Malaysian Journal of Medical Sciences, Vol. 7, No. 2, July 2000 (32-37)

CASE REPORT

BIVENTRICULAR CENTRAL NEUROCYTOMA

A R Ariff, AS Muda, JM Abdullah*
Gurjeet K, B M Biswal*****

Department of Radiology, *Neuroscience Unit
****Department of Pathology, *****Department of Radiotherapy**
School of Medical Sciences, Universiti Sains Malaysia
16150 Kubang Kerian, Kelantan, Malaysia

A case of biventricular neurocytoma is reported. A 36 year old Malay lady presented with headache of 8 months duration. Physical examination revealed signs of increase intracranial pressure. CT-scan and MRI showed tumour in both lateral ventricles. Patient underwent tumour debulking followed by adjuvant radiotherapy. The radiological appearances of central neurocytoma are discussed.

Key words : *Central neurocytoma, treatment, radiology.*

Case report

A 36-year old Malay lady presented with an 8-month history of frontal headache which was throbbing in nature. There was no history of blurring of vision, nausea, numbness, vomiting, trauma, loss of weight, loss of appetite or neck stiffness. Prior to admission, the headache became more frequent and severe and was not relieved by analgesics. Her husband noticed a change in her behaviour. She had no previous significant medical history. Her social and family history were unremarkable.

On examination, the patient was alert and conscious. The vital signs were normal. Funduscopic examination revealed papilloedema on the left side. Other systems were essentially normal. Routine

Figure 1*: Axial NCCT brain reveals lobulated mass in both lateral ventricles. The mass has solid and cystic components.*

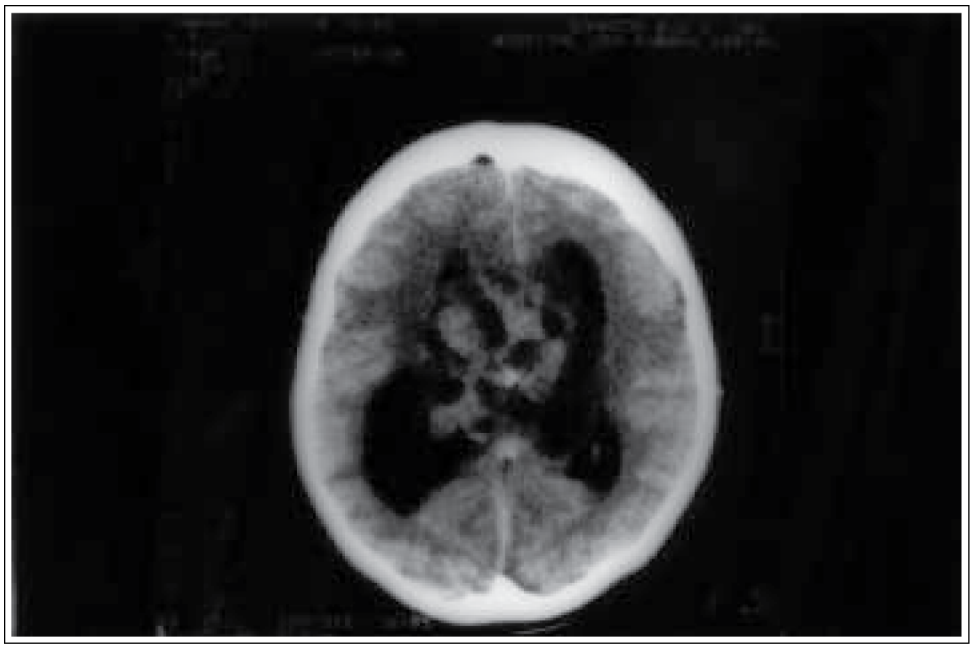
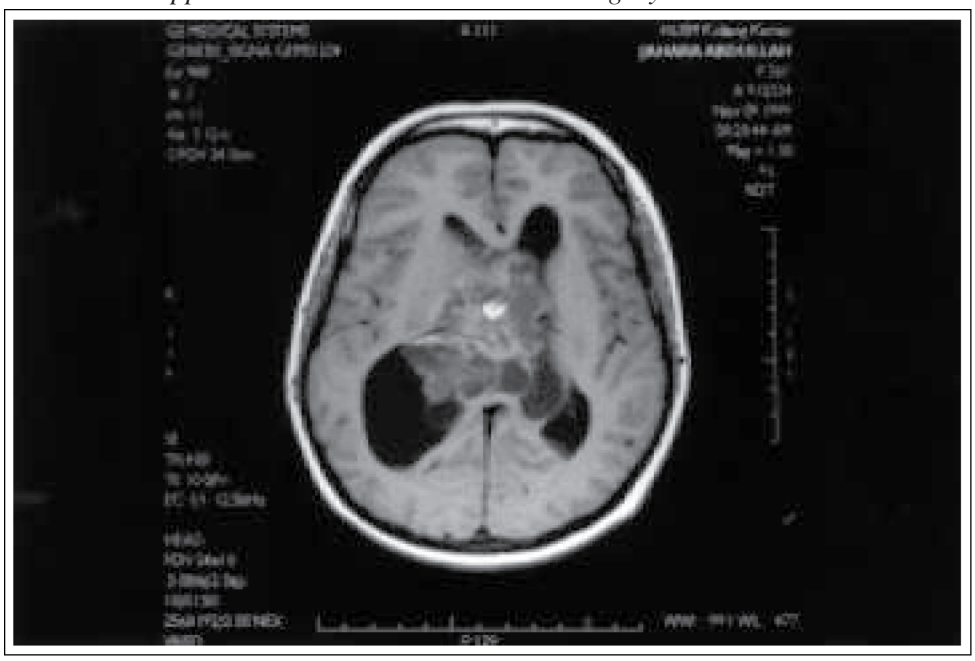


