Prevalence of Medulloblastoma in Shiraz, Southern Iran

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Dear Editor

Brain tumors are the second most common malignancy among children less than 20 years of age.1 Medulloblastoma (MB), as a specific brain tumor, is mostly prevalent in children between the ages of 3-8 years.2,3 It is a highly malignant primary brain tumor that develops in the cerebellum, in the posterior fossa, but may spread to other parts of the brain and to the spinal cord through the cerebrospinal fluid.2,3 The tumor's origin in the cerebellum is infratentorial as they occur below the tentorium.4 Medulloblastoma is diagnosed histologically; its histological classification has gained in importance and newer treatment protocols will include histology stratification.1 Correct diagnosis of MB may require ruling out atypical teratoid rhabdoid tumor (ATRT)5 and primitive neuroectodermal tumor (PNET). It is rare in adults (2%)6 and more common in males (62%) than females (38%).7 The tumor is distinctive on T1 and T2-weighted MRI and is well circumscribed, solid, pink-gray in color and very cellular.5 Treatment is dependent on staging and age of onset while these findings cannot differentiate which standard-risk patients will relapse and die. Outcome has been shown to be affected by tumor subtype and molecular alterations.8 Dose-intense chemotherapy can optimize treatment, especially in the context of risk stratification.9 This study aims to determine the epidemiology MB in Shiraz, Southern Iran.

During the last 4 years, we enrolled 41 cases of 5-18 year-old children who referred to the neurosurgery wards affiliated with Shiraz University of Medical Sciences. Of these, 20 patients were histologically diagnosed with MB or PNET and 90% had hydrocephaly. The majority (95%) of patients suffered from classic PNET and 5%
from the desmoplastic type. For 80%, the tumors were located in the midline posterior fossa. The fourth ventricle was involved in 35% of patients, and 10% had evidence of spinal metastasis. The male to female ratio was 3:2. Total resection was attempted for 70% of subjects. The survival rate at one year was 90%; at two years, 80%; and at three years, 20%. All patients underwent radiotherapy.

Chan et al. reported that 41% of patients had spinal metastases. Total resection was performed in 65% of patients. The prospect of survival at one year was 85%; at two years, 70%; and at three years, 15%. McNeil et al. showed a survival of 86% at one year and 56% at five years. These findings were identical to our results. All patients underwent radiotherapy except two who underwent both radiotherapy and chemotherapy. Our findings have shown that MB in our area is similar to that observed in other areas worldwide.

References