35 years old male patient presented clinically with progressive lower limb weakness and impotence of 3 years duration, disturbance of micturation with few drops of urine passing without the patient control was a prominent symptom. On examination the patient had atrophy of the anterior tibial group of muscles on the right side, with bilateral spasticity in both lower limbs and a sensory level at D8 spinal segment (D4-D5 vertebral segments). Reflex examination showed lost ankle reflexes in both lower limbs and diminished knee reflex on the left side while the knee reflex on the right side was exaggerated. Bilateral extensor planter responses were also noted. No other abnormalities were noted and the upper limbs were free. Clinical diagnosis of long spinal lesion extending from D8 spinal segment to the conus /epiconus regions was made.

**RADIOLOGICAL FINDING**

**Figure 1.** Intramedullary ependymoma of the conus medullaris and filum terminale. A,B,C, The tumor is cystic with intramedullary transverse bands and thin walls. The tumor extends from D10-D12. D, Dilatation of the central canal of the spinal cord rostral to the tumor mass, the spinal cord is probably atrophic at the region of dilatation of the central canal, notice absence of transverse bands inside the dilated central canal. Also notice an extramedullary long space occupying lesion mildly pushing the spinal cord anteriorly and has the same CSF signal intensity on both
T1,T2. (arachnoid cyst). The associated arachnoid cyst could be an incidental asymptomatic radiological finding or could be secondary to the spinal neoplasm. Some degree of enhancement is seen in the tumor walls after contrast injection (C).

Figure 2. Intramedullary cystic ependymoma with intracystic transverse bands. Notice an extramedullary extradural mass of the same CSF signal intensity pushing the spinal cord anteriorly and flattening its posterior surface (arachnoid cyst).

Figure 3. Intramedullary crescentic hypointense lesions on the T2 image. These hypointense lesions probably represent hemosiderin deposition secondary to tumor bleeding, which is common in ependymomas.
DIAGNOSIS:

- INTRAMEDULLARY CYSTIC EPENDYMOMA (MYXOPAPILLARY EPENDYMOMA) OF THE CONUS / EPICONUS REGIONS AND FILUM TERMINALE (EXTENDING FROM D10-D12 SPINAL SEGMENTS).
  - SECONDARY DILATATION OF THE CENTRAL CANAL OF THE SPINAL CORD EXTENDING FEW SPINAL SEGMENTS ABOVE THE TUMOUR MASS WITH SPINAL CORD ATROPHY.
  - POSSIBLE RETROMEDULLARY MULTISEGMENTAL EXTRADURAL ARACHNOID CYST PUSHING THE SPINAL CORD ANTERIORLY.

DISCUSSION

- Spinal ependymoma
  - Prevalence

Ependymoma is the most common intramedullary spinal neoplasm in adults, accounting for up to 60% of all glial spinal cord tumors (19). From the four largest studies of patients with spinal cord ependymomas reported in the literature (5,19,24,25), the following demographic information emerges: These lesions tend to manifest in young adulthood, with a mean age at presentation of 38.8 years and are more common in male patients (57.4%). Cord ependymomas occur most commonly in the cervical region, with 44% involving the cervical cord alone and an additional 23% extending into the upper thoracic region. About 26% are located in the thoracic cord alone. Only 6.5% involve either the distal thoracic cord or the conus medullaris (5,19,24,25). Myxopapillary ependymoma, a variant type, is, in rare cases, found in the subcutaneous tissue of the sacrococcygeal region, usually without any connection with the spinal canal (26). It is believed that these arise from either heterotopic ependymal cell rests or vestigial remnants of the distal neural tube during canalization and retrogressive differentiation (26,27).

  - Clinical Presentation

As with most primary intramedullary tumors, there is frequently a long antecedent history before the diagnosis of an intramedullary ependymoma is established. The mean duration of symptoms was 36.5 months for the 183 patients from the four largest studies (5,19,24,25). A large majority of patients with spinal cord ependymomas have relatively mild clinical symptoms. Most of the 183 patients (81%) could walk without assistance at presentation (5,24,25). In general, the less preoperative neurologic deficit existing at presentation, the better the postoperative outcome (5,25). Typically, patients initially present with mild symptoms, and there is no objective evidence of neurologic deficits, which often leads to a delay in diagnosis (5,19,24,25). Some ependymomas may even be a source of subarachnoid hemorrhage (28,29).