Figure 2: Axial T1WI shows lobulated mass in both lateral ventricles which appear to be isointense to the cortical gray matter.



blood investigations were also within normal limits.

Computed tomography (CT-scan) revealed a lobulated mass predominantly in the body of both lateral ventricles associated with hydrocephalus (Figure 1].

Areas of coarse calcification were not seen on CT-scan as expected in intraventricular neurocytoma. Attachment to the septum pellucidum could not be ascertained.

Magnetic resonance imaging revealed alobulated mass in both lateral ventricles which was of isointense signal intensity relative to cortical gray matter on T1WI (Figure 2). The mass showed evidence of cystic spaces and vascular flow void areas on T2WI (Figure 3). MRI appearance suggested an attachment of mass to the septum pellucidum (Figure 4). There were no evidence of haemorrhage. The solid component enhanced intensely following intravenous gadolinium (Figure 5).

Figure 3: Axial T2WI reveals cystic spaces and vascular flow void areas. Note the isointense solid component (arrow).

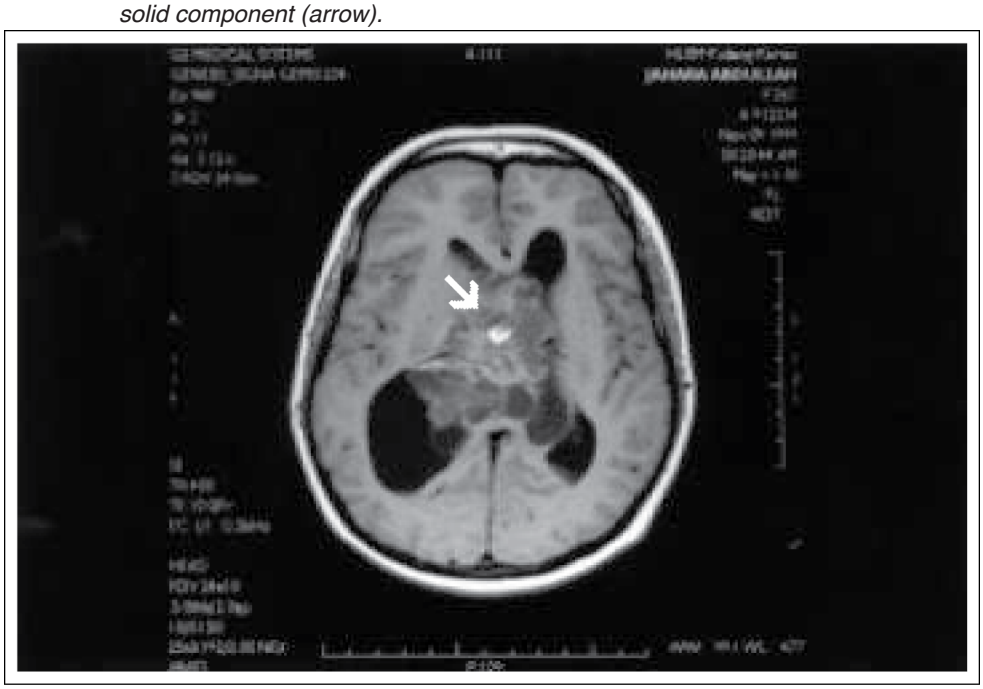
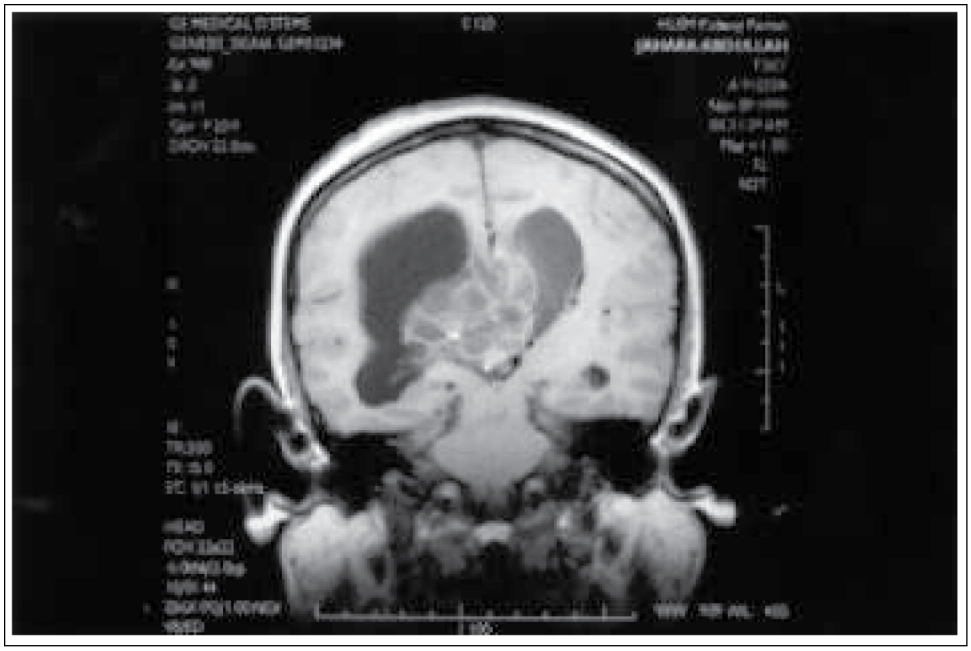


Figure 4. Coronal TIWI shows biventricular mass attached to the septum



Cerebral angiography revealed a lesion supplied by the branches of right anterior cerebral and middle cerebral arteries (Figure 6). There were no supply from the anterior or posterior choroidal arteries. Immediate treatment with intravenous dexamethasone and Rickham's catheter insertion were initiated to reduce the intracranial pressure.

Subsequently, the patient underwent tumour debulking. Histopathological examination showed uniform round cells with central nuclei, clear cytoplasm and well defined cell membranes. Immunohistochemical staining for glial fibrillary acid protein (GFAP) and synaptophysin were positive.

