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Case Report

Central Neurocytoma: Case Report and Literature Review

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Abstract: Intraventricular neoplasms are readily seen on cross-sectional images,consideration of the tissue within and the clinical findings provide the means to limit the differential diagnosis when analyzing an intraventricular mass on an imaging study. Central neurocytomas are rare, slow-growing, intraventricular tumors that are of neuronal origin, typically located in the lateral ventricles, near the foramen of Monro with characteristic imaging features. Central neurocytoma is a slow growing, benign neoplasm with a favorable prognosis and by using clinical and imaging findings one can significantly limit the differential diagnosis.
Keywords: Central Neurocytoma, Intraventricular Neoplasm, Magnetic Resonance Imaging, Cross-sectional Imaging.

INTRODUCTION

Central neurocytoma (CN) is a rare WHO Grade II neuroepithelial intraventricular tumors, constituting only 0.25-0.50% of all intracranial tumors [1]. CN is a slow growing, benign neoplasm with a favorable prognosis and affects mainly young adults (20-40years of age) [2-4].There is no reported gender predilection [5] .CN is typically located in the lateral ventricles, near the foramen of Monro, with a characteristic attachment to the septum pellucidum. We report here a 34 years old man who had a large, intraventricular brain mass with characteristic imaging features on MR. After resection, pathological diagnosis was central neurocytoma.

CASE REPORT

A 34 years old, previously healthy male presented with 3 months of worsening daily headaches. These headaches were diffuse, lasted for several hours, and mostly occurred in the morning. He was initially diagnosed and treated for migraines. In spite of being on migraine medications, however, his headaches worsened, and started to become associated with nausea, vomiting and blurry vision.

On clinical examination, there was mild papilloedema, his vital signs were unremarkable and had no other focal neurological deficit. His laboratory findings were within normal limits.

MRI Brain study revealed a large, lobulated, well circumscribed, heterogeneous signal intensity intra ventricular mass of size approximately 3.9 x 3.7 x 3.7 cm attached to septum-pellucidum near foramen of monro and epicentral in the left lateral ventricle body region (Fig. 1& 2).

Mass was heterogeneous isointense to grey matter on T1 weighted imaging (Fig. 2) and iso to hyperintense on T2 / FLAIR imaging (Fig. 1) & showed multiple intratumoral & peritumoral cystic areas (soap-bubble appearance). Punctuate foci of T1 / T2 hyperintensities were noted with corresponding marked blooming effect on susceptibility-weighted imaging suggestive of presence of calcification.Fluid- fluid levels of differential signal intensity were noted in the few tumoral cyst suggestive of hemorrhages (Fig. 3).

There was moderate dilatation of lateral ventricles with entrapment of left frontal horn showing slightly higher signal intensity fluid as compared to CSF – likely due to long duration stagnation of CSF.

On post contrast imaging there was moderate inhomogeneous enhancement of mass lesion (Fig. 2b). Superiorly lesion was adherent to inferior margin of body of corpus callosum. Diffuse band like increased signals were noted in the posterior body & splenium of corpus callosum.Lesion was closely abutting the ependymal linening in the superior aspect in body of

ventricles with periventricular T2/ FLAIR hyperintensity although no obvious abnormal enhancement was noted.MR spectroscopy showed the elevated choline peak with markedly suppressed NAA.

Imaging characteristic & location of the lesion were highly suggestive of central neurocytoma.Pathological diagnosis of tumor was central neurocytoma.