At diagnosis, patients with spinal cord ependymomas typically have back or neck pain (67%), sensory deficits (52%), motor weakness (46%), or bowel or bladder dysfunction (15%) (5,19,24,25). The predominance of sensory symptoms (85% of patients with pain and other sensory deficits combined) may be directly related to the more central location of these tumors (5). Spinal cord ependymomas are believed to arise from ependymal cells that line the central canal. Theoretically, this central location makes it likely that the crossing spinothalamic tracts will be compressed or interrupted. Dominant motor symptoms are commonly associated with very large ependymomas and a poorer postoperative outcome secondary to the increased surgical risk associated with resection of these larger lesions (5). Hoshimaru et al (25) found that patients with a shorter duration of symptoms tended to have a better postoperative outcome. Lesions of the thoracic cord are associated with poorer surgical outcomes, perhaps because
of its relatively tenuous vascular supply, compared with lesions of the cervical spinal cord. This was especially true in patients with evidence of arachnoid scarring or cord atrophy at surgery (4).

Intramedullary ependymomas are characterized by slow growth and tend to compress adjacent spinal cord tissue rather than infiltrate it. Accordingly, there is almost always a cleavage plane, which facilitates microsurgical resection, the treatment of choice (5,19,24). Patients frequently have worsened symptoms in the immediate postoperative period secondary to edema and possibly transient interference with spinal cord blood flow (5,25,30). Postoperative radiation therapy is reserved for recurrent disease, which is much more commonly seen in cases of subtotal resection (5,25,30). The patient's preoperative neurologic status is the most important predictor of outcome (5,19,24). Earlier surgical resection is associated with fewer and less severe neurologic deficits. Recurrence is substantially reduced when a complete gross total resection can be performed (5,19,30). The 5-year survival rate for patients with spinal cord ependymomas is approximately 82%, regardless of how severe the preoperative neurologic deficits. The 20-year survival rate, however, is much worse for patients who present with a major neurologic dysfunction (50%) than for patients with a minor neurologic impairment (33%) (1). The lungs, retroperitoneum, and lymph nodes are the most common extraspinal sites of metastatic spread (31).

- **Pathologic Characteristics**

Most ependymomas displace rather than infiltrate adjacent neural tissue. Because ependymomas are believed to arise from ependymal cells of the central canal within the spinal cord, symmetric cord expansion is the rule. These soft, friable, well-margined lesions are frequently gray and have associated syringomyelia (32). Polar cysts are a common finding (62% in the study by Brotchi and Fischer [24]). True tumoral cysts are less common (22% of cases) (24). Small feeding vessels at the ventral surface are commonly noted at surgical resection (5,25). The myxopapillary variant is virtually always located along the filum terminale with occasional extension to the conus medullaris and may appear as a soft, tannish "bag" of tissue (32).

Uniform, moderately hyperchromatic nuclei are typical findings at histologic examination (32). Six histologic types are recognized: cellular (the classic and most common type), papillary, clear cell, tanycytic, myxopapillary, and melanotic (the least common type) (32). Perivascular pseudorosettes are virtually required to establish the diagnosis of ependymoma but may be less conspicuous in less cellular types of ependymomas. With use of the World Health Organization (WHO) classification (24), almost all spinal cord ependymomas can be classified as either grade I or grade II. Malignant types are rare (32). Cystic degeneration is seen in 50% of cases, and hemorrhage is common (especially at the superior and inferior margins of the tumor). In contrast to intracranial ependymomas, calcification is uncommon.

Rarely, a cord ependymoma may extend exophytically and present a diagnostic challenge (8). Ependymomas may even arise outside the CNS (sacroccygeal region, broad ligament of the ovary). Up to one-third of these ectopically located ependymomas are associated with spina bifida occulta (33).