The post-operative course was complicated by an extradural haematoma which was completely evacuated.

In view of the residual tumour, she was subjected to radiotherapy, with a total dose of 54 Gy in 27 fractions over 6 weeks.

Discussion

Figure 5. Coronal T1WI post gadolinium DTPA shows enhancement of the solid component.

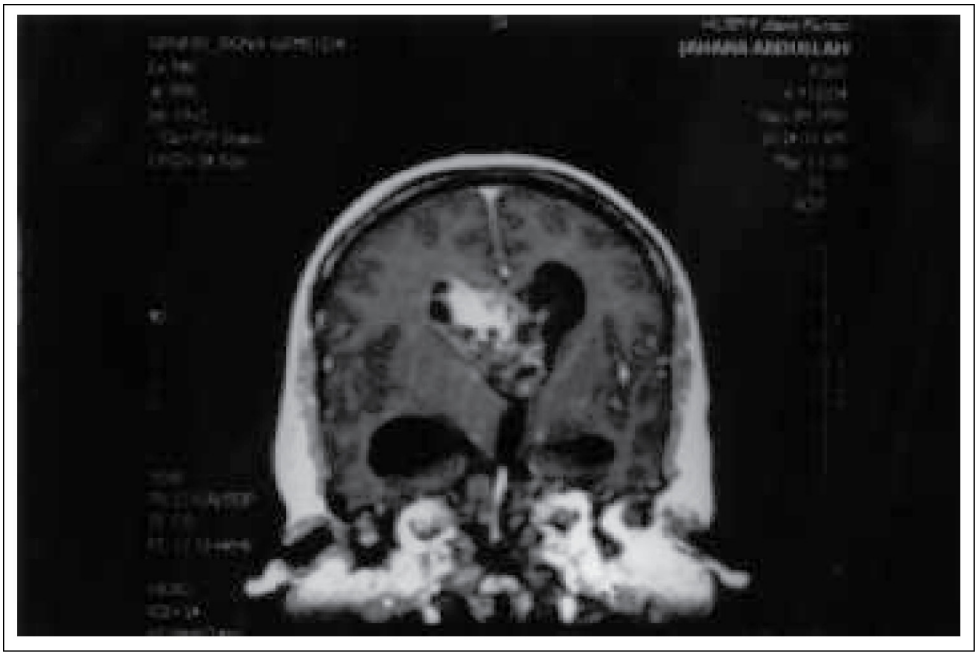
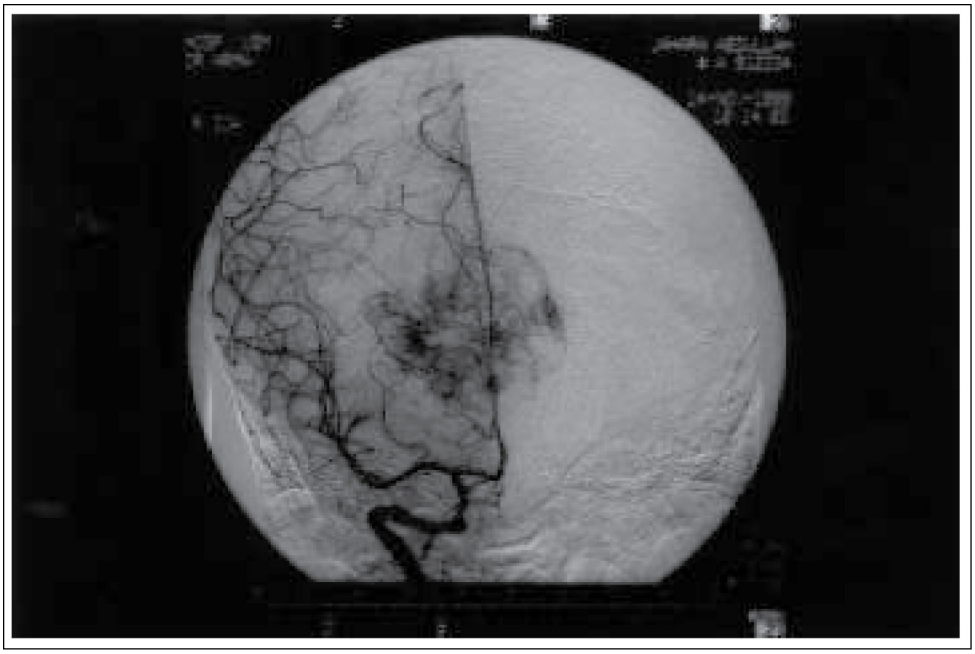


