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Spinal Ependymoma and its Radiological Findings

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Abstract

Spinal Ependymomas are the most common intramedullary neoplasm in adults, comprising 60% of all glial spinal cord tumors. They are the second most common intramedullary neoplasm in the pediatric population, accounting for 30% of pediatric intramedullary spinal neoplasms. Peak incidence is in the fourth decade, with 39 years being the median age at presentation. Males are more affected than females. There is an increased coincidence with neurofibromatosis type II.

Keywords: Ependymoma • Spinal Cord • Neurofibromatosis

Introduction

There are no clear risk factors for ependymomas in adults, except, as in children, that patients with neurofibromatosis type II have an increased risk of developing ependymomas, particularly spinal ependymomas. NF2related ependymomas have a slower growth rate and more benign behavior and are therefore often treated less aggressively than sporadic ependymomas [1,2].

The clinical presentation is similar to that of other intramedullary spinal tumors, with common pain, weakness, and sensory changes. The present study was qualitative in nature, with the elaboration of a literature review, using as means of theoretical foundation the academic and scientific journals available

Case Report

Spinal ependymomas are the most common spinal cord tumor in general, seen in both adult and pediatric populations. Peak incidence is in the fourth decade, with 39 years being the median age at presentation. Males are more affected than females. There are no clear risk factors for ependymomas in adults, with the exception, as in children, of patients with type II neurofibromatosis. The present study was of qualitative nature with elaboration of literature review, using as means of theoretical foundation the academic and scientific journals available, comparing the different data found and relating them to the radiological findings in patients with spinal ependymoma. From the results obtained through the different data found, it is evident that ependymomas may occur anywhere along the spinal cord; however, the cervical spinal cord is the most common site (44%). Simple film features that can be seen with a spinal ependymoma include several. Most ependymomas are slow growing. They tend to compress the adjacent spinal cord tissue rather than infiltrate it, almost always leaving a cleavage plane between the tumor and the spinal cord tissue [3].

Results and Discussion

From the results obtained through the different data found, it is evident that ependymomas can occur anywhere along the spinal cord; however, the cervical cord is the most common location (44%). An additional 23% occur within the cervical cord and extend to the upper thoracic cord, and 26% occur only in the thoracic cord. Sensory symptoms can be explained by the proximity of these centrally located tumors to the spinoalamic tracts. Dominant motor symptoms are commonly associated with very large ependymomas.

Ependymomas were believed to arise from ependymal cells lining the central canal of the spinal cord, but more recently it has become evident that they actually arise from progenitor cells or radial glia-like stem cells. On histological evaluation, uniformly hyperchromatic nuclei are usually observed. Perivascular pseudo-rosettes are the classic finding [1].

In a small subset of patients, MYCN amplification is identified. This is a distinct tumor and is now recognized as a separate entity in the 5th Edition (2021) of the WHO classification of CNS tumors. The vast majority of spinal ependymomas are WHO grade 2, with grade 3 tumors being rare. Spinal ependymomas with MYCN amplification are aggressive high-grade tumors; however, they have not yet received a formal grade in the 5th Edition (2021) of the WHO classification of CNS tumors. Online and in printed versions, gathering and comparing the different data found in the sources that were consulted and listing the main factors related to the radiological findings in patients with spinal ependymoma, as well as the characteristic signs and symptoms that were observed in the patients' radiographic images. Simple film features that can be seen with a spinal ependymoma include: scoliosis, spinal canal widening, scaly vertebral body, pedicle erosion, laminar thinning [4].

CT can demonstrate non-specific canal widening, this to slightly hyperattenuating compared to normal spinal cord, intense enhancement with iodinated contrast, large lesions can cause flaking of the posterior vertebral bodies and enlargement of the neural outflow foraminal. MRI is the modality of choice for evaluating suspicious spinal cord tumors [5].

enlarged 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 expansion of the cord); although not encapsulated, they are well circumscribed; tumor cysts are present in 22%; non-tumor cysts are present in 62%; syringomyelia occur in 9-50% of cases; in contrast to intracranial ependymomas, calcification is uncommon; average length of four vertebral body segments (Figure 1).

Typical signal characteristics on, T1: most are isointense to hypointense; mixed signal lesions are seen if cyst formation, tumor necrosis, or hemorrhage has occurred. T2: hyperintense - peritumoral edema is seen in 60% of cases; associated hemorrhage leads to the "cap sign" (a hypointense hemosiderin rim on T2-weighted images) in 20-33% of cases. The cap sign is suggestive, but not pathognomonic, for ependymoma, as it can also be seen in hemangioblastomas and paragangliomas. T1 C+ (Gd): virtually all strongly enhancing, somewhat inhomogeneous (Figure 2) [6,7].

