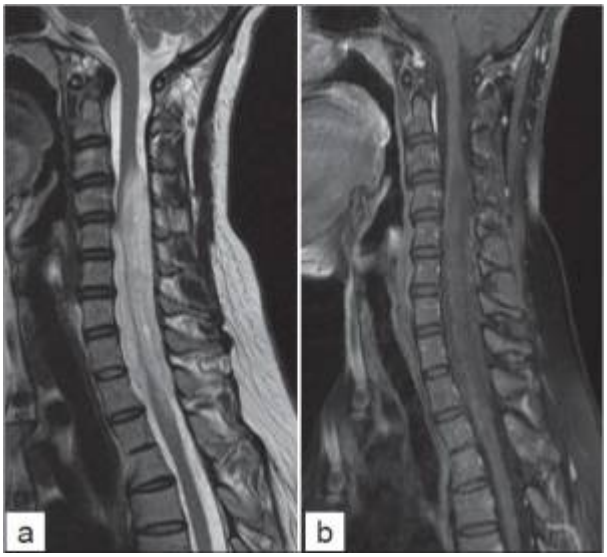
Spinal cord subependymoma



A spinal cord subependymoma (SCSE) is a benign, non-invasive, slow-growing, WHO Grade I spinal cord tumor [1], first reported by Boykin et al. in 1954. [2].

Epidemiology

Their most common site of occurrence is the fourth ventricle followed by the lateral ventricles. Spinal cord subependymomas typically manifest as cervical and cervicothoracic intramedullary or, rarely, extramedullary mass lesions.

Pathology

Histologically, there are hypocellular areas with occasional clusters of cells and frequent microcystic changes, calcifications, and hemorrhage. Radiologically, subependymomas generally manifest as eccentric well circumscribed nodular lesions with mild-to-moderate enhancement.

Clinical features

They often present clinically with pain and neurologic symptoms, including motor, sensory, urinary, and sexual dysfunction.

Diagnosis

Toi et al., made an important discovery of what seems to be a characteristic appearance for spinal subependymoma on sagittal MRI. Swelling of the spinal cord is extremely steep, providing unusually large fusiform dilatation resembling a bamboo leaf. They termed this characteristic MRI appearance

as the “bamboo leaf sign.” This characteristic was apparent in 76.2% of cases of spinal subependymoma for which MRI findings were reported. Conclusion. The bamboo leaf sign on spinal MRI is useful for differentiating between subependymoma and other intramedullary tumors. Neurosurgeons encountering the bamboo leaf sign on spinal MRI should consider the possibility of subependymoma [3].

Differential diagnosis

It is not easily differentiable from a spinal cord ependymoma with radiological findings.

Treatment

Spinal cord subependymomas are not dissected easily from the spinal cord. Considering the rather indolent nature of spinal cord subependymomas, subtotal removal without the risk of neurological deficit is another option [4].

Outcome

Surgical findings and outcomes differ from those of an ependymoma, including a high risk of neurological deficit in the event of a poor dissection plane from the spinal cord with a low rate of recurrence.

Case series

Mikula et al., present a series of spinal cord subependymomas with a detailed description of the clinical, radiological and pathological features, and characterization by chromosomal microarray analysis. Briefly, the four patients included two men and two women, between the ages of 22 and 48 years. The most common presenting symptoms were neck and arm pain with upper extremity weakness. By imaging, the tumors were found to involve multiple spinal levels, including cervical/cervico-thoracic (three patients) and thoracic (one patient), were all eccentric, and had minimal to no post-contrast enhancement. Two patients underwent gross total resection, one had a sub-total resection, and one underwent biopsy alone with a decompressive laminectomy. Follow up ranged from 6 months to 22 years. One patient (case 4) had recurrence 15 years following gross total resection and chromosomal microarray analysis revealed deletions on the long arm of chromosome 6. Our limited series suggests that spinal cord subependymomas can rarely recur, even following gross total resection, suggesting a possible role for long-term surveillance for these rare tumors [5].

Yuh et al., retrospectively reviewed the medical records of ten spinal cord subependymoma patients (M : F=4 : 6; median 38 years; range, 21-77) from four institutions.

The most common symptoms were sensory changes and/or pain in eight patients, followed by motor weakness in six. The median duration of symptoms was 9.5 months. Preoperative radiological

diagnosis was ependymoma in seven and astrocytoma in three. The tumors were located eccentrically in six and were not enhanced in six. Gross total resection of the tumor was achieved in five patients, whereas subtotal or partial resection was inevitable in the other five patients due to a poor dissection plane. Adjuvant radiotherapy was performed in two patients. Neurological deterioration occurred in two patients; transient weakness in one after subtotal resection and permanent weakness after gross total resection in the other. Recurrence or regrowth of the tumor was not observed during the median 31.5 months follow-up period (range, 8-89).

Spinal cord subependymoma should be considered when the tumor is located eccentrically and is not dissected easily from the spinal cord. Considering the rather indolent nature of spinal cord subependymomas, subtotal removal without the risk of neurological deficit is another option [6].

Case reports

A 51-year-old man presented with a 2-year history of progressive muscle weakness in the right lower extremity. Sagittal magnetic resonance imaging (MRI) showed spinal cord expansion at the Th7-12 vertebral level. Surgical resection was performed and the tumor was found to involve predominantly subpial growth. Histological diagnosis was subependymoma, classified as Grade I according to criteria of World Health Organization. They made an important discovery of what seems to be a characteristic appearance for spinal subependymoma on sagittal MRI. Swelling of the spinal cord is extremely steep, providing unusually large fusiform dilatation resembling a bamboo leaf. They termed this characteristic MRI appearance as the “bamboo leaf sign.” This characteristic was apparent in 76.2% of cases of spinal subependymoma for which MRI findings were reported. Conclusion. The bamboo leaf sign on spinal MRI is useful for differentiating between subependymoma and other intramedullary tumors. Neurosurgeons encountering the bamboo leaf sign on spinal MRI should consider the possibility of subependymoma [7].

A case report of a single patient in whom a subependymoma was resected from the cervical spinal cord with return to normal functioning.

Clinical examination, magnetic resonance imaging evaluation, surgical resection, and histological analysis were performed for diagnosis and treatment of this patient.

The patient experiencing myelopathy symptoms underwent a surgical resection of cervical spinal cord subependymoma that resulted in return to normal function.

Subependymoma should be included in the differential diagnosis of atypical presentations for myelopathy, as discrete surgical resection can result in good outcome [8].

A 53 year old man with a progressive paraparesis, paraesthesias of the lower limbs and sphincter disturbance. The tumour was partly removed, without progression 5 years after surgery [9].

References

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