Results of recent investigations reveal mutations of the type 2 neurofibromatosis transcript in some cases of sporadic spinal cord ependymomas that occurred in patients without type 2 neurofibromatosis. These changes have not been observed in intracranial ependymomas and, therefore, are suggestive of a different molecular basis for ependymomas that arise in the spinal cord (34).

- **Imaging Characteristics**

Radiographs of patients with ependymomas may reveal scoliosis (16% of cases) or canal widening (11%) with associated vertebral body scalloping, pedicle erosion, or laminar thinning (1). Conventional myelography frequently reveals either a complete or partial block in the flow of contrast material (1). At unenhanced CT, ependymomas are either isoattenuated or slight hyperattenuated compared with the normal spinal cord (1). Ependymomas enhance intensely after intravenous administration of iodinated contrast material. CT myelography shows nonspecific cord enlargement (1).

Most spinal cord ependymomas are iso- or hypointense relative to the spinal cord on T1-weighted MR images.
In rare cases, they may manifest as a hyperintense mass, usually secondary to the effects of hemorrhage (24,35,36). On T2-weighted images, the lesions are typically hyperintense relative to the spinal cord (35,36), although in the single largest review of spinal ependymomas, isointense tumors were as common as hyperintense tumors (24). About 20%—33% of ependymomas demonstrated the "cap sign," a rim of extreme hypointensity (hemosiderin) seen at the poles of the tumor on T2-weighted images. This finding is thought to be secondary to hemorrhage, which is common in ependymomas and other highly vascular tumors (eg, paraganglioma, hemangioblastoma) (16,35). Most cases (60%) also showed evidence of cord edema around the masses (24). The average number of vertebral segments involved with abnormal signal intensity is 3.6; however, some ependymomas may involve as many as 15 segments (19,24,25,35,36). Despite the theoretic support for cord ependymomas having a central location on the basis of the presence of ependymal cells in the central canal, only 62.5%—76% of these tumors have been reported to arise from a central location (16,35).

Cysts are a common feature, with 78%—84% of ependymomas having at least one cyst, most of which are the nontumoral (polar) variety (24,35,36). The prevalence of tumoral cysts appears to be more variable (4%—50% of cases) (24,35,36). Syringohydromyelia is also quite variable in previously published reports, occurring in 9%—50% of cases (24,36). Analysis in one published case revealed that the syringomyelia fluid had a protein content consistent with that of an exudate, which supports the hypothesis that the cavity resulted from a disruption of the blood-brain barrier (37). When the data from the three largest studies evaluating contrast enhancement on MR images are combined, the vast majority of spinal cord ependymomas (84%) enhanced to at least some degree following the intravenous administration of gadolinium-based contrast material and even more (89%) had well-defined margins on the contrast-enhanced images (5,24,36).

60% of intramedullary tumors, which tend to be primary (ependymomas, astrocytomas, hemangioblastomas) rather than secondary, include a syringomyelic cavity. The cavity is located at the extremities of the tumor (above and/or below) and is probably created by tumoral fluid secretion with secondary dilation of the ependymal canal, although CSF circulatory disorders induced by the tumor may also be responsible. A syringomyelic cavity must be distinguished from a possible intratumoral cystic component (which enhances after gadolinium injection), and from tumor-associated intramedullary edema (T2 hyperintense and T1 hypointense, but less so than CSF). This distinction is important because a syringomyelic cavity does not need to be surgically removed and generally regresses after tumor ablation. (43)

Intraoperative ultrasonography reveals ependymomas as regions of sharply defined uniform echogenicity. Cysts are easily seen with this modality (5).

- **Myxopapillary Ependymoma**

A special variant of ependymoma, the myxopapillary ependymoma constitutes about 13% of all spinal ependymomas (38). This tumor tends to have an earlier clinical presentation (mean age, 35 years) and is more commonly seen in male patients. These mucoid tumors have a distinct predilection for the conus medullaris or filum terminale and are thought to arise from the ependymal glia of the filum terminale. Consequently, myxopapillary ependymomas are the most common neoplasm (83% of cases) in this region (38). Occasionally, they occur in the extradural space, probably arising from the coccygeal medullary vestige at the distal portion of the neural tube (39). Multiple lesions have been variably reported in about 14%—43% of cases (39).

