Spinal cord ependymoma

Epidemiology

Intramedullary ependymoma is a rare neoplasms, comprising approximately 5% of all CNS tumors and 15% of all spinal cord tumors.

Ependymomas are the most frequent spinal cord tumors in adult patients.

Some authors believe it is more common in the fourth and fifth decade while others propose a wider distribution spanning between the second and sixth decade of life [1) 2) 3) 4) 5)]

In a population-based survey of 467 patients with primary intraspinal neoplasms, intramedullary ependymomas accounted for 34.5% of all ependymomas of the central nervous system. According to the same study, the age-adjusted incidence rate for the primary intraspinal neoplasm is 0.5 in females and 0.3 in males per 100,000 population per year [6)].

Spinal ependymomas commonly present as intramedullary tumors in the cervical or thoracic cord or as tumors arising from the conus medullaris or the filum terminale

Sometimes the tumor is located outside of the spinal cord and affects the cauda equina nerve roots.

Classification

Spinal anaplastic ependymoma, spinal extramedullary ependymoma are rare.

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see Spinal myxopapillary ependymoma.

see Spinal tanycytic ependymoma.

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see High cervical spinal cord ependymoma

see Cervical spinal cord ependymoma

see Thoracic spinal cord ependymoma

Dissemination

Spinal cord ependymomas presenting with regional dissemination along the neuroaxis are rare, with a yet undetermined standard of care.

Diagnosis

Plain radiograph

Plain film features that may be seen with a spinal ependymoma include:

scoliosis

spinal canal widening

vertebral body scalloping

pedicle erosion

laminar thinning

CT

CT may demonstrate:

non-specific canal widening

iso to slightly hyper-attenuating compared with normal spinal cord

intense enhancement with iodinated contrast

large lesions may cause scalloping of the posterior vertebral bodies and neural exit foraminal enlargement

MRI

MRI is the modality of choice for evaluating suspected spinal cord tumours. Features include:

widened spinal cord (as ependymomas arise from ependymal cells lining the central canal, they tend to occupy the central portion of the spinal cord and cause symmetric cord expansion)

although unencapsulated, they are well-circumscribed

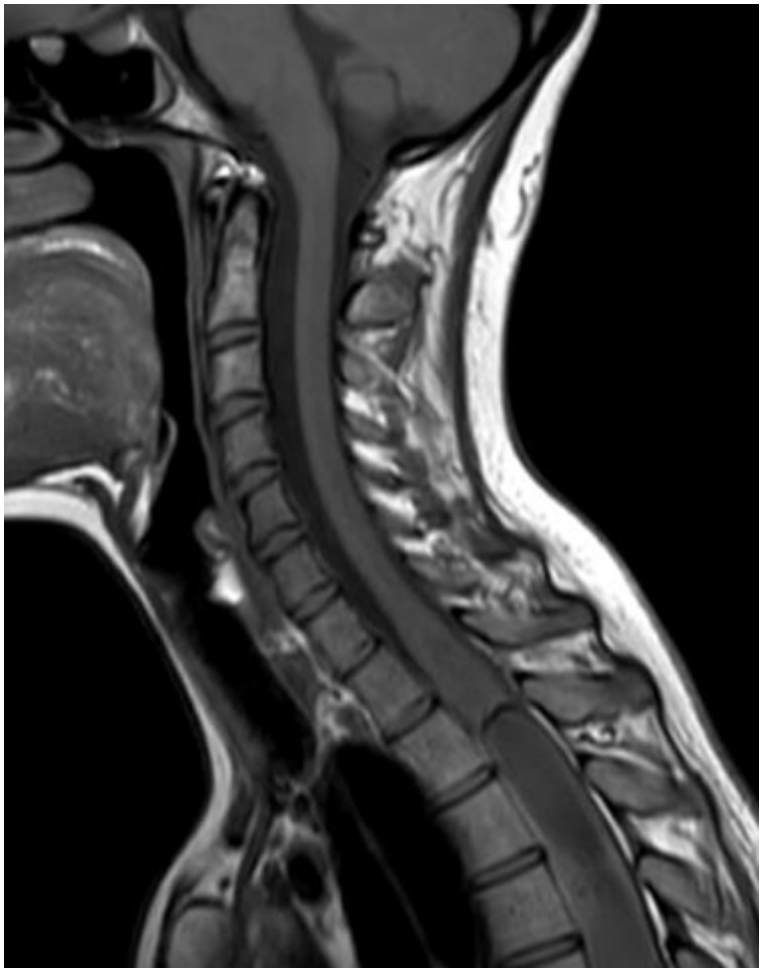
tumoral cysts are present in 22%. Non-tumoral cysts are present in 62%.

