

Gangliogliomas: Issues of Prognosis and Treatment

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Gangliogliomas are uncommon tumors found in approximately 1% of primary CNS neoplasms. These lesions predominantly occur in children and young adults with the peak incidence in the second decade of life. The tumors consist of atypical, dysplastic neurons and glial cells, which are usually astrocytes, that show neoplastic features. They can be found in all sites, but most often are seen in the temporal lobe. The tumors are generally well circumscribed and slow growing. Although they have an overall good prognosis, attempts to predict the clinical outcome on the basis of histologic features have generally been unsuccessful. Malignant transformation may occur, usually within the glial component of the neoplasm.

Patel et al (see page 808) emphasize that these neoplasms may not be as rare as heretofore believed, particularly in the spinal cord. They reported an incidence of approximately 25% in their series of 11 cases, although the absence of juvenile pilocystic astrocytomas was surprising. Problems with arriving at the proper diagnosis are discussed. As the tumor may have a heterogeneous appearance, the small tissue samples that are often available for examination may not include the diagnostic ganglion cells. Additionally, ganglionic elements may be easily mistaken for gemistocytic astrocytes. The use of synaptophysin immunohistochemistry by these investigators has disclosed that many tumors previously diagnosed as astrocytomas are in fact gangliogliomas.

A word of caution, however, may be in order regarding the use of synaptophysin immunohistochemistry for the diagnosis of gangliogliomas. While its utility has been of major value in the diagnosis of ganglioglioma (1), Zhang and Rosenbloom (2) maintain that normal neurons in the spinal cord may stain for this protein in a manner similar to that of neo-

plastic ganglion cells. While Patel et al reasonably address these concerns, appropriate caution is still necessary.

Overall, this important paper highlights the frequency of ganglioglioma and provides useful guidelines for the diagnosis of this tumor. It certainly indicates that gangliogliomas need to be seriously considered in the differential diagnosis of all intramedullary neoplasms of the spinal cord. As the prognosis of gangliogliomas is different from that of other commonly occurring neoplasms of the spinal cord, its proper diagnosis is of utmost importance.

The paper by Kincaid et al (see page 801) points out useful radiologic criteria for the diagnosis of ganglioglioma. Such background information will be useful to the pathologist in deciding to carry out a more diligent search for the presence of a ganglioglioma. In their series they found a positive correlation between histologic malignancy and poor clinical outcome. Such correlations have not been uniformly found in several series, including the one by Patel et al. This issue remains controversial.

In summary, both papers highlight clinical, pathologic and radiologic features of an unusual, bicellular neoplasm that occurs more often than previously recognized. Future studies should settle issues of prognosis and appropriate treatment for this interesting neoplasm.

References

1. Miller DC, Koslow M, Budzilovich GN, Burstein DE. Synaptophysin. A sensitive and specific marker for ganglion cells in central nervous system neoplasms. *Hum Pathol* 1990;21:93-98
2. Zhang PJ, Rosenbloom MK. Synaptophysin expression in the human spinal cord—diagnostic implications of an immunohistochemical study. *Am J Surg Pathol* 1996;20:273-276

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