomas. Surgical intervention in such cases should be primarily limited to confirming the histological diagnosis of the tumor.

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References


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