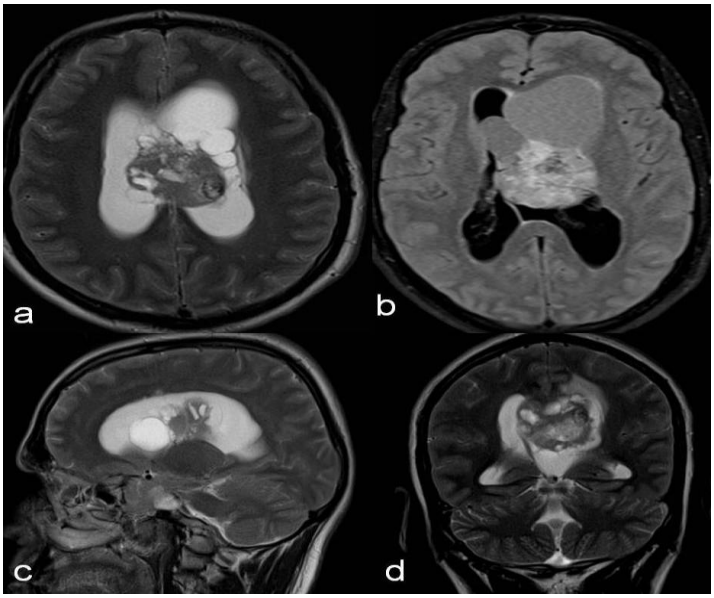

Fig.1(a, b, c, d): (a) T2 Axial,(b) Flair,(c) T2 Saggital and (d) T2 Coronal MR Images shows a large, lobulated, heterogenous, iso to hyperintense,intra ventricular mass showing multiple intratumoral and peritumoral cystic areas.(soap-bubble appearance)

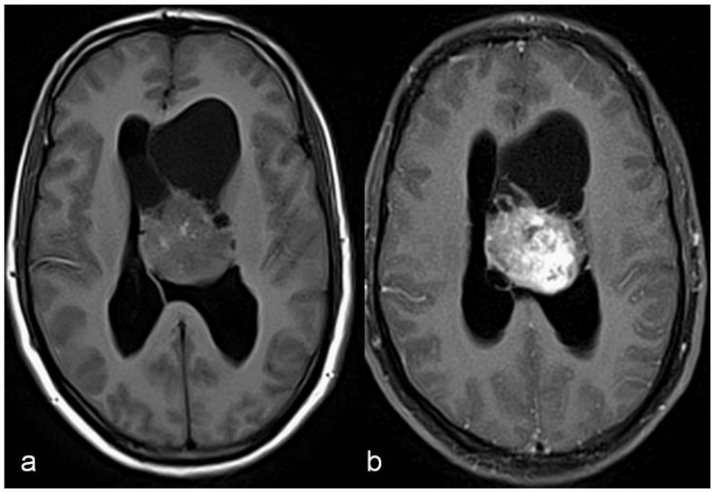

Fig. 2 (a, b): T1W Axial (a)Pre and(b) Post contrast MR Images showing heterogenous, isointense, intraventricual mass with moderate, inhomogenous enhancement

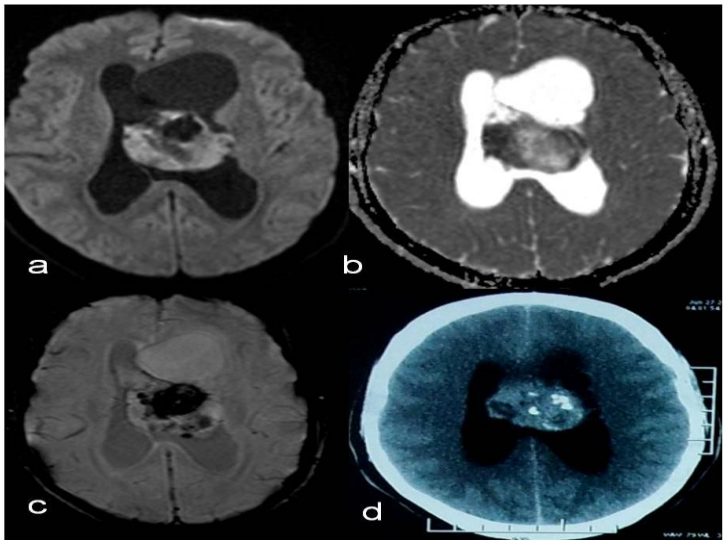

Fig. 3(a, b, c, d): (a)DWI and (b)ADC MR Axial Images-solid appearing areas of mass showing diffusion restriction suggestive of high cellularity.(c)SWI shows marked blooming in mass suggesting hemorrhage/calcification.(d) CT Axial Image confirming tumoral amorphous calcification