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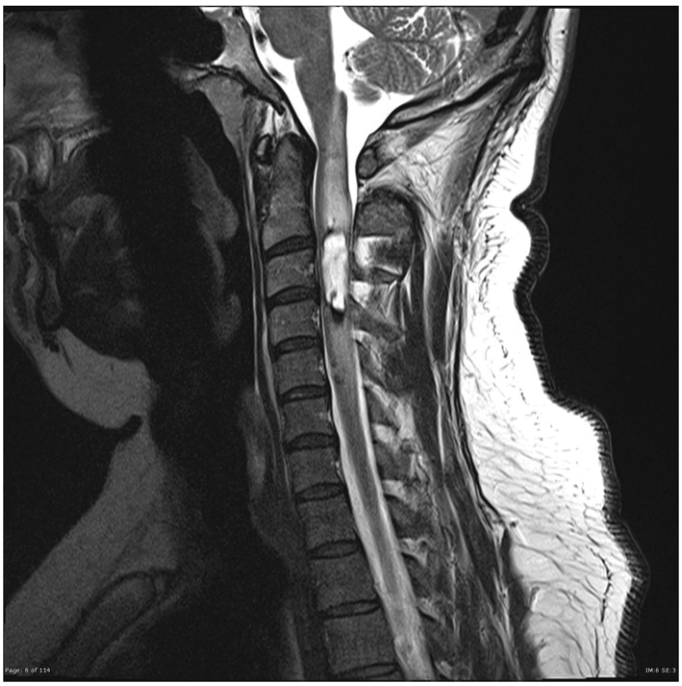

Figure 1. MRI image extending from C2/3 to the midpoint of C4 is an intramedullary mass that is high T2 signal, low T1 signal and demonstrates prominent contrast enhancement (visible on axial images). It has a prominent hemosiderin boundary with some intrinsic high T1 signal consistent with hemorrhage. A moderate amount of vasogenic edema extends above and below the lesion from the cervical crater junction to C7. The remainder of the cervical cord, cervical vertebrae, and paraspinal soft tissues are unremarkable. The vertebral arteries are codominant to demonstrate normal flow voids.

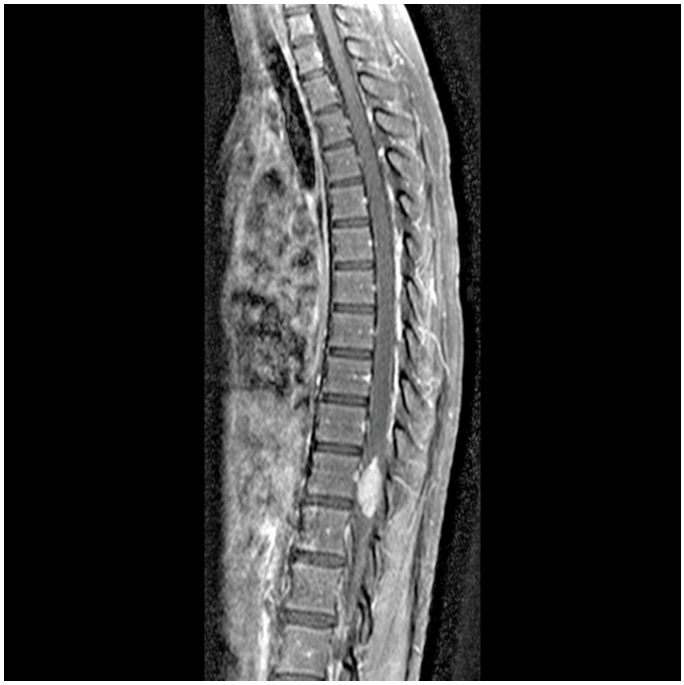

Figure 2. Image from an MRI, showing a welldemarcated central intramedullary tumor at T11T12, which is isointense on T1, heterogeneously hyperintense on T2 and has intense contrast enhancement. There is a small polar cyst component at the inferior margin of the tumor. This mass promotes local expansion of the cord. The lesion measures 1.2 x 1.2 x 2.8 cm. There is a hyperintense T2 signal in the spinal cord segment above the tumor, which may represent edema and some component of syringomyelia.

Conclusion

Most ependymomas are slow growing. They tend to compress the adjacent spinal cord tissue rather than infiltrate it, almost always leaving a cleavage plane between the tumor and the spinal cord tissue. A complete curative excision can be achieved in approximately 50% of cases. In these patients, the 5-year survival rate is approximately 85%. In patients who cannot achieve complete resection, the 5-year survival rate is approximately 57%. Recurrence is rare after complete excision. Although metastatic spread is rare, the most common sites for metastasis include the retroperitoneum, lymph nodes, and lungs. Spinal ependymomas with MYCN amplification are, in contrast, aggressive tumors with poor prognosis and frequent early dissemination throughout the central nervous system and recurrence despite resection.

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