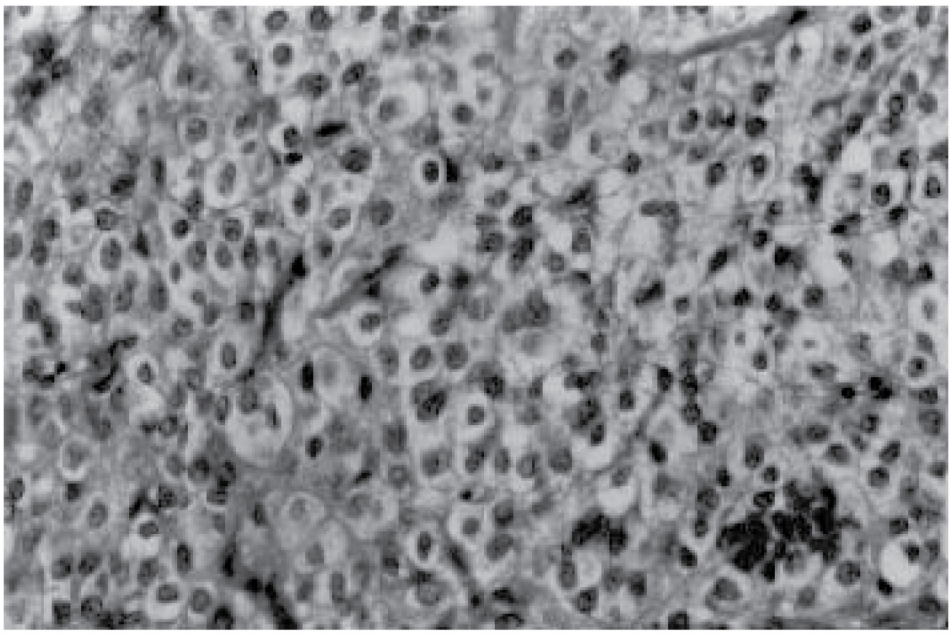
Figure 6: Right internal carotid angiogram reveals lesion supplied by the branches of right ACA and MCA.



Central neurocytoma is a small cell neuronal tumour that occurs in the lateral and third ventricles (1). It is composed of mature neuronal cells, giving an exception to the rule that neuronal cells do not replicate after fetal life. It was described previously by Hassoun et. al (2). However it was erroneously labelled as intraventricular oligodendroglioma. Following that it was termed as central neurocytoma by the World Health Organisation (3).

Typically the tumour presents in young adults (mean age : 25-30 years). The patient usually presents with nausea, vomiting, headache and papillooedema due to obstructive hydrocephalus (4). Commonly the lesion is intraventricular and attached to the septum pellucidum. It is a slow growing benign tumour, usually without extraventricular extension. A few unusual cases of extraventricular

Figure 7: Microscopic appearance of neurocytoma. Haematoxylin and Eosin, x 400.



extension have been reported (5).

Diagnosis

The general management guidelines for central neurocytoma are not yet clearly defined. The typical CT appearance of central neurocytoma is that of a coarsely calcified, well circumscribed intraventricular tumour found in the body or frontal horn of the lateral ventricle. It is usually in close proximity to the foramen of Monro and typically attached to the septum pellucidum (6). There may be bleeding within the tumour mass and hydrocephalus is almost always present. The tuntur enhances mild to moderate with intravenous contrast.