DISCUSSION

Initially described in 1982 by Hassoun et al, central neurocytoma (CN) is a rare tumor of neuroglial origin [6]. The initial description classified them as WHO grade I lesions, however this was upgraded in 1993 to WHO grade II as it was recognized that at least some of these tumors exhibited more aggressive behavior [5]. Central neurocytomas constitutes only 0.25-0.50% of all intracranial tumors. However, they are the most common primary intraventricular tumors in adults. CN typically affects young adults around the third decade. They are characteristically located in the supratentorial ventricular system. Half of the cases involve the lateral ventricles near the foramen of Monro, whereas 15% are located in both the lateral and third ventricles. About 13% of central neurocytomas are bilateral and only 3% occur in the third ventricle as an isolated location. There is a single case report of a central neurocytoma that arose in the fourth ventricle [7]

The typical clinical presentation is with signs and symptoms of increased intracranial pressure induced by obstructive hydrocephalus. Patients may present with acute symptoms related to sudden development of ventricular obstruction and elevated intracranial pressures, generally there is more insidious onset of symptoms. Schild et al analyzed 27 patients with central neurocytomas regarding their presenting symptoms, and 93% of patients complained of headaches, 37% had visual changes, and 30% experienced nausea and vomiting at presentation [8]. In another study by Wang et al, out of 27 patients, 21 presented with headache and 6 with vomiting [9].

On MRI, central neurocytoma (CN) is typically heterogeneous on all sequences, but it is isointense to cerebral cortex on T1-weighted images and isointense-to-hyperintense on T2-weighted images. Areas of low signal intensity or absent signal on both T1- and T2-weighted images can represent calcification, cyst, hemorrhage, and tumor vessels. Most tumors show some degree on enhancement [9]. CN may show a characteristic metabolic profile on proton MR spectroscopy, consisting of high choline and low NAA peaks plus glycine and/or alanine peaks [11, 12].

At CT, central neurocytoma (CN) is isoattenuating or slightly hyperattenuating and is a well-demarcated, lobulated mass with moderate to strong heterogeneous contrast enhancement. Calcification is seen in about 50% of cases and is usually clumped, amorphous, or globular [1, 6].

Tumor location is a key imaging finding. CN is typically centered on the midline near the foramen of Monro with a broad-based attachment to the septum pellucidum. The imaging differential diagnosis consists of other intraventricular tumors that occur in young adults, including, but not limited to, endymoma, subependymomas, subependymal giant cell astrocytoma, choroid plexus papilloma, choroid plexus carcinoma, meningioma and intra ventricular metastasis.

Ependymomas are typically calcified, are more common in children, are more common in the fourth ventricle, and show intense enhancement on contrast-enhanced images. Subependymomas and central neurocytomas have an affinity for the anterior portion of the lateral ventricle, and both commonly demonstrate a heterogeneous cystic appearance on cross-sectional images. Subependymomas are more common in older adults, whereas central neurocytomas are more common before 40 years of age. Subependymal giant cell astrocytomas always lie near the foramen of Monro and are characterized by frequent calcification, intense enhancement on contrast-enhanced studies, and the presence of other stigmata seen in tuberous sclerosis.

When a mass is centered on the choroid plexus, a highly vascular tumor—either choroid plexus papilloma, choroid plexus carcinoma, meningioma, or metastasis—should be suspected. The characteristic heavily lobulated appearance of a choroid plexus tumor favors this diagnosis over other possibilities, although it is not always possible to distinguish between the more common benign form, the choroid plexus papilloma, and the less common malignant counterpart, the choroid plexus carcinoma [7, 10]

Using dynamic susceptibility contrast-enhanced MR perfusion to assess intraventricular tumors, central neurocytomas reveal intermediate vascularity (mean relative cerebral blood volume [rCBV] of between 2 and 3), compared with highly vascularized tumors such as papillomas, meningiomas, and renal carcinoma metastases, which have a mean rCBV of greater than 3, and poorly vascularized tumors such as ependymomas and subependymomas, showing mean rCBV of less than 2[13].