Syringohydromyelia occurs in 9-50% of cases

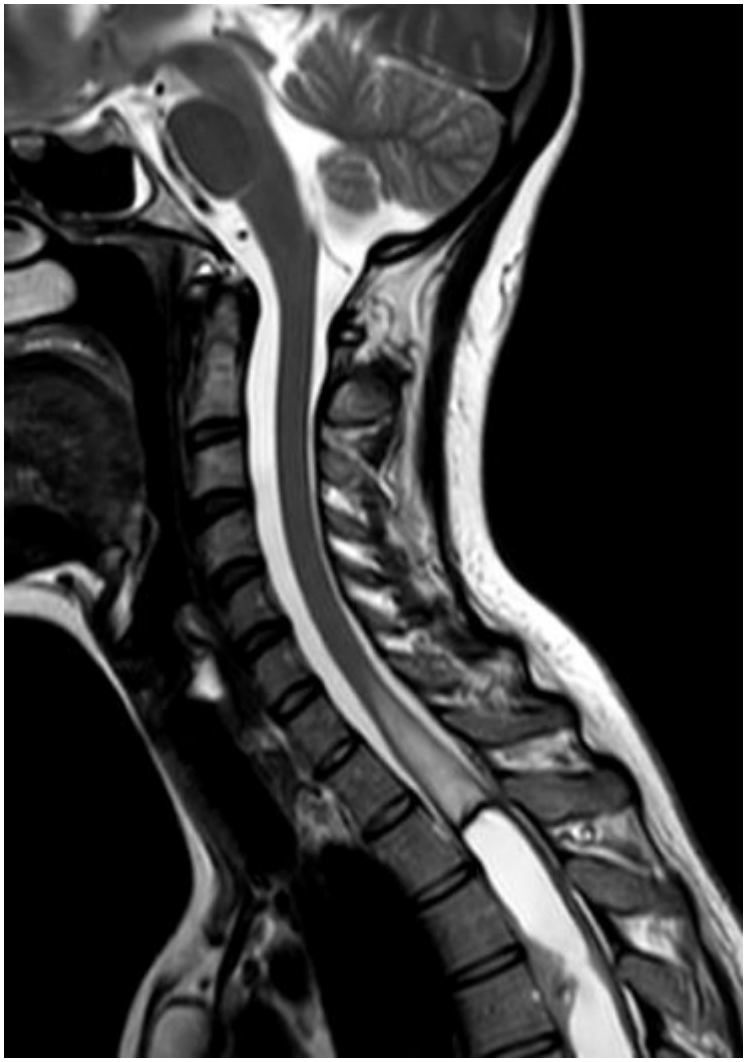
In contrast to intracranial ependymomas, calcification is uncommon

average length of four vertebral body segments

Typical signal characteristics:



T1: most are isointense to hypointense; mixed signal lesions are seen if cyst formation, tumour necrosis or haemorrhage has occurred.



