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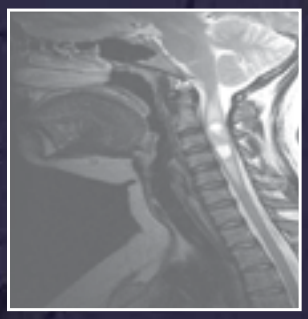
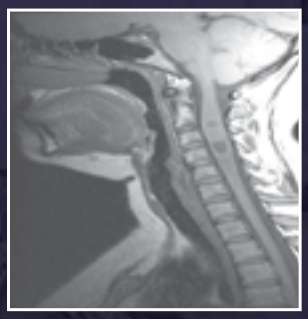
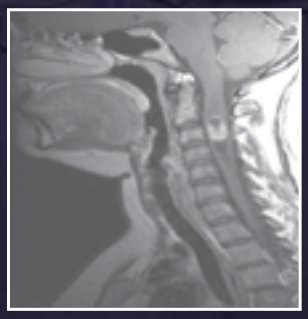
These three MRI scans show sagittal images of a patient with an intramedullary ependymoma of the cervical spinal cord.



The first is a T2 weighted sagittal image of the cervical spine showing the tumor surrounded by a typical syrinx (white fluid). Above and below the syrinx is edema in the spinal cord.



The second is a T1 weighted image.



The third is a T1 after gadolinium enhancement revealing the tumor turning white.

Treatment Strategies for Intramedullary Tumors

by Thomas H. Jones, MD, Neurosurgeon and Medical Director, Santa Barbara Neuroscience Institute

Intramedullary tumors (IMT) of the spinal cord are rare, representing only 2 to 4 percent of all CNS intrinsic tumors. However, in the pediatric cohort, they account for 10 percent of CNS neoplasms.

ASTROCYTOMAS, EPENDYMOMAS AND hemangioblastomas, appearing largely benign histologically, represent the majority of intramedullary neoplasms. These lesions are typically slow-growing and expansile, and patients generally present with back pain and/or sensorimotor dysfunction. Astrocytomas represent 90 percent of the IMT in children under age 10 and 60 percent in adolescents. Ependymomas predominate in midlife and after age 60, are roughly statistically equivalent to astrocytomas.

The differential diagnosis of intramedullary mass lesions includes subependymomas, metastases from extraneural cancers, demyelinating disease, vascular malformations, parasitic cysts, sarcoidosis and subacute myelomalacia related to extrinsic compression from spondylosis and/or disc herniations.

DIAGNOSTIC EVALUATION

A thorough history is essential. The physician must look for the clinical signatures of other disease processes. For instance, demyelinating disease is frequently suggested by a history of recurring neurologic events affecting disparate anatomic areas of the CNS. Such historical clues should prompt a diagnostic brain MRI or CSF analysis. Pulmonary symptoms might trigger a more thorough workup looking for evidence of sarcoidosis. A rapidly deteriorating clinical course, particularly in a patient with a history of cancer (esp., small cell lung cancer or melanoma), would raise the specter of an intramedullary metastases.

MRI is the diagnostic tool of choice and, renal function permitting, should be ordered with and without the IV administration of gadolinium. Ependymomas tend to be slightly hyperintense on T2 images, often have clear tumor margins, uniformly enhance with gadolinium and the majority are associated with syringomyelia. Astrocytomas are less likely to enhance or have clear tumor margins and are slightly less likely to have an associated syrinx. Hemangioblastomas are generally smaller, circumscribed, highly vascular lesions which are pia-based, dorsally located and are also commonly associated with large syrinxes.

Remember the increased likelihood of IMT in patients with neurofibromatosis (NF1 or NF2), one of the most common autosomal dominant heritable diseases. Additionally, von Hippel-Lindau syndrome is an autosomal dominant neurocutaneous disease strongly linked to multifocal CNS hemangioblastomas as well as other systemic tumors, including those in the pancreas, kidney and pheochromocytomas.

CLINICAL MANAGEMENT

In symptomatic patients, IMT consistent with ependymoma or astrocytoma should undergo surgical exploration and attempted resection. Typically, most ependymomas can be grossly excised through a midline myelotomy and long-term cure anticipated. In patients with astrocytomas, the surgical treatment hinges on the development of a plane of dissection. In most series, a total gross resection is possible in only 50 to 70 percent. Hemangioblastomas can be resected in most patients, although those with von Hippel-Lindau syndrome, who tend to develop multiple lesions, should be operated upon only if worsening while being followed. A standard surgical approach includes use of microsurgical techniques supplemented by sensory and motor-evoked potentials. Radiation therapy is reserved for incompletely resected astrocytomas, in patients considered too ill for surgery or those who have lost most neurologic function by the time of clinical presentation.

For more information, please e-mail sbni@sbch.org or visit our website at www.sbni.org.