The tumour is mainly isointense to cortical gray matter on both T1 and T2WI. They may have cystic spaces and calcification, giving rise to signal void areas or heterogenous intensity within the tumour mass. Magnetic resonance imaging is an excellent modality in demonstrating the characteristic attachment of the tumour to the septum pellucidum (7). There is no specific angiographic feature for central neurocytoma. Computed tomography still plays a very important role especially in demonstrating the calcification within the tumour.

Histologically, the tumour has a uniform appearance of small, round cells that mimics

Table 1: Differential diagnoses of an intraventricular tumour

Tumour/ Features	Central Neurocytoma	Meningioma	Choroid plexus papilloma	Oligodendroglioma	Ependymoma
Origin	Mature neurons	Meningothelial cells concentrated in arachnoid villi.	Choroid epithelium	Oligodendrogliocytes ±Astrocyte	Intraventricularly/Ependymal rest in the white matter
Age	Young adult (25-30) years	35-75years		Any age.Most 4-5D	Predominantly <5y.o Smaller peak in 3rdD
Sex(M:F)	-	M>F	3:2	5:4	0.8:1
Location	Body±frontal horn+3rd Ventricle	Exclusively in trigone	Adult:3rd v/4th V Children:Trigone	Frontal>Temporal> parietal> brainstem. Less commonly extension into thesubarachnoid space	Infratentorial:Floor of 4th V. Supratentorial:Frontal> parietal>temporo-parietal.
CT	Well circumscribed. Coarsely calcified. Mild-moderate contrast enhancement. Peritumoral oedema extremely uncommon	Sharply marginated Intense contrast enhancement Calcification in radial/circular pattern 20%	Well demarcated Smooth/ Lobulated. Homogenously increase density. Dense homogenous enhancement. Small foci of Ca2+(common) N/B:Other masses can be noted within plexus i.e AVM Cavernous haemangioma Dermoid Epidermoid	Mixture of hypodensity, isodensity, calcification and occasionally haemorrhage. Cystic areas due to myxoid accumulation. Ca2+ 91% especially where the tumour has infiltrated the cortical grey matter (nodular/massive/ dotdash/curvilinear/stippled/ diffuse) Calvarial erosion	Isointense on plain CT. Frequently but not always, show contrast enhancement. Ca:50%. Small seeding into the ventricular ependyma, subarachnoid space and spinal canal do not show contrast enhancement until the tumour reaches a certain degree of enlargement.
MRI	Isointense relative to cortical grey matter on T1&T2WI with heterogenous areas due to calcifications, cystic spaces or vascular flow voids. Attach to septum pellucidum (characteristic)	Hypo-iso onT1 Iso-hyper on T2	Hypo- to- isointense to brain on all pulse sequences. Hypo-Ca/Vascular signal void. Intense enhancement. (Ideal method to demonstrate cerebellar/ brainsteminvasion.	Heterogenous on all pulse sequences. Hyperintensity from previous haemorrhage may be noted on T1WI.	Low on T1WI. High on T2WI. Mixed signal due to haemorrhage, calcification and blood vessel.
Angio	Supply from striate arteries of MCA, Anterior Choroidal and branches of ACA.	'mother in law' sunburst/spoke-wheel pattern. Supply : choroidal vessel	Dilatation of PICA/ Choroidal branches. Dense tumour stain in arterial phase.	The fine capillary meshwork is too small to be seen on angio.	
Prognosis after diagnosis and treatment		Cx: Local invasion of venous sinuses.	Papilloma is often totally resectable, while a carcinoma, because of invasion, is not. Long term survival of patient with choroid plexus papilloma.	<50% at 5 years	Wrap around the blood vessel and cranial nerve making total resection difficult. Tended to recur locally and disseminate with time.

oligodendroglioma, but central neurocytoma is purely neuronal in origin. Presence of neuronal tissue can be shown by electron microscopy or neuronal specific markers such as synaptophysin (8).