In this particular patient the tumor involved few spinal segments at the conus region, that is probably responsible for the lower limb neurological manifestations, with atrophy, diminished reflexes etc. The higher sensory level is probably caused by the dilatation of the central canal of spinal cord that extended rostrally for 4 to 5 spinal segments above the tumor mass. The tumor itself was cystic, however the dilatation of the central canal of spinal cord above the tumor mass is not a part of the tumor, it is rather secondary to CSF flow obstruction secondary to the tumor and in this respect it is similar to obstructive hydrocephalus. It is also possible that the multisegmental retromedullary arachnoid cyst is also implicated in symptom formation in so far as the higher sensory level is concerned as it was seen exerting mass effect on the spinal cord (which is pushed forward by the arachnoid cyst with flattening of its posterior border). The arachnoid cyst extended many spinal segments above the tumor mass.
Myxopapillary ependymomas usually manifest with lower back, leg, or sacral pain and weakness or sphincter dysfunction. Although most of these tumors are slow growing, those located near the sacrum may be more aggressive, creating large, lytic areas of bone destruction (27,40,41).

Myxopapillary ependymomas are characteristically lobulated, soft, sausage-shaped masses that are often encapsulated (38). The histologic hallmark is heterogeneity, resulting from generous mucin production and papillary zones mixed with cellular areas composed of rosettes and pseudorosettes (38).

Myxopapillary ependymomas have a nonspecific radiologic appearance and are typically isointense relative to the spinal cord on T1-weighted MR images and hyperintense on T2-weighted MR images. Hyperintensity on both T1- and T2-weighted images may be noted occasionally, a finding that reflects mucin content or hemorrhage (36). Superficial siderosis may be seen but is not specific, as it has been noted in association with other highly vascular tumors (42). Enhancement is virtually always seen after the intravenous administration of contrast material. The predilection of these tumors for the conus medullaris should be suggestive of the diagnosis (39).

Radiologically, astrocytoma is a close mimicker of ependymoma. Ependymomas are central in location since they arise from central canal ependymal cells, whereas astrocytomas are eccentric with infiltrating borders as they originate from the cord parenchyma. Findings such as hemorrhage within the tumor and hemosiderin deposition or calcification are more frequent in ependymomas due to rich connective tissue stroma. Contrast enhancement is also intense and homogenous in ependymomas whereas it is patchy and irregular in astrocytomas. Ependymomas especially the myxopapillary variety have particular predilection for conus medullaris which is not so in the case of astrocytomas. Ependymomas can be differentiated from hemangioblastoma by virtue of enhancing mural nodule, which is the hallmark of hemangioblastoma. Congenital syringomyelia should be ruled out because of the location of the cavitations (Congenital syringomyelia is more common in the cervical region) and because of absence of Chiari malformation at the cranio-cervical junction. (43)

Rostral and caudal cysts (syrinxes) usually are associated with intramedullary tumors of all histologic types, whereas intratumoral cysts also are frequent. Syrinxes reflect a reactive process within the spinal cord, do not contain neoplastic cells, have gliotic linings, are filled with fluid similar to CSF, rarely are proteinaceous or hemorrhagic, and do not need to be resected but merely drained at surgery. Tumor cysts generally are smaller, may have irregular walls, and are eccentric in position within the cord. They are lined by abnormal glia and are xanthochromic or blood-filled.

The associated arachnoid cyst could be an incidental asymptomatic radiological finding and might be secondary to the spinal neoplasm. Arachnoid cysts usually occur in association with normal arachnoid cisterns, and such cysts are congenital, arising from arachnoid clefts and arachnoid duplications. Glioependymal cysts are rare; only a few instances of interhemispheric glioependymal cysts are known. Glioependymal cysts may be associated with agenesis of the corpus callosum, heterotopia, and other dysplasias.

