Spinal intramedullary ependymal cyst and tethered cord in an adult

Case report

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Spinal intramedullary ependymal cysts are extremely rare. Fourteen cases have been reported in the literature, and only one was associated with another congenital lesion. The authors describe the case of a 33-year-old man in whom an intramedullary ependymal cyst and filar lipoma were present. These lesions were diagnosed using magnetic resonance imaging and were treated surgically. Pathological examination confirmed the diagnosis. At 6 months postsurgery, there was no evidence of recurrence.

KEY WORDS • ependymal cyst • intramedullary cyst • filar lipoma • spinal cord

S PINAL intramedullary ependymal cysts are rare congenital lesions and only 14 cases have been pathologically proven to date. Their association with a concomitant congenital lesion, such as a tethered cord, is even more rare. We describe the case of an adult male in whom an intramedullary cyst and filar lipoma caused tethering of the cord.

Case Report

Examination. This 33-year-old man presented with a 2-month history of gradually increasing back pain and lower-extremity paresthesias. Neurological examination showed decreased touch and pain sensation in his legs. Motor weakness and reflex changes were absent. Spine MR imaging showed a well-circumscribed, 26 × 13-mm intramedullary oval cystic lesion at L-3; the lesion was hypointense on T1- and hyperintense on T2-weighted images without contrast agent administration. There was also a filar lipoma associated with cord tethering. An additional intradural cystic lesion was observed at S-3 (Fig. 1).

Operation. The patient underwent an L3–5 laminectomy. The dura was opened under the microscope. The cord was expanded. A 10-mm midline myelotomy was made in the laminectomy region. A cyst cavity filled with clear fluid was encountered. The cyst could not be totally removed because there was no cleavage line between the cystic cavity and spinal cord. Thus, a portion of the cyst wall was obtained for biopsy examination, and adequate communication was created between the cyst and the subarachnoid space. Using an ultrasonic aspirator, the lipoma was separated from the roots of the cauda equina, and the terminal filum was sectioned.

Histopathological Examination. The cyst wall lining was composed of either columnar or cuboidal ciliated and nonciliated epithelium that lacked a basement membrane and rested on fibrous tissue (Fig. 2).

Postoperative Course. After surgery the patient’s symptoms improved gradually. He was discharged on the 7th postoperative day.

Discussion

Intramedullary ependymal cysts of the spine are very rare. In the 14 reported cases, each had only a single ependymal cyst. Only one was present in association with a filar lipoma–related tethered cord in an infant. Our case shares certain rare features with the patient described by Balasubramaniam, et al. To date, there have been no reports of both intramedullary ependymal cyst and filar lipoma in an adult.

The association of the ependymal cyst with other intraspinal anomalies suggests that the basic cause is dysembryogenesis. It has been believed that ependymal cysts originate in the neural tube wall after ectopic displacement. The most widely accepted hypothesis is that the floor plate of the neural tube is evaginated on the anterior side, becomes isolated, and later develops into a cyst. In our case, we believe that the tethered spinal cord and/or the location/presence of the lipoma may have caused the cyst to form.

On histological examination, the ependymal cyst is characterized by a lining of epithelium, either columnar or cuboidal, with or without cilia, that lacks a basement mem-
brane and rests on fibrous tissue. Some pseudostatification may be seen.

In the reported cases the patients ranged in age from 1 to 71 years, and there was no sex preference (seven males and seven females). The lesion was most commonly localized at the conus medullaris. Including our case, the conus medullaris was involved in eight patients.

Clinical signs and symptoms depend on the lesion’s location. The clinical presentation varied from progressive weakness, radicular pain, and paresthesia to tetraparesis. Magnetic resonance imaging is the best modality with which to diagnose and differentiate intramedullary cystic lesions. The MR imaging features of the cyst are a well-defined lesion, sharply delineated from the cord and isointense with cerebrospinal fluid on T₁- and T₂-weighted MR images and proton density images without administration of contrast material. The cyst is usually found in the anterior aspect of the cord, does not communicate with the central canal, is located off center, and should be differentiated from syringomyelia or a terminal ventricle.

Surgery seems to be the optimal treatment in patients with related symptoms. The goals of surgery are to create a communication between the cyst cavity and the subarachnoid space and to obtain a biopsy sample from the cyst wall. The communication is achieved by means of fenestration or marsupialization. In several reported cases a cystosubarachnoid shunt was placed. We chose to avoid this therapy because of the high possibility that the route of communication would become blocked.

Because ependymal cysts may recur, it is necessary that patient undergo thorough follow-up examinations. The guidelines for the follow-up interval and imaging studies have not been clearly defined in the literature; we performed follow-up MR imaging 6 weeks after surgery to allow for comparison of these studies with others obtained at later follow-up intervals. After 6 weeks, an imaging study really only needs to be performed when signs of deterioration or cessation of improvement appear. We are planning to conduct follow-up studies in our patient periodically for at least one year.

In summary, our case represents a very rare condition. The best method for diagnosing spinal intramedullary ependymal cysts is with MR imaging, and surgery seems to be the optimal treatment in symptomatic patients.

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Fig. 1. Sagittal T₁-weighted (A), sagittal T₂-weighted (B), and axial T₁-weighted (C) MR images showing the intramedullary cyst and the tethering due to a filar lipoma.

Fig. 2. Photomicrograph of the specimen demonstrating columnar epithelial cells lining the neuroglial stroma. H & E, original magnification × 200.
References


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