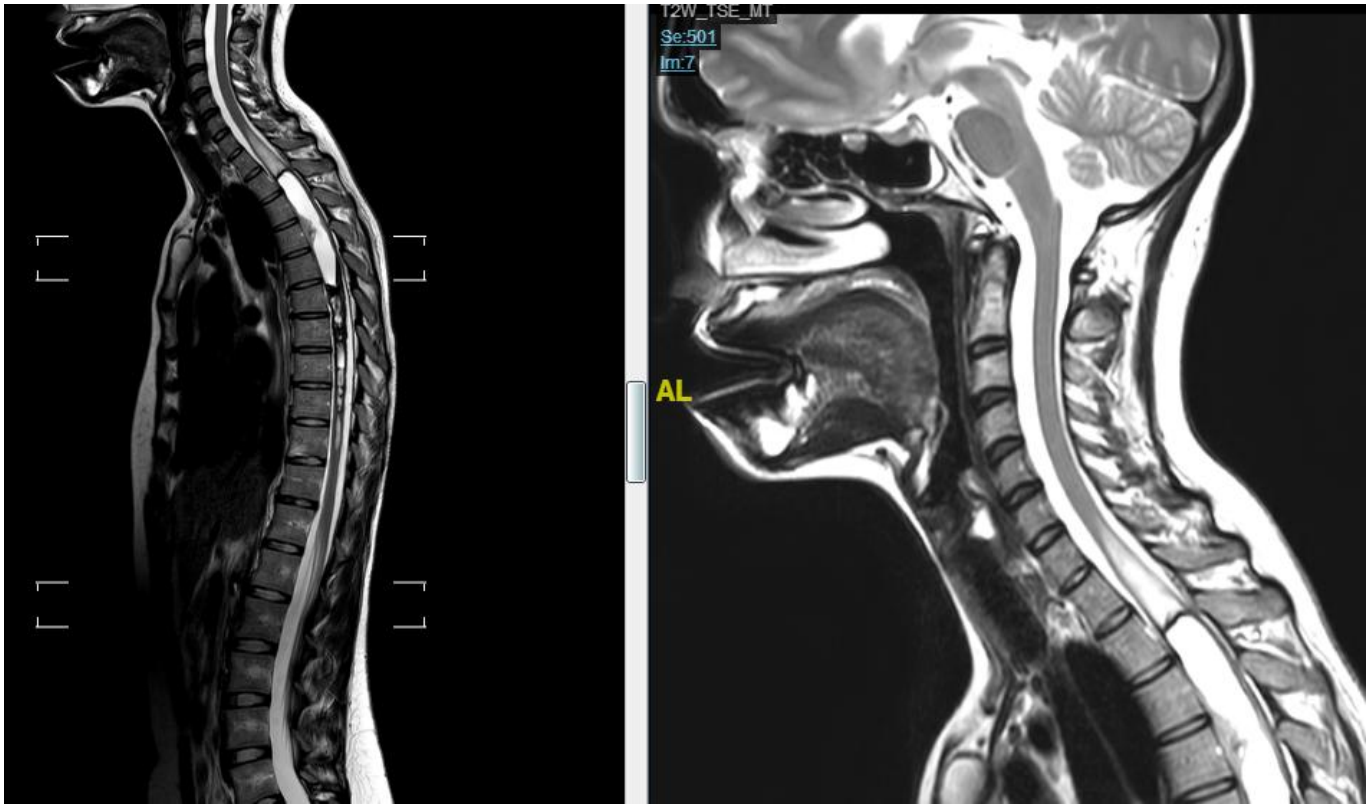
T2: hyperintense

peritumoural oedema is seen in 60% of cases

associated haemorrhage leads to the “cap sign” (a hypointense haemosiderin rim on T2 weighed images) in 20-33% of cases.

The cap sign is suggestive of but not pathognomonic for ependymoma as it may also be seen in Spinal cord hemangioblastomas and paragangliomas.

T1 C+ (Gd): virtually all enhance strongly, somewhat inhomogeneously.



