Radiological Features and Differential Diagnosis of Lateral Ventricle Central Neurocytoma

Central Neurocytoma (CN) accounts for less than 1% of total brain tumors. It is typically localized in supratentorial ventricular region in relation to septum pellucidum and foramen monro. It is of pure neuronal origin predominantly affecting young individuals between 20-40 years of age. Aim of this study is to evaluate neuroradiological feature of CN in Magnetic Resonance Imaging (MRI) with pathological correlation.

We retrospectively reviewed five cases of CN diagnosed in last ten years and tried to study their locations and radiological variations in MRI.

The age distribution was 20-54 years, the mean age being 37 years. All five lesions were intra ventricular in origin; biventricular in three, right ventricular in one and left ventricular in one. Two cases showed extension to the occipital horn of the right lateral ventricle. In one case intra ventricular lesion was completely cystic. In all the five cases, cystic areas and obstructive hydrocephalus were detected and hemorrhage was seen in only two cases.

Though relatively rare, CN is not that uncommon. In case of intraventricular lesions, if it is in relation to septum pellucidum causing obstructive hydrocephalus, containing cystic areas and occasional hemorrhage in MRI with mild to moderate contrast enhancement, CN should always be suspected.

Key words: central neurocytoma, intraventricular lesions, MRI, radiological features

Central Neurocytoma (CN) is one of the relatively rare tumors of the central nervous system (CNS). It is grade II tumor according to WHO classification. It is a biologically benign, well differentiated tumor of purely neuronal origin and has better outcome after complete surgical resection. It was first described by Hassoun et al in 1982 with two cases of intra ventricular tumors of neuronal origin and benign in nature. Since then more than 500 cases have been identified and reported. Most of the neurosurgeons and neuropahtologists have published their cases of CN as a case report or a short series.

CN is found to be originated at the midline area in the supratentorial region straddling in lateral and third ventricles. It is usually attached to the septum pellucidum, fornix or walls of the lateral ventricles producing mass effect on foramen monro causing obstruction and subsequent hydrocephalus. Cystic lesions are frequently seen and 50% of the lesions contain calcification. Hemorrhagic presentation is less common. It has a propensity to occur mostly in young age group between 20-40 years with equal sex predilection. Radiological and light microscopy appearance of CN resembles to oligodendroglioma and ependymoma causing misdiagnosis.

Materials and methods
This is a simple retrospective analytical study of small number of cases. Five histopathologically diagnosed cases
of CN from 2000-2010 were retrospectively studied. They were evaluated radiologically and immunohistochemically. Various aspects of CN were analyzed and evaluated. Among them were two males and three were females ranging from 20 to 54 years of age (mean age=37). All the patients were studied by MRI. Patients presented with headache (N=5), dizziness (N=4), seizure (N=1), blurred vision (N=1) and hemiparesis (N=1) as the main clinical manifestations.

All the radiological investigations were performed by GE signa excite 3.0 Telsa MRI (general electronic, USA). Fast Spin echo sequences with T1Weighted image (T1WI) (TR 500, TE 15), T2Weighted image (T2WI) (TR 5100, TE 137), FLAIR (TR 9602, TE 116) and Simens Magnetom Vision Plus 1.5 Telsa (Simens co., Germany) Spin echo sequences with T1WI (TR 450, TE 14), T2WI (TR 5000, TE 128), FLAIR (TR 9000, TE 120). Contrast agent used for accurate imaging was Gadolinium diethyle triamine penta acetic acid (Gd-DTPA) in the dose of 0.2 ml per kg body weight.

### Results

Among 5 cases, 2 were males and 3 were females. Age distribution was between 20-54 years old (Mean=37). Of 5 cases 3 were biventricular lesions, 1 was in the left ventricle and 1 in right lateral ventricles invading left lateral ventricle. All 5 tumors were attached to the septum pellucidum (Figure 1). Two of them showing extension into the occipital horn of the lateral ventricle (Figure 2) and three were extending towards the anterior horn of the lateral ventricle. Hydrocephalus was seen in all 5 cases causing obstruction in foramen monro (Figure 3). Cystic lesions was also seen in all 5 cases and hemorrhage in 2 cases. MRI characteristic features are given in table 1.

In immunohistochemistry, all the specimens were examined with Synaptophysin, Neuron–specific enolase (NSE), Glial Fibrillary acidic protein (GFAP). Among them Synaptophysin and Neuron–specific enolase (NSE) were positive in all cases and Glial Fibrillary Acidic Protein (GFAP) was only positive in 2 cases (Table 2).

