Central neurocytoma

Central neurocytoma (CN), is a intraventricular tumor of neuroepithelial origin with intermediate malignancy (WHO grade II) first described in 1982 by Hassoun and colleagues [1].

Typically forms from the neuronal cells the septum pellucidum.

Epidemiology

Central neurocytomas are typically seen in young patients (20-40 years of age) and account for less than 1% (0.25-0.5%) of intracranial tumors. There is no reported gender predilection.

Central neurocytomas (CNs) occurs typically in the lateral ventricle, adjacent to the septum pellucidum and foramen of Monro. [2].

Isolated lesions in the third ventricle are uncommon [3].

Classification

Atypical central neurocytoma with extracranial metastases is a rare variant of benign central neurocytoma (CN).

Molecular Genetics

1p/19q co-deletion has not been reported in central neurocytoma, but it can be seen in extraventricular neurocytoma.

Clinical features

The clinical presentation varies and many are incidentally detected.

Typically central neurocytomas present with symptoms of intracranial hypertension, headaches being most frequent, or seizures (especially tumours with extra ventricular extension).

A relatively short clinical course, typically only a few months, is most common. Rarely central neurocytomas may be associated with sudden death secondary to acute ventricular obstruction. Also rare, is a sudden presentation due to intraventricular hemorrhage.

see Obstructive hydrocephalus from central neurocytoma.

Only 3 previous cases in which CN presented with co-occurring psychotic symptoms were found in the PubMed database. A report presents the case of a 27-year-old patient with paranoid syndrome without neurological symptoms, in whom magnetic resonance imaging confirmed a large intracranial

tumor located predominantly in the right lateral ventricle and third ventricle reaching down to the hypothalamus. Resection of the tumor (histopathologically a CN) resulted in complete remission of the psychotic symptoms. This case supports the need for neuroimaging in all patients with first-episode psychosis because of the possibility of neurologically silent brain tumors. Quick diagnosis in such cases is crucial for the selection of treatment methods and prognosis [4].

Complications

The incidence of spontaneous hemorrhage at presentation is rare. Chandshah et al. reported a case of the intraventricular lesion with bleed which was operated on an emergency basis as it caused obstructive hydrocephalus and the patient was in altered sensorium. The exact cause of hemorrhage in CNs is not known, and the previously discussed explanations are discussed [5].

see Obstructive hydrocephalus from central neurocytoma.

Diagnosis

Detectable with both computed tomography and magnetic resonance imaging.

Fairly characteristic imaging features, appearing as heterogeneous masses of variable size and enhancement within the lateral ventricle.

The vast majority of central neurocytomas are located entirely within the ventricles. Typical locations include:

lateral ventricles around foramen of Monro (most common): 50%

both lateral and 3rd ventricles: 15%

bilateral: 15%

3rd ventricle in isolation: 5%

Extra ventricular neurocytomas (or cerebral neurocytomas) are distinctly uncommon, and thought to be a separate entity due to the tendency to have prominent ganglionic or glial differentiation.

CT

Central neurocytomas are usually hyperattenuating compared to white matter. Calcification seen in over half of cases, usually punctate in nature.

Cystic regions are frequently present, especially in larger tumours. Contrast enhancement is usually mild to moderate. Accompanying ventricular dilatation often present.

MRI

T1 isointense to grey matter heterogenous

T1 C+ mild-moderate heterogeneous enhancement

T2/FLAIR typically iso to somewhat hyperintense compared to brain numerous cystic areas (bubbly appearance), many of which completely attenuate on FLAIR prominent flow voids may be seen

GE/SWI calcification is common, typically punctate haemorrhage (especially in larger tumours) is common uncommonly results in ventricular haemorrhage

MR spectroscopy may have a strong choline peak glycine peak (3.55ppm) has also been reported

Angiography

A tumor blush is frequently identified, with the mass supplied by choroidal vessels. No large feeding arteries are usually seen.

Pathology

Central Neurocytoma Pathology.

Differential diagnosis

Central Neurocytoma Differential Diagnosis.

Treatment

see Central neurocytoma treatment.

Outcome

They generally have a good prognosis provided a complete resection can be achieved.

Cases of CSF dissemination have been reported, but are rare.

Case series

see Central neurocytoma case series.

Case reports

Central neurocytoma case reports.

References

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