Extraventricular neurocytomas (EVN) have already been recently described. They can be situated in the cerebral hemispheres, pons, cerebellum, spinal cord, and retina.Extraventricular neurocytomas are typically well-demarcated masses, may have cystic components in up to 50%, enhance variably, and may or may not be associated cerebral edema.EVN is a rare neoplasm that can have significant overlap in imaging appearance with other primary brain neoplasms;therefore, it is difficult to make an accurate preoperative diagnosis.EVN should be considered in the differential diagnosis when a large cerebral parenchymal mass with cystic necrosis,calcification, and/or hemorrhage foci, and extensive enhancement is encountered in younger patients[14].

Central neurocytomas have good prognosis. The best treatment is complete surgical resection. Patients with incomplete excision may benefit from radiotherapy. In contrast with the more aggressive atypical neurocytomas,well-differentiated neurocytomas are associated with an excellent long-term survival (5 year survival 81%).Cases of CSF dissemination have been reported, but are rare[5].

CONCLUSION

Central neurocytomas are rare, slow-growing, intraventricular 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. 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.

REFERENCES

1. Hassoun J, Söylemezoglu F, Gambarelli D, Figarella-Branger D, von Ammon K, Kleihues P; Central neurocytoma: a synopsis of clinical and histological features. Brain Pathol., 1993; 3(3): 297-306.
2. Figarella-Branger D, Soylemezoglu F, Kleihues P, Hassoun J.; Central neurocytoma. In Kleihues P, Cavenee WK. Pathology and genetics of tumors of the nervous system. Lyon: IARC Press; 2000:107-109.
3. De Tommasi A, D'Urso PI, De Tommasi C, Sanguedolce F, Cimmino A, Ciappetta P; Central neurocytoma: two case reports and review of the literature. Neurosurg Rev., 2006; 29: 339-347.
4. Schmidt MH, Gottfried ON, von Koch CS, Chang SM, McDermott MW; Central neurocytoma: a review. J Neurooncol., 2004; 66(3): 377-384.
5. Smith A, Smirniotopoulos J, Horkanyne-Szakaly I; From the Radiologic Pathology Archives: Intraventricular Neoplasms: Radiologic-Pathologic Correlation. Radiographics, 2013; 33 (1): 21-43.
6. Hassoun J, Gambarelli D, Grisoli F, Pellet W, Salamon G, Pellissier JF; Central neurocytoma. An electron-microscopic study of two cases. Acta Neuropathol. (Berl)*;* 1982;56(2):151-156.
7. Koeller KK, Sandberg, GD; From the Archives of the AFIP: Cerebral Intraventricular Neoplasms: Radiologic-Pathologic Correlation. RadioGraphics, 2002; 22:1473-1505.
8. Schild SE, Scheithauer BW, Haddock MG, Schiff D, Burger PC, Wong WW et al.; Central neurocytomas. Cancer, 1997; 79: 790-795.
9. Wang M, Jia D, Shen J, Zhang J, Li G; Clinical and imaging features of central neurocytomas. J Clin Neurosci, 2013; 20(5): 679-685.
10. Koeller KK, Rushing EJ; From the Archives of the AFIP: Oligodendroglioma and Its Variants: Radiologic-Pathologic Correlation. RadioGraphics, 2005; 25:1669-1688.
11. Chuang MT, Lin WC, Tsai HY, Liu GC, Hu SW, Chiang IC; 3-T proton magnetic resonance spectroscopy of central neurocytoma: 3 case reports and review of the literature. J Comput Assist Tomogr., 2005; 29(5): 683-688.
12. Jayasundar R, Shah T, Vaishya S, Singh VP, Sarkar C; In vivo and in vitro MR spectroscopic profile of central neurocytomas. J Magn Reson Imaging, 2003; 17: 256-260.
13. Holveck A, Grand S, Boini S, Kirchin M, Le Bas JF, Dietemann JL et al.; Dynamic

susceptibility contrast-enhanced MRI evaluation of cerebral intraventricular tumors: preliminary results. J Neuroradiol., 2010; 37(5): 269-275.

14. Yang GF, Wu SF, Zhang LJ, Lu GM, Tian W, Shah K; Imaging Findings of Extraventricular Neurocytoma: Report of 3 Cases and Review of the Literature. AJNR Am J Neuroradiol., 2009; 30: 581– 585.