### Discussion

Oligodendroglioma mostly arises in the fronto temporal region and rarely found in lateral ventricle. Calcification can be seen anywhere within the tumor and is larger than CN, It can erode inner table of the calvarium which is the distinguish feature of the oligodendroglioma.

Ependymoma occurs early in life and frequently cystic and hemorrhagic in nature. It is often centrally necrotic and primarily located at the body of lateral ventricle in Delta area with a broad base connected to lateral wall. Calcification is less common than in CN with significant enhancement after contrast injection.

Subependymal giant cell astrocytoma is a childhood disease, mostly seen near the foramen monro and attached to the ependymal surface under head of caudate nucleus. 90% of them are associated with tuberous sclerosis with fine calcification. Sometimes peritumoral edema can be found which is rare in CN.

Choroid plexus papillomas usually have irregular surface involving atria of the lateral ventricle. It is commonly seen in children less than 10 years old and have tendency to

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### Table 1: Clinical and MRI findings of central neurocytoma, +=positive, -= negative, hetero= heterogenous, Mod= moderate

<table>
<thead>
<tr>
<th>Serial No</th>
<th>Age/ Sex</th>
<th>Clinical presentation</th>
<th>Size (cm)</th>
<th>Location</th>
<th>Hydrocephalus</th>
<th>Cyst</th>
<th>Hemorrhage</th>
<th>MRI Findings</th>
<th>T1</th>
<th>T2</th>
<th>FLAIR</th>
<th>Contrast</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>32/F</td>
<td>Headache, dizziness, blurred vision</td>
<td>5.6x4.8x4.0</td>
<td>Both lateral ventricles</td>
<td>+</td>
<td>+</td>
<td>small</td>
<td>-</td>
<td>Iso SI</td>
<td>Mixed SI</td>
<td>High SI</td>
<td>Hetero+++</td>
</tr>
<tr>
<td>2</td>
<td>54/F</td>
<td>Headache, seizures</td>
<td>5.0x4.5x4.0</td>
<td>Left lateral ventricle</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Low SI</td>
<td>High SI</td>
<td>Low SI</td>
<td>--</td>
</tr>
<tr>
<td>3</td>
<td>20/F</td>
<td>Headache, Dizziness</td>
<td>8.0x5.0x4.0</td>
<td>Right lateral ventricle</td>
<td>+</td>
<td>+</td>
<td>small</td>
<td>-</td>
<td>Iso SI</td>
<td>Mixed SI</td>
<td>Iso SI</td>
<td>Mild Hetero+</td>
</tr>
<tr>
<td>4</td>
<td>26/M</td>
<td>Dizziness, headache, left limb weakness</td>
<td>7.4x5.8x5.0</td>
<td>Both lateral ventricles</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Iso SI</td>
<td>Mixed SI</td>
<td>Mixed SI</td>
<td>Mild +</td>
</tr>
<tr>
<td>5</td>
<td>31/M</td>
<td>Dizziness, headache, vomiting</td>
<td>5.0x4.5x4.0</td>
<td>Both lateral ventricles</td>
<td>+</td>
<td>+</td>
<td>small</td>
<td>+</td>
<td>Mixed SI</td>
<td>Iso to High SI</td>
<td>-</td>
<td>Hetero+++</td>
</tr>
</tbody>
</table>
invade outer margin of the ventricle causing vasogenic edema with significant contrast enhancement.

Meningioma: 5% of it localizes within the ventricular system, generally found in elderly women involving lateral or fourth ventricle with strong contrast enhancement.\(^1,3,15\)

CN is an intra ventricular WHO grade II tumor of the pure neuroepithelial origin. It is recognized as a rare CNS tumor with an incidence rate of less than 1% (0.2%- 0.5%) mainly affecting young population between 20- 40 years of age with no sex predominance.\(^1,7\) CN is typically located in the supratentorial ventricular system mostly involving anterior half of either lateral ventricle.\(^2\) It is found to be attached to septum pellucidum with a broad base in the region of foramen monro, it extends into 3\(^{rd}\) ventricle and or even to opposite lateral ventricle. According to literature (Nashio et al, Patil et al, Barbosa et al and Hassoun et al,) nuclei of the septum pellucidum could be the possible source of the tumor growth.\(^3,12\) Though pure neuronal tumors are uncommon but owing to their less aggressiveness nature they have better prognosis than common glial tumors.

The available reports mainly focused on their histopathological features, rather than on neuroradiological appearances. Though radiological appearance can strongly suggest the diagnosis, it alerts the neuropathologist for the need to perform additional immunohistochemistry or ultrastructural evaluation of the tumor specimen. Besides its typical location, MRI delineates CN as Iso-intense to gray mater in both T1 and T2 weighted images.

