Uterus-like mass of mullerian origin in the lumbosacral region causing cord tethering

Report of two cases

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The presence of mullerian-origin tissue in the lumbosacral region is extremely uncommon. The authors report two cases of mullerian-origin tissue in that region. In the first case a 33-year-old woman harbored a conus medullaris mass lesion. Spinal dysraphism, tethered cord syndrome (TCS), and diastematomyelia were also present. In the second case a 24-year-old woman presented with low-back pain and a conus medullaris lesion, which was a cause of the TCS. Pathological examination in both cases revealed a uterus-like structure with evidence of fresh and old hemorrhage. The rarity of this lesion and its association with diastematomyelia requires documentation.

Key Words • choristoma • heterotopia • tethered cord syndrome • mullerian tissue • uterus • spinal dysraphism • spina bifida

Case Reports

Case 1

History and Presentation. This 33-year-old woman presented with a swollen mass on her lower back that had been present since birth. The size of this swelling remained unchanged. There was a 2-year history of right leg numbness and a 4-month history of urinary dysfunction (straining during micturition). She developed a nonhealing ulcer on the right foot and gangrene of a great toe, which underwent autoamputation.

Examination. We noted, in the lumbar region, a 5-cm-diameter swelling covered with a tuft of hair. Sensory loss was 50% in the L4–S2 dermatomes, but no motor deficit was observed. Computed tomography and MR imaging of the spine revealed L2–5 spina bifida with an L2–3 osseous spur. The cord was split at L-2 and L-3 and the cord re-united at the lower border of L-4. The terminal cord was tethered to a subcutaneous lipoma (Fig. 1). We performed
a microsurgical excision of the meningocele and osseous spur, cord detethering, and duraplasty. In the subcutaneous lipomatous tissue there was a small mass with hemorrhage on its surface. This mass and the subcutaneous lipoma were resected, and tissue specimens were sent for histopathological examination.

Histopathological Examination. Grossly, the surgical specimen measured $3 \times 2 \times 1.5$ cm and was composed of firm nodular fibrous tissue and an osseous spur. This nodular mass was embedded in the lipomatous tissue lying outside the meningocele. Once the surface of the globular mass was cut, we observed several areas of hemorrhage. Microscopic examination of the globular mass showed an infantile uterus-like structure consisting of smooth-muscle bundles and endometrial tissue. At places these glands were surrounded by smooth-muscle bundles. The glands were lined by columnar-to-cuboidal epithelium, and some contained hemosiderin-laden macrophages suggestive of old hemorrhage (Fig. 2).

Based on these features, a final diagnosis of TCS with a uterus-like structure of müllerian origin was made.

Case 2

History. This 24-year-old woman presented with a 4-year history of low-back pain. The pain radiated to both lower limbs and was accompanied by paresthesia, numbness, and weakness of both lower extremities. The pain worsened during menstruation.

Examination. We noted a sensory loss of 50 to 90% from the T-12 level and below. There was wasting of the bilateral calf muscles, and hip and knee strength was Grade 4+/5. Magnetic resonance imaging showed enlargement of the conus medullaris with an oval well-defined space-occupying lesion, which was hyperintense on T1-weighted images but with a hypointense periphery on T2-weighted images (Fig. 3). Based on the MR imaging findings, we established a diagnosis of an intraconus lipoma. Through an L-3 laminectomy, we excised the terminal filum and conus medullaris lesion. On cutting open the lesion, we observed dark brown thick fluid; the possibility of dermoid cyst was considered.

Histopathological Examination. The cut surface of the intraconus mass was dark brown with cystic areas that were well demarcated from the neural tissue of the conus medullaris. Microscopic examination revealed well-formed endometrial tissue consisting of endometrial glands and stroma (Fig. 4). This tissue was well demarcated from the neural tissue. In addition, glands surrounded by smooth-muscle bundles were also seen. No immature tissue or other derivatives of ectoderm or endoderm.
were identified. Examination of a specimen of the osseous spur showed cartilaginous tissue only.

Based on these features, a final diagnosis of TCS with a uterus-like structure of müllerian origin was made.

**Discussion**

Tethered cord syndrome is a condition in which there is a low-lying conus medullaris and a thickened terminal filum and/or lipoma. The most common associated vertebral anomaly is spina bifida. Various skin lesions have been described including a hemangioma, skin dimple, tuft of hair, hypertrichosis, subcutaneous lipoma, and associated imperforate anus.

The presence of tissue of müllerian origin has been described in four other cases (Table 1). Young and Feder reported the first case of spinal dysraphism in a patient in whom they found endometrial tissue in the subcutaneous tissue of the lumbosacral region. Several decades later, Kurman et al. reported a similar case. In a case reported by Molleston et al., endometrial tissue was present subcutaneously and extended into the conus medullaris. The features in all three of these cases were similar and all were associated with spina bifida. Because only endometrial tissue was documented histologically, a diagnosis of endometriosis was considered in these cases. Rougier et al. reported the case of an 18-year-old woman who presented with paresthesia and weakness of the left leg, spinal dysraphism, and an intramedullary mass. Pathological examination revealed smooth-muscle bundles with interspersed endometrial tissue and evidence of old hemorrhage. The features in our Case 1 are similar to the case described by Rougier et al.; we observed a uterine-like structure with evidence of old hemorrhage suggestive of bleeding during menstruation. However, the unique features in Case 1 were the split cord malformation, osseous spur, and associated spina bifida. In Case 2, there was well-formed uterus-like structure with distinct separate endometrium and myometrium.

The histogenesis of this type of lesion in the lumbosacral region is debatable. The term choristoma was proposed by Kurman et al. to indicate the presence of this type of müllerian-origin tissue. A choristoma is defined as “a mass of tissue histologically normal for an organ or part of the body other than the site at which it is located.” It should be differentiated from ectopia, a condition in which an organ or tissue is present in an abnormal position but absent at its normal location. In each of our cases the uterus was present in its normal location. The possibility of teratoma was excluded because other derivatives of germ layers were absent.

The embryological development of the distal part of the spinal cord is different from that of the rostral portion. The spinal cord’s caudal end undergoes dedifferentiation and redifferentiation. Some authors have described the loss of the caudal portion of the spinal cord and preformed ganglia, which are necessary for the normal development of the conus medullaris and terminal filum. The disturbance of this normal ontogenetic process may lead to displacement of mesenchymal or müllerian-origin tissue within the spinal cord. Schwartz proposed that the distal portion of the spinal cord develops from the Hensen node (also known as the primitive node) and that the existence of some pluripotent cell rests might be a source of these types of nonneoplastic or neoplastic conditions.

Although rare, this differential diagnosis of müllerian-origin choristoma should be considered by neurosurgeons when evaluating patients with adult TCS and a cystic mass in the conus medullaris. The tissue excised from the tethered cord should be examined carefully because this lesion can be the cause of spinal cord tethering.

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**TABLE 1**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
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<th>Sex</th>
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<th>Associated Anomalies</th>
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<td></td>
<td>subcutaneous mass</td>
<td>spina bifida</td>
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<tr>
<td>Kurman et al., 1969</td>
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<td>spina bifida, tethered cord</td>
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<td></td>
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<td>tethered cord</td>
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<td>present cases</td>
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<td>24, F</td>
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<td>low-back pain; numbness &amp; weakness in both lower limbs</td>
<td>tethered cord</td>
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**FIG. 4.** Case 2. Photomicrographs showing endometrial glands with stroma (A) and evidence of old hemorrhage in the form of hemosiderin (B), as well as smooth-muscle bundles and thick-walled vessels (C). H & E, original magnification × 100.
References


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