Hypointensity at the tumor margin was found to be a relatively firm pseudocapsule, and hypointensity within the tumor corresponded to intratumoral hematoma. All of the tumors with hypointensity were ependymomas at histologic examination. When MR imaging shows an intramedullary tumor with hypointensity at the tumor margin, it is suggestive, but not pathognomonic, of an ependymoma [7].

Differential diagnosis

see Intramedullary ependymoma differential diagnosis

Treatment

see Spinal cord ependymoma treatment.

Complications

Despite their usually well-defined dissection plane, surgical morbidity has been documented to be considerably higher compared with other intramedullary entities.

Although surgery was once reserved for diagnosis alone, the evolution of surgical practices has elevated resection to the treatment of choice for these lesions. While technological advances continue to improve the capacity for gross-total resections and thus decrease the risk of recurrence, ependymoma spinal surgery still contains a variety of potential complications. The presence of neurological deficits and deterioration are not uncommonly associated with spinal cord ependymoma surgery, including sensory loss, dorsal column dysfunction, dysesthetic syndrome, and bowel and bladder dysfunction, particularly in the immediate postoperative period. Surgical treatment may also lead to wound complications and CSF leaks, with increased risk when radiotherapy has been involved. Radiation therapy may also predispose patients to radiation myelopathy and ultimately result in neurological damage. Additionally, resections of spinal ependymomas have been associated with postoperative spinal instability and deformities, particularly in the pediatric population. Despite the

advances in microsurgical techniques and intraoperative cord monitoring modalities, there remain a number of serious complications related to the treatment of spinal ependymoma tumors. Identification and acknowledgment of these potential problems may assist in their prevention, early detection, and increased quality of life for patients afflicted with this disease [8].

Outcome

see Intramedullary ependymoma outcome.

There have been anecdotal reports of radiographic response of spinal ependymomas in neurofibromatosis type 2 (NF2) patients being treated for progressive vestibular schwannomas with bevacizumab.

Neurofibromatosis 1 (NF1) and ependymoma

Ependymoma with Neurofibromatosis type 1 (NF1) has rarely been reported. To date, only four cases have been reported in English literature

Case series

see Spinal cord ependymoma case series.

Case reports

Spinal cord ependymoma case reports.

1)
Traveras JM, Ferrucci J. Radiology. Diagnosis-Imaging-Intervention. Neuroradiology and radiology of the head and neck. Lippincott. 1994;3(110):1–12.
2)
Rawlings CE, Giangaspero F, Burger PC, Bullard DE. Ependymomas: a clinicopathologic study. Surg Neurol. 1988;29:271–81.
3)
Choi JY, Chang KH, Yu IK, Kim KH, Kwon BJ, Han MH, Kim IO. Intracranial and spinal ependymomas : review of MR Images in 61 patients. Korean J Radiol. 2002;3:219–28.
4)
Asazuma T, Toyama Y, Suzuki N, Fujimura Y, Hirabayshi K. Ependymomas of the spinal cord and cauda equina : an analysis of 26 cases and a review of the literature. Spinal Cord. 1999;37:753–9.
5)
Schwartz TH, McCormick PC. Intramedullary ependymomas : clinical presentation, surgical treatment strategies and prognosis. J Neurooncol. 2000;47:211–8.
6)
Helseth A, Mørk SJ. Primary intraspinal neoplasms in Norway, 1955–1986, a population-based survey of 467 patients. J Neurosurg. 1989;71:842–5.
7)
Nemoto Y, Inoue Y, Tashiro T, Mochizuki K, Oda J, Kogame S, Katsuyama J, Hakuba A, Onoyama Y. Intramedullary spinal cord tumors: significance of associated hemorrhage at MR imaging. Radiology. 1992 Mar;182(3):793-6. PubMed PMID: 1535896.
8)
Nagasawa DT, Smith ZA, Cremer N, Fong C, Lu DC, Yang I. Complications associated with the treatment for spinal ependymomas. Neurosurg Focus. 2011 Oct;31(4):E13. doi: 10.3171/2011.7.FOCUS11158. Review. PubMed PMID: 21961857.