The differential diagnoses of an intraventricular tumour based on radiological findings are many (Table 1). However, central neurocytoma should be considered as the main differential diagnosis in a young adult with a calcified intraventricular mass attached to the septum pellucidum. The possible diagnosis of central neurocytoma based on radiological features should be noted to the pathologist since further staining will be required to arrive at the correct diagnosis.

The general management guidelines for central neurocytoma are not yet clearly defined. However from the summary of published literature, it is evident that, tumour debunking surgery should be the initial treatment modality. The completeness of surgery predicts post treatment outcome. Though central neurocytoma is a benign tumour, repeated recurrence and CSF spread of disease can occur. There are clear indications that local radiotherapy help to reduce local relapse rate.

Response to radiation is usually slow as it is a benign disease. The shrinkage of tumour following radiotherapy takes almost 6months to 2 years post treatment (9). Recently there is a report of use of combination chemotherapy in recurrent central neurocytoma (11). However it's efficacy has yet to be proven.

Correspondence :

Dr. Abdul Rahman Mohd Ariff MMed (Radiology),
Department of Radiology,
School of Medical Sciences,
Universiti Sains Malaysia,
16150 Kubang Kerian, Kelantan, Malaysia.

References

1. Figarella-Branger D, Pellissier JF, Daumas-Duport C, Delisle MB, Pasquier B, Parent M, Gambarelli D, Rougon G, Hassoun J. Central Neurocytoma: Critical evaluation of small-cell Neuronal Tumour. *Am J Surg Pathol* 1992; **16(2)**: 97-109

2. Hassoun J, Gambarelli D, Toga M. Central Neurocytoma. An electron microscopic study of two cases. *Acta Neuropathol* 1982; **56**:151-56

3. World Health Organisation (WHO) *Book for "Histological Typing of Tumours of the Central Nervous System,* 1993.

4. Dong Gyu Kim, Je G. Kim DG, Chi IG, Park SH, Lee SH, Jung HW, Kim HJ, Cho BK, Choi KS, Han DH. Intraventricular neurocytoma: clinicopathological analysis of seven cases. *J. Neurosurg* 1992; **76**:759-65.

5. Eng DY, De Monte F, Ginsberg L, Fuller GN, Jaeckle K. Craniospinal dissemination of central neurocytoma. Report of two cases. *Neurosurg* 1997; **86**:547-52.

6. Yasargil MG, Von Ammon K, Von Deimling A, Valvanis A, Wichmann W, Wiestler OD. Central neurocytoma. Histopathological variants and therapeutic approaches. *J. Neurosurg* 1992; **76**:32-37.

7. Kim DG, Kim JS Chi JG, Park SH, Jung HW, Choi KS, Han DH. Central neurocytoma: proliferative potential and biological behaviour. *J Neurosurg*. 1996; **84**:742-47.

8. Jay V, Edwards V, Hoving E, Rutka J, Becker L, Zielenska M, Teshima I. Central neurocytoma: Morphological, flow cytometric, polymerase chain reaction, fluorescence in situ hybridization and karytypic analyses. *J. Neurosurg* 1996;**90**:348-54.

9. Kim DH, Paek SH, Kim IH Chi JE, Jung HW, Han DH, Choi KS, Cho BK. Central neurocytoma: The role of radiation therapy and longterm outcome. *Cancer* 1997;**79**:1995-2002.

10. Weil MD, Wara WM. Pediatric tumour of the central nervous system. In Leihel SN and Philips TL eds. *Textbook, of radiotion oncology*. WB Saunders company. Philadelphia, 1998; 934-35.

11. Brandes AA, Amista P, Gardiman M Volphin D, Guylielmi B, Carolloc, Pinnag, Turazzis, Momfardini's. Chemotherapy in patients with recurrent and progressive neurocytoma. *Cancer* 2000; **88**:169-74