# Symtomatic Subependymoma Of The Lateral Ventricle: A Rare Entity –A Case report and review of literature

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**Citation**

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**Abstract**

Objective: Subependymomas of the lateral ventricle are very rare, benign, noninvasive and slowly growing tumors. The most common site being fourth ventricle followed by lateral, third ventricles, septum pellucidum and spinal cord. Many cases are asymptomatic and discovered at autopsy in elderly patients. Symptomatic cases often present in middle aged patients with hydrocephalus.This tumor was first described by Scheinker in 1945 as a separate entity. Settings: Grant Medical College & Sir J.J Group of Hospitals, byculla, Mumbai, Maharashtra, India. Methods: A 25 year old male presented with a history of sudden onset headache and giddiness. CT scan and MRI scans with contrast were suggestive of a heterogeneous mass lesion in the atria of lateral ventricle. Patient was operated upon with craniotomy and complete excision of lesion. Results: Patient had a stormy course post-operatively. He has been following up regularly with gradual recovery. Conclusion: Subependymal tumors are rare especially in the lateral ventricles. Complete surgical excision with follow up should be the goal to achieve cure.

## INTRODUCTION

Subependymomas are rare, noninvasive, benign tumors. Most commonly they arise in the fourth (50-60%) and lateral ventricles (30-40%). Rarely do they arise in the third ventricle, septum pelludicum and the spinal cord. They are classified histological, according to the World Health Organization (WHO) classification as Grade I tumors. Due to the widespread use of pre-operative imaging an increasing number of asymptomatic subependymomas has been detected. However due to their rarity and variable imaging characteristics, reliable preoperative diagnosis remains challenging.

## CASE REPORT

A 25 year old male was admitted to our hospital with a 5 day old history of headache and giddiness which was sudden in onset and gradually progressive in nature. Headache was associated with occasional visual obscurations with blurring of vision while getting up in the morning.

On neurological examination, higher mental functions were normal; Visual acuity was 6/9 in right eye and 6/36 in left eye and Fundus was normal. The rest of the cranial nerves, as well as motor and sensory examinations were normal. Cerebellar signs, Romberg's test and gait were normal.

X-ray skull was normal.

On C.T. scan ( Fig.1) with contrast there was ill defined heterogenous mass lesion in atrium of left lateral ventricle and differential diagnosis were given as Hemorrhage, Ependymoma and Astrocytoma.

MRI scans (Fig.2) were showing ill defined heterogenous lesion hypo intense on T1 weighted images, hyper intense on T2 weighted images with minimal contrast enhancement.

The patient was operated upon with left parieto-occipital craniotomy with endoscopic assisted transcortical excision of left ventricular lesion. Intra-operatively, lesion was present in the atrium of left lateral ventricle with thalamic extension. The lesion was soft, suckable, vascular, with areas of gray and black. Near total excision was achieved and brain was lax at the end of procedure.

Frozen report was given as low grade glioma.

The patient had a stormy post operative recovery with ventilation for 7 days and gradual weaning off. The patient had convulsions post-op which were controlled with anticonvulsants. Tracheostomy was done on post-operative day 4. The patient was gradually weaned off tracheostomy and was removed. 3 sittings of hyperbaric oxygen therapy were given. The patient was gradually weaned off Ryles tube feeds and started on oral feeds. At the time of discharge patient was conscious, dull, obeying commands and

accepting oral feeds.

Histopathological examination (Fig.3) was showing vaguely lobular pattern with clustering of tumor cells. Individual cells were monomorphic, round to oval nuclei with bland chromatin. Focal nuclear pleomorphism was noted. Mitoses were absent. Stroma is gliofibrillary with presence of microcysts. Hemorrhage and hemosiderin is noted suggestive of Subependymoma (WHO grade-1)

Review from other institute was also suggestive of Subependymoma.

Post-operative CT scans (Fig.4) were suggestive of near complete excision of lesion with intraventricular blood which was resolved on follow up scans.

Pt was advised regular follow up at discharge. In follow up, patient had gradually improved in sensorium with increased alertness and weight gain.

**Figure 1**

Fig.1- C.T. scan with contrast showing ill defined heterogeneous mass lesion in atrium of left lateral ventricle with minimal contrast enhancement.