Most likely, arachnoid cysts expand when hydrodynamic pulse waves of cerebrospinal fluid (CSF) become entrapped in arachnoid locations. The cysts may be unilocular or loculated by septations. The wall of the cyst is usually smooth. Most cysts are filled with clear colorless fluid of low protein content comparable to CSF. A few cysts may contain elevated protein content. Gross appearances of glioependymal cysts usually are indistinguishable from arachnoid cysts; however, their microscopic appearances vary. Glioependymal cysts are epithelial lined and may bear cilia, but only glial tissue is seen lining the lesion if the epithelial lining becomes dehiscent or lost. Glioependymal cysts are believed to derive from displaced neuroectodermal tissue. Histologic classification is only of systematic interest and has little bearing on prognosis.

Acquired arachnoid cysts may develop following surgery, trauma, subarachnoid hemorrhage, neonatal infections and can occasionally occur in association with extra-axial or intramedullary neoplasm. Arachnoid cysts associated with tumors develop as a consequence of CSF loculation surrounded by arachnoid scarring, with expansion of osmotic filtration or via a ball-valve mechanism. (arachnoid scarring in this case is probably secondary to arachnoiditis induced by CSF seeding of the primary neoplasm, which is common in ependymoma, the resulting arachnoid cysts frequently are associated with syrinx formation and/or cord atrophy). These acquired arachnoid
cysts have been described variably as acquired secondary or leptomeningeal cysts. The reason arachnoid cysts grow and become space occupying is far from clear. No inner lining is present through which active transport can take place. Neurosurgeons have observed ostia with pulsating fluid in exposed cysts suggesting a hydrodynamic flap-valve or ball-valve mechanism.

Table 1. Pathological findings secondary to spinal ependymoma and not considered a part of it

<table>
<thead>
<tr>
<th>Pathological finding</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rostral and caudal cysts (syrinxes)</td>
<td>Usually are associated with intramedullary tumors of all histologic types. These syringes reflect a reactive process within the spinal cord, do not contain neoplastic cells, have gliotic linings, are filled with fluid similar to CSF, rarely are proteinaceous or hemorrhagic, and do not need to be resected but merely drained at surgery. Tumor cysts generally are smaller, may have irregular walls, and are eccentric in position within the cord. They are lined by abnormal glia and are xanthochromic or blood-filled.</td>
</tr>
<tr>
<td>Arachnoiditis, arachnoid cysts and spinal cord atrophy</td>
<td>Arachnoid cysts associated with tumors develop as a consequence of CSF loculation surrounded by arachnoid scarring, with expansion of osmotic filtration or via a ball-valve mechanism. (arachnoid scarring in this case is probably secondary to arachnoiditis induced by CSF seedling of the primary neoplasm, which is common in ependymoma, the resulting arachnoid cysts frequently are associated with syrinx formation and/or cord atrophy)</td>
</tr>
</tbody>
</table>

• Addendum
  - A new version of this PDF file (with a new case) is uploaded in my web site every week (every Saturday and remains available till Friday.)
  - To download the current version follow the link "http://pdf.yassermetwally.com/case.pdf".
  - You can also download the current version from my web site at "http://yassermetwally.com".
  - To download the software version of the publication (crow.exe) follow the link: http://neurology.yassermetwally.com/crow.zip
  - The case is also presented as a short case in PDF format, to download the short case follow the link: http://pdf.yassermetwally.com/short.pdf
  - At the end of each year, all the publications are compiled on a single CD-ROM, please contact the author to know more details.
  - Screen resolution is better set at 1024*768 pixel screen area for optimum display
  - Click here for an archive of the previously reported cases in downloadable PDF files.
  - For an archive of the previously reported cases go to www.yassermetwally.net, then under pages in the right panel, scroll down and click on the text entry "Downloadable case records in PDF format" and "Downloadable short cases in PDF format"

REFERENCES

References


36. Kahan H, Sklar EML, Post MJD, Bruce JH. MR characteristics of histopathologic subtypes of spinal


43. Metwally, MYM. Textbook of neuroimaging, CD-ROM based publication, in Metwally (ed), WEB-CD agency for electronic publication, version 81a ; 2007