Spinal cord lymphoma

Lymphoma of the spinal cord is an uncommon manifestation of lymphoma. Although spinal lymphoma more commonly involves the vertebral body (vertebral body tumours) or epidural compartment, intramedullary lymphoma may rarely occur.

Apparent intramedullary spinal cord lymphoma may often in fact represent secondary invasion of the spinal cord by leptomeningeal carcinomatosis.

Epidemiology

Intramedullary spinal lymphoma accounts for 3.3% of all CNS lymphoma, which constitutes only 1% of all lymphomas in the body.

The mean age at presentation is 47 years. Females are more commonly affected than males.

see also Cauda equina lymphoma.

Types

Primary intramedullary lymphoma

Secondary intramedullary lymphoma

Risk factors

Risk factors for developing CNS lymphoma include:

AIDS

Transplant recipients

Congenital immune deficiency

Epstein-Barr virus infection

Clinical presentation

Clinical presentation is similar to that of other intramedullary spinal tumours, with pain, weakness and sensory changes common.

Pathology

Primary lymphoma of the spinal cord is most often predominantly histiocytic or mixed histiocytic and

lymphocytic. 85% are non-Hodgkin lymphomas.

The principal microscopic feature is a dense, perivascular, mononuclear infiltrate composed predominantly of large lymphocytes with prominent nucleoli. Most tumours are B-cell derivatives.

Radiographic features

The most common intramedullary location is the cervical cord, followed by the thoracic cord then the lumbar cord.

Most are solitary lesions, however there may be multiple lesions throughout the spinal cord.

MRI

Although spinal cord expansion is usually present, in some patients there is relatively minimal enlargement of the cord. The lesions are generally poorly defined and tumoural cysts are generally not a feature and secondary syringomyelia is rare . Lymphoma usually does not have a haemorrhagic component.

Reported signal characteristics include:

T1: isointense to spinal cord

T2: hyperintense (this is in contrast to the characteristic low T2 signal intensity that is seen in intracranial lesions)

T1 C+ (Gd): usually solid and homogeneous enhancement

Treatment and prognosis

Radiotherapy is the primary therapy for potential preservation of neurologic function and extension of survival. Minimal data are available on the role of chemotherapy for primary spinal cord lymphoma.

The prognosis for patients with intramedullary spinal lymphoma is poor. The survival rate at 2.5 years is less than 50% 3.

Differential diagnosis

General imaging differential considerations include:

spinal astrocytoma

tumoural cysts are more common

spinal ependymoma

tumoural cysts are more common

heamorrhage is more common

spinal intramedullary metastases

usually more sharply circumscribed

transverse myelitis

variale contrast enhancement

rapidly progressive clinical course

spinal cord contusion

an appropriate history is usually present spondylotic myelopathy lacks enhancement multiple sclerosis with spinal involvement most have brain lesions typically located eccentrically enlargement of the cord is uncommon may not demonstrate contrast enhancement.

Case series

Mayo Clinic medical records, lymphoma database, and autopsies from 1996 to 2009 were searched. Inclusion criteria were clinical myelopathic presentation, intramedullary spinal cord abnormalities, and pathologically confirmed CNS lymphoma. Exclusion criteria were extramedullary lymphoma, secondary intramedullary lymphoma, or other myelopathic etiology. Clinical features, diagnostic methods, neuroimaging, treatment, and outcomes were assessed.

The 14 patients' median age at presentation was 62.5 years (range 41-82 years) and 10 were men (71%). Two had lymphoma risk factors (HIV infection 1; chronic immunosuppression postorgan transplant 1). Most had initial presumptive diagnoses of CNS demyelinating disease and definitive diagnosis of lymphoma was delayed a median of 8 months (range 1-22 months). CNS lymphoma was pathologically confirmed by biopsy (brain 6; spinal cord 4), CSF cytology (3), and autopsy (1). Most patients had multifocal, persistently enhancing lesions on spinal MRI and 8 had involvement of conus medullaris, cauda equina, or both. IV methotrexate was the initial treatment in 9 of 12 (75%) but lymphoma recurred in the majority. Half of the patients were wheelchair-dependent at 10 months and 2-year survival was 36%.

PISCL mimics other causes of myelopathy. Spinal MRI demonstrating multifocal lesions, persistent gadolinium enhancement, and conus medullaris or cauda equina involvement is characteristic. Pathologic confirmation often requires CNS biopsy. Despite chemotherapy, morbidity and mortality is high [1)].

Case reports

Primary spinal lymphoma carries a low incidence and poor prognosis. The most reliable diagnostic signs have been segmental swelling of the cord on myelography and increased cellularity of cerebrospinal fluid. Biopsies of spinal tumors or their cerebral extensions showed lymphoma of the non-Hodgkin type only. Peroxidase-antiperoxidase staining for light chains helps to distinguish well differentiated lymphomas from infectious processes in some instances. A lymphoma in an immunosuppressed patient fulfilling all criteria for primary growth in the spinal cord is described, and the case compared with others in the literature. Combined radiotherapy and chemotherapy provided

a 48-month survival in one patient [2].

1)
Flanagan EP, O'Neill BP, Porter AB, Lanzino G, Haberman TM, Keegan BM. Primary intramedullary spinal cord lymphoma. Neurology. 2011 Aug 23;77(8):784-91. doi: 10.1212/WNL.0b013e31822b00b9. Epub 2011 Aug 10. PubMed PMID: 21832220.
2)
Hautzer NW, Aiyesimoju A, Robitaille Y. “Primary” spinal intramedullary lymphomas: a review. Ann Neurol. 1983 Jul;14(1):62-6. PubMed PMID: 6614871.