Magnetic Resonance Imaging of a Case of Central Neurocytoma

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1. INTRODUCTION

Central neurocytomas are low-grade and slow growing primary brain tumours of neuronal origin that were firstly described by Hassoun et al. in 1982 (1). They comprise 0.25 to 0.5% of all primary brain tumors (2). They develop predominantly in young adults and are most frequently located in the lateral ventricles near the foramen of Monro or in the septum pellucidum and into the third ventricle. Neurocytoma can also be found into the fourth ventricle (3, 4) and spinal cord (5). This tumor can spread through cerebrospinal fluid and disseminate along craniospinal structures (6) and organs outside central nervous system, such as the peritoneum by means of a ventriculoperitoneal shunt (7). Neurocytoma localized outside the ventricular system is rarely found (8). Central neurocytomas of the aqueductal and pineal regions are also rare (9). Central neurocytomas are WHO Grade II neuroepithelial intraventricular tumours with fairly characteristic imaging features, appearing as homogeneous masses of variable size and enhancement within the lateral ventricle. They are typically seen in young patients, and generally have a good prognosis provided a complete resection can be achieved. Central neurocytomas are typically seen in young patients (20–40 years of age), and accounts for less than 1% (0.25–0.5%) of intracranial tumours. There is no reported gender predilection. In CT seen 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 ventricu-
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ular dilatation often present. MRI in T1 isointense to grey matter, heterogeneous, T1 with contrast mild-moderate heterogeneous enhancement, T2/Flair typically is 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 punctuate, haemorrhage (especially in larger tumours) is common, uncommonly results in ventricular haemorrhage (10).

2. CASE REPORT

A 45 year old man with 3 months of worsening daily headaches. These headaches were diffuse, lasted for several hours, and mostly occurred in the morning. She was initially diagnosed and treated for migraines but later he had epileptic atace and diplopia and neurolog recomended MRI.

We did MRI with protocol: pre-contrast MRI images of the brain were obtained using TSE/T2W sequence in axial/coronal planes; 3D–Hi-resolution T1W sequence in sagittal plane; FLAIR/T2W sequence in axial plane; thin slice FLAIR/T2W and Flash/T2W sequences in oblique coronal plane (perpendicular to temporal lobes) GRE/T2W sequence in axial plane for detection of heme products. Post-contrast images were acquired using TSE/T1W sequence in axial, coronal and sagittal planes. Diffusion weighted and ADC mapping MRI images were acquired using EPI sequence in axial plane.

Results: a 23x12mm heterogeneous mass within aqueductus cerebri, with calcified and/or hemorrhagic foci and extending downwards till fourth ventricle. It’s originating from the right paramedian posterior aqueductal wall (tectum), and also extending to and involving the tegmentum of mesencephalon at its right paramedian aspect. CSF flow obstruction secondary to described aqueductal mass, with resultant triventricular hydrocephalus. Marked transependymal CSF leak can be noted at periventricular white matter, secondary to severe hydrocephalus.

3. DISCUSSION

Central neurocytomas are more frequent from 20 to 40 years of age (about 70% of the cases). It is extremely rare in children. Signs and symptoms are caused by increased intracranial pressure due to cerebrospinal fluid block. The ma-
Majority of central neurocytomas are benign. Approximately 25% of these rare central nervous system tumors are more aggressive, with an MIB-1 labeling index >2% or atypical histologic features, and are classified as atypical neurocytomas (11).

MRI was helpful in refining the differential diagnosis. Central neurocytomas are usually T1 isointense, with variable intensity on T2 and a heterogeneous enhancement pattern and globular calcifications imparting the classic “soap-bubble” appearing lateral ventricular lesion.

Another characteristic feature of central neurocytomas is that they usually appear to arise from either the septum pellucidum or foramen of Monro.

Central neurocytomas are slow-growing tumors that are of neuronal origin. Histologically they resemble oligodendrogliomas and were originally thought to be unusual intraventricular oligodendrogliomas until their neuronal character was established by electron microscopy. Currently, immunostaining for synaptophysin can confirm the neuronal nature (12, 13). Treatment is with complete surgical resection, but radiation can be used for recurrence or incomplete resection.

Teaching Points: a) Central neurocytoma should be at the top of the differential in a young adult with a “soap bubble” appearing intraventricular lesion that appears to arise from the septum pellucidum; b) Complete surgical resection implies cure.

4. CONCLUSION

Central neurocytomas are rare, slow-growing, intraventricular tumors that are of neuronal origin. Immunohistochemically stains and transmission electron microscopy can provide useful diagnostic information. Currently, immunostaining for synaptophysin can confirm the neuronal nature. It is classified as WHO grade II. By using clinical, demographic, and imaging findings, one can significantly limit the differential diagnosis for many of the most common intraventricular tumors.

Total tumor excision is associated with favorable prognoses. Postoperative radiotherapy may be considered for cases of subtotal excision, anaplastic histological variants, or recurrent tumors.

Characteristic MR Findings include a main solid portion with intratumoral cystic changes and broad attachment to the septum pellucidum and /or lateral wall of the lateral ventricle. MRI is helpful in defining tumor extension, which is important in preoperative planning. Although central neurocytoma is a relatively rare lesion, it should be considered in the differential diagnosis of intraventricular lesions in the presence of such typical MR findings. However, a definitive diagnosis requires immunochemical study and electron microscopy.

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