ATYPICAL CENTRAL NEUROCYTOMA WITH BIVENTRICULAR INVOLVEMENT: A RARE CASE REPORT

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ABSTRACT

Atypical central neurocytomas are rare tumors of central nervous system usually encountered in lateral ventricle but rarely may involve the third ventricle and also extraventricular zones. It can be diagnosed by presence of histological anaplasia/high proliferative index confirmed by immunohistochemistry. It affects mostly young adults of 2nd and 3rd decades though infrequently it may be seen in children also. The prognosis of atypical neurocytoma is not good with recurrence and dissemination to surrounding brain material. We describe a case of atypical central neurocytoma extending to third ventricle and surrounding brain with high proliferative potential proved by immunohistochemistry.

Key words: Central Neurocytomas, Immunohistochemistry, Biventricular.

INTRODUCTION

Central neurocytomas are rare benign tumors comprising 0.1 to 0.5% of the primary brain tumors and predominantly seen in intraventricular location. Extraventricular neurocytomas have been described to affect brain parenchyma like cerebral hemisphere, thalamus, cerebellum, pons, amygdale and retina (Juratli et al., 2013). It occurs mainly in young adults in 2nd & 3rd decade.

Few of them show nuclear atypia and high proliferative index with MIB1 more than 2% and have a poor prognosis. It is thought to arise from the precursor cells of septum pellucidum and subependymal cells of lateral ventricles. WHO included neurocytoma as a separate entity in its new classification in 1993 and graded it as Grade I tumour (Louis et al., 2007). But it was suggested to label it as Grade II if atypia or high proliferative index is present.

We describe a case of central atypical neurocytoma in a 27 year old female involving lateral and third ventricle with immunohistochemical confirmation.

CASE REPORT

A 27 year old female presented with headache for two and half years and forgetfulness for one year. On examination her visual acuity was 6/24 (B/L), higher motor function-normal, bilateral papilledema, no motor/sensory deficit. (Figure 1) CT scan showed a large hyperdense globular intraventricular lesion intensely enhancing on contrast. MRI revealed large globular T1 hypointense, T2 heterogeneous hyperintense mass lesion showing heterogeneous enhancement extending to left side ependymal region and filling the entire third ventricle. (Figure 2a & b) A clinicoradiological diagnosis of choroid plexus papilloma/gliala was given.

Squash cytology was reported as oligodendroglioma due to presence of small round cells with moderate amount of pale eosinophilic to clear cytoplasm and round vesicular nuclei. The gross received was multiple bits of greyish white tissue together measuring 1x.5x.5 cm. Hematoxyline & Eosin stained...
sections showed presence of diffusely arranged tumour tissue with clear to pale eosinophilic cytoplasm, mild nuclear atypia and fine chromatin, intervening areas of acellular eosinophilic fibrillary zones and proliferation of branching thin walled blood vessels. Few area showed perivascular arrangement of tumor cells showing a pseudorosettoid pattern (Figure.3a,b,c). Due to presence of clear to pale eosinophilic cytoplasm in tumor cells possibility of oligodendroglioma, central neurocytoma, ependymoma and clear cell meningioma were considered as differential diagnosis. For confirmation immune histochemistry was done. It showed strong positive reaction for synaptophysin (Figure.4a) and CD56 (NCAM), negative for GFAP (Figure.4b), chromogranin and EMA (Figure.4c). MIB 1(Figure.4d) index was high around 5-7%. So our diagnosis was confirmed as atypical central neurocytoma.

Figure 1. Photograph of patient with neurocytoma before operation

Figure 2. A&B-MRI findings showing lesion in lateral and third ventricles

Figure 2a

Figure 2b

Figure 3. A,B,C-H&E stained section showing small round cells with clear cytoplasm and perivascular arrangement

Figure 3a.

Figure 3b.
Figure 3c.

DISCUSSION AND CONCLUSION

Neurocytomas are rare benign intraventricular tumors of the central nervous system usually located in the lateral ventricle in the zone of foramen of Monro. Depending on location, it can be lateral ventricular, biventricular (both lateral and third ventricle) or rarely only third ventricular (3% of cases). Besides it can also be seen in extraventricular location (Chinnikatti et al., 2010).

These are usually benign with a favourable prognosis accounting for 75% of neurocytomas. But neurocytomas with a high proliferative index i.e. MIB-1 labelling index >2% with/without anaplastic features are designated as atypical and are noted to have worse clinical outcome and higher recurrence rate compared with normal neurocytomas (Söylemezoglu et al., 1997; Brat et al., 2007). MIB-1 LI score >3% is considered as a bad prognostic indicator and said to have an adverse outcome (Rades et al., 2004). A retrospective analysis of 15 cases of neurocytomas done by Mackenzie et al showed that the proliferative potential is a better predictor for clinical outcome and prognosis than histological features. The terms “atypical” and “anaplastic” are not suitable to describe these lesions as they indicate a typical histologic appearance. Therefore most accurate designation suggested would be “proliferating neurocytoma” (Mackenzie, 1999).

In the above said case also there was no hostomorphological atypia but the proliferative index was high (6-7%) which also was again confirmed as the patient did not survive more than 6 months. The limitations of the pathological diagnosis consist of morphology very similar to ependymoma, which also contains perivascular acellular areas, but subtly fewer
monomorphic nuclear shapes and a more fibrillar matrix. In this case, the first diagnosis of an oligodendroglioma, which is much more likely because of the perinuclear clearing of cytoplasm and delicate branching vasculature, was based only upon morphology. However, using immunohistochemistry, it was possible to reach the correct diagnosis of neurocytoma.

Our case was GFAP negative which rules out ependymoma and oligodendroglioma. Other differential diagnosis was clear cell meningioma, which are usually seen in cerebellopontine angle and must show predominantly clear cell morphology. These are also biologically aggressive tumors and the diagnosis should be made carefully. Since the present case didn’t have the criteria satisfying clear cell meningioma and was negative for EMA, further confirmed our diagnosis as neurocytoma. The proliferative index using MIB1 was around 5-7% in the highest proliferating zones. So a final diagnosis of atypical central neurocytoma was given. This patient died within 6months of surgery indicating the poor prognosis of these tumors. The treatment protocol usually followed for these cases are complete resection (CR), complete resection plus radiotherapy, incomplete resection (IR) and IR plus radiotherapy. Even cases of incomplete resection can also be treated by chemotherapy and have a better prognosis (Dirk et al., 2004). This patient expired before receiving radiotherapy. But there are reports documenting decrease in Ki67 after radiotherapy.

In summary, the case is of interest to the readers due to its unusual course, extensive involvement of both lateral and third ventricles as well as its monomorphic histological appearance with high proliferative index proved by immunohistochemistry.

REFERENCES