The cystic component has high signal intensity in T2 weighted and FLAIR images with areas of signal void due to tumor blood supply and due to calcification. The tumor shows mild to moderate enhancement after contrast injection.\(^4,9\) Though cystic component is common finding in CN, intratumoral hemorrhage is less common but if hemorrhage is seen within intra ventricular lesion then it strongly points toward CN. Previous reports have identified neurocytoma also arising in extra ventricular sites such as cerebral hemispheres (commonly frontal and followed by

Figure1: A 26 year old male with mass in bilateral lateral ventricle attach to septum pellucidum of size 7.4 cm × 5.8 cm x 5.0 cm extending to bilateral anterior horn containing cystic area. A: T1WI showing mass with iso intensity and high intensity is seen in rt. anterior horn corresponding to hemorrhage, B: mass is heterogeneous showing high intensity on T2WI, C: mixed SIintensity is seen on FLAIR image, D axial and E coronal contrast enhanced scan showing mild to moderate enhancement of hemorrhagic foci and tumor mass.
Central Neurocytoma

<table>
<thead>
<tr>
<th>Serial No</th>
<th>Synaptophysin</th>
<th>Neuron specific enolase (NSE)</th>
<th>Glial Fibrillary acidic protein (GFAP)</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>4</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

Table 2: Immunohistochemical study, + Positive, - Negative

parietal lobe), thalamus, cerebellum, pons, and spinal cord. Even atypical and malignant cases have also been recorded with pathological findings of atypical cell and mitotic behavior.

In this study, the maximum age was 54 years which is slightly higher. Among them one case was seen on the right lateral ventricle extending to occipital horn and invading into contralateral ventricle with small cystic area showing iso signal intensity (SI) on T1WI, mixed SI on T2WI and iso SI on FLAIR image with presence of mild contrast enhancement. One case was on seen in the left lateral ventricle involving septum pellucidum cystic in nature with septation showing low SI on T1WI, high SI on T2WI and FLAIR image with mild contrast enhancement. Three cases had bilateral lateral ventricular involvement with mass extending to frontal horn in two cases and occipital extension in one case, showing cystic areas and hemorrhage. All of them were iso SI on T1WI and iso to mixed SI on T2WI and on FLAIR image with mild to moderate contrast enhancement.

Regarding preoperative radiological diagnosis, one was CN, three were ependymoma and one was choroid plexus papilloma. Although CN has a characteristic neuroimaging finding, its rarity and variable presenting features create dilemma in radiological diagnosis. Therefore, radiologists are reluctant to give CN as a first possible diagnosis and finally diagnosis was confirmed as CN by Neuropathologists.

Neuropathologically CN is characterized by presence of small uniform round cells with a clear cytoplasm and round nuclei. Cells are arranged in honey combed architecture. Calcifications are observed within the tumor, which have resemblance toward Oligodendroglioma and Ependymoma under light microscopic examination leading to frequent misdiagnosis if further examination is not performed. On immunohistochemical studies, a positive reaction to the neuronal marker proteins such as, neuron specific enolase (NSE) and synaptophysin were found to be positive. Glial fibrillary acidic protein (GFAP) was positive only in some scattered reactive astrocytes within the periphery of the tumors. Ultrastructural evaluation with Electronmicroscope is most reliable, it shows numerous well formed synapses containing parallel bundle of microtubules which are specific for neuronal differentiation.

Ganglioneurocytoma is rare variant of CN which is characterized by differentiation towards ganglion cell.

Figure 2: A 37 year old female with mass of size 5.6 cm x 4.8 cm x 4.0 cm involving bilateral lateral ventricle extending to anterior and posterior horn of the right lateral ventricle causing hydrocephalus due to obstruction of the Foramen Monro. A: Mass showing isointensity in T1WI, B: T2WI showing mixed intensity within the lesion, C: FLAIR image showing high intensity and periventricular edema, D: Axial and E: Coronal view T1WI contrast scan showing heterogeneous significant enhancement.

Figure 3: A 54 year old female with cystic mass in the left lateral ventricle compressing contralateral ventricle showing obstructive hydrocephalus. A: FLAIR image showing iso SI of the septa B: No enhancement seen on contrast scan.
Conclusions

Even though CN are rare CNS tumors it should be first considered when a young patient with lesion in the supratentorial ventricular system in relation to septum pellucidum showing Iso signal intensity on T1 and T2 weighted images with some contrast enhancement, containing cystic component and occasional hemorrhage.

References
2. Chang EF, Gupta N. Pediatric CNS Tumors, chapter 8: Neuronal Tumors, pp168