Intraventricular Subependymoma

Subependymomas are benign tumors of the ventricles that grow from the ventricular wall into the cerebrospinal fluid spaces within the brain, obstructing the flow of the cerebrospinal fluid and causing obstructive hydrocephalus. It is estimated that ependymomas represent between 0.2% and 0.7% of all intracranial tumors. They arise most frequently in the fourth ventricle (50-60%) and the lateral ventricles (30-40%) [1].

Epidemiology

They occur in middle to late adulthood.

Representing approximately 10% of ependymal tumors, subependymomas most often “present” as incidental autopsy findings in the brains of the elderly.

Most frequently they arise in the fourth ventricle (50-60%), followed by the lateral ventricle (30-40%), and less frequently in the septum pellucidum and spinal cord [2] [3].

Classification

Third Ventricular Subependymoma

Subependymoma in the lateral ventricle.

Subependymoma of the fourth ventricle.

Subependymoma of the septum pellucidum.

Histology

Subependymomas are small, discrete tumors of adults lying most often at the foramen of Monro or the fourth ventricle. It is composed of clusters of ependymal and astrocyte-like cells in a dense fibrillary stroma. It is typically attached to a ventricular wall and the most common site is the fourth ventricle.

Histologically, the tumor may be either compact or microcystic, but extensive microcystic change is rarely reported.

The histogenesis of subependymomas is still a matter of debate, with candidates including subependymal glia, astrocytes, ependymal cells, or some mixture of these cells. A recent theory hypothesizes that they originate from tanycytes, which are cells normally located in the subependymal zone [4].

Clinical features

They are likely to remain asymptomatic throughout life and some were found by autopsy. If symptomatic, tumor location and size are critical factors for presentation.

Diagnosis

Subependymomas have typical image morphologic characteristics that differentiate them from tumors of other entities, however, the rare subgroup of histopathological mixtures of subependymomas with ependymal cell fractions has no distinctly different imaging properties.

Knowing the imaging characteristics of subpendymoma and their differential diagnoses is of particular importance in order to be able to decide between the necessity of follow-up controls, an early invasive diagnosis or, depending on the entity, tumor resection.

KEY POINTS:

· Subependymomas have typical imaging characteristics that are clearly distinguishable from other entities.. · Increased incidence in middle/ older aged men, most frequent localization: 4th ventricle..

· Symptomatic subependymomas, often located in lateral ventricles, are usually characterized by hydrocephalus..

· Radiological identification of mixed subependymoma with ependymal cell fractions is not possible..

· Image based differentiation from other entities is important for the procedure.. [5)].

Treatment

The surgical aims are the maximal safe tumoral resection, the decompression of neural elements, and establishment of a pathological diagnosis and the restoration of normal CSF pathways. As subependymomas are low-grade lesions with low rates of cell proliferation and a benign clinical course, complete surgical removal is usually curative.

Case series

33 patients with subependymoma, including 4 patients with a mixture of subependymomas with ependymal cell fractions in terms of imaging and clinical aspects and with reference to a current literature review.

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With the SEER-18 registry database, information from all patients with intracranial subependymoma diagnosed during 2004-2013 were extracted, including age, sex, race, occurrence of surgery, extent of primary surgery, receipt of radiation, tumor size, and follow-up data. Age-adjusted incidence rates, overall survival, and cause-specific survival were calculated. Cox proportional hazards model was used for both univariate and multivariate analyses.

Four hundred sixty-six cases were identified. The overall incidence of intracranial subependymoma is 0.055 per 100,000 person-years (95% confidence interval, 0.05-0.06). Through multivariate analysis, age <40 years (hazard ratio [HR], 0.21; P = 0.03), female sex (HR, 0.34; P = 0.03), location within ventricles or near brainstem (HR, 0.49; P = 0.04), and occurrence of surgery (HR, 0.50; P = 0.02) were significant independent positive prognostic factors. Receipt of radiation did not show a significant relationship.

Clinical factors such as younger age, female sex, and location within ventricles or near brain stem demonstrated positive relationship with overall survival. For treatment options, surgery remains a mainstay option. No support for radiation therapy was identified [7].

Forty-three cases of pathologically confirmed, surgically treated intracranial subependymoma were identified. Thus in this patient population, subependymomas accounted for approximately 0.07% of intracranial tumors (43 of an estimated 60,000). Radiologically, 79.1% (34/43) of intracranial subependymomas were misdiagnosed as other diseases. Pathologically, 34 were confirmed as pure subependymomas, 8 were mixed with ependymoma, and 1 was mixed with astrocytoma. Thirty-five patients were followed up for 3.0 to 120 months after surgery. Three of these patients experienced tumor recurrence, and one died of tumor recurrence. Univariate analysis revealed that shorter progression-free survival (PFS) was significantly associated with poorly defined borders. The association between shorter PFS and age < 14 years was almost significant (p = 0.51), and this variable was also included in the multivariate analysis. However, multivariate analysis showed showed only poorly defined borders to be an independent prognostic factor for shorter PFS (RR 18.655, 95% CI 1.141-304.884, p = 0.040). In patients 14 years of age or older, the lesions tended to be pure subependymomas located in the unilateral supratentorial area, total removal tended to be easier, and PFS tended to be longer. In comparison, in younger patients subependymomas tended to be mixed tumors involving the bilateral infratentorial area, with a lower total removal rate and shorter PFS.

Intracranial subependymoma is a rare benign intracranial tumor with definite radiological features. Long-term survival can be expected, although poorly defined borders are an independent predictor of shorter PFS. All the features that differ between tumors in younger and older patients suggest that they might have different origins, biological behaviors, and prognoses [8].

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24 pathologically proved cases of intracranial subependymomas in 17 male and seven female patients with a mean age of 48.1 years. All patients were symptomatic. CT and MR images were used to characterize the size, shape, and location of the subependymomas; the degree of hydrocephalus; tumor calcification; and the density, signal, and enhancement characteristics of the tumors.

Eighteen of 24 tumors were 3 cm or more in greatest dimension. Nineteen were lobulated, and hydrocephalus was seen in 21. Fourteen were in the lateral ventricle, and 10 were in the posterior fossa. Calcifications were present in five (all fourth ventricular) and absent in 10 (all lateral ventricular) subependymomas imaged with unenhanced CT. On 18 contrast-enhanced CT scans, five of six subependymomas with heterogeneous enhancement were in the fourth ventricle, and nine of 12 tumors with minimal or no enhancement were in the lateral ventricle. Small internal foci with a signal intensity similar to that of CSF were seen on images of all 10 lateral ventricular subependymomas obtained with both T1-weighted and T2-weighted sequences. On 13 contrast-enhanced T1-weighted images, seven of eight tumors with heterogeneous enhancement were in the fourth ventricle, and all five with minimal or no enhancement were in the lateral ventricle.

Intracranial subependymomas were seen in symptomatic middle-aged adults and showed different CT and MR imaging features, depending on their anatomic location. Calcification and heterogeneous contrast enhancement were common features of fourth ventricular subependymomas showed a lack of calcification as well as minimal or no contrast enhancement of CT and MR images [9].

Case reports

Intraventricular subependymoma presenting as subarachnoid hemorrhage [10].

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