Successful Central Neurocytoma Management: A Rare Brain Tumor

Sir,

Central neurocytoma (CN) is a benign-low malignant (WHO Grade-II) extremely rare primary brain tumor with frequency of 0.1 - 0.5%, and male : female ratio of 1:1.1.[1,2]

A 24-year man, presented with history of headache, vision and gait problems, and generalised fits with urinary incontinence. He had no focal neurological deficit (GCS 15/15), brisk lower limb reflexes, and up-going planters bilaterally. CT scan brain showed a large tumor in the lateral ventricles, grossly dilating them, later confirmed on MRI (Figure 1a-d). The tumor was totally removed (piecemeal but radical) with the help of an operating microscope. No ventricular drains were used. The tumor was greyish and friable, histologically comprising of sheets of cells, central nuclei, salt and pepper chromatin, fibrillary background, areas of chicken wire vascular pattern, and rosette formation. Synaptophysin was positive. Post-operatively, the patient regained full consciousness with intact memory and no neurological deficit, but minimal personality changes. On follow-up review after four weeks, he had fully recovered. Follow-up MRI showed no residual or recurrent tumor.

The second patient was a 30-year male, presenting with generalised fits, confusion (GCS = 12/15), pupils bilaterally reactive, no focal neurological deficit, brisk deep tendon reflexes, and bilateral up-going planters. An urgent CT brain, followed by MRI brain, showed large intraventricular tumor (Figure 1e-h). Radical removal of a large, necrotic, soft tumor was done, which was arising from the septum pellucidum and adherent to the lateral ventricular floor in a few places; but it was easily separated. Histopathology report confirmed CN with synaptophysin positive and Ki-67 labelling index <1%. Patient regained consciousness immediately after the surgery; but had mild dysphasia, and personality changes. On follow-up after four weeks, his speech and behaviour were found to have improved. Postoperative MRI showed no residual tumor or recurrence, and the ventricles had regressed in size. He resumed his job three months after the surgery, as a lecturer, and is able to drive to work.

Both patients underwent frontal osteoplastic craniotomy, using right inter hemispheric transcallosal microsurgical approach, followed by oral dexamethasone course, tapered off over two weeks, with oral sodium valproate. Both the patients were being followed up at regular intervals.

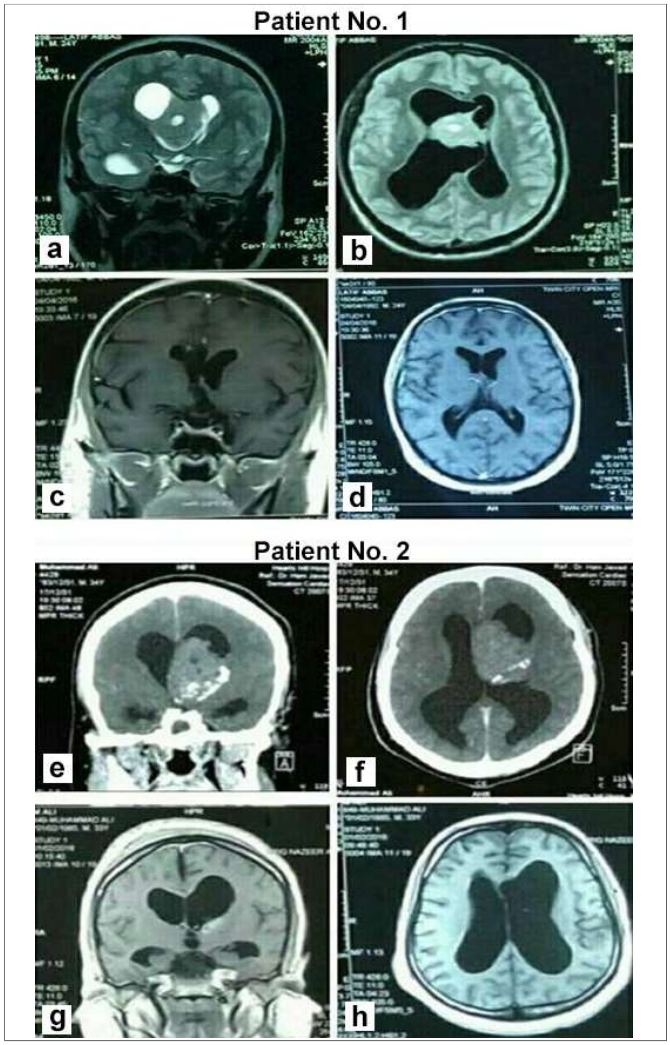

Figure 1: Patient 1: Preoperative T2W-MRI brain coronal **(a)** T1W-MRI axial sections **(b)** large right intraventricular tumor, gross dilatation of right lateral ventricle, compression the left lateral ventricle. Postoperative T1W-MRI brain scans **(c)** coronal **(d)** axial sections showing successful complete resection, with no residual tumor.
Patient 2: Preoperative coronal **(e)** axial **(f)** sections CT scan brain: large tumor involving left lateral ventricle with calcifications. Postoperative T1W-MRI brain scans coronal **(g)** axial sections **(h)** after successful GTR.

CN clinically presents with confusion, seizures,[2] signs of raised intracranial pressure or intraventricular hemorrhage.[3] On MRI, it appears heterogeneous. Moderate to strong enhancement with contrast (in solid portions of tumor),[2] ventricular dilatation, hemorrhage, necrotic or cystic changes, vascular signal voids or calcifications are commonly noted.[4] On CECT, it appears as well demarcating heterogeneous mass with isoattenuation/slight hyper attenuation.

Immunohistochemistry is strongly positive for synaptophysin and antibody to ßIII Tubulin, marker Ki67/MIB-1 labelling index being positive in upto 5%.[4] Morphologically, tumor cells are round, in clusters with nuclear-free fibrillary areas mimicking neuropil, calcifications and delicate arborizing capillaries.[3]

Treatment of choice is gross total resection (GTR), *via* an open microsurgical approach or with a neuro-endoscope. Recurrence is seen in 20% cases.[5] Adjuvant radiotherapy, in case of incomplete resection or evidence of atypia, improves local control but not the survival.[5] Role of chemotherapy has not been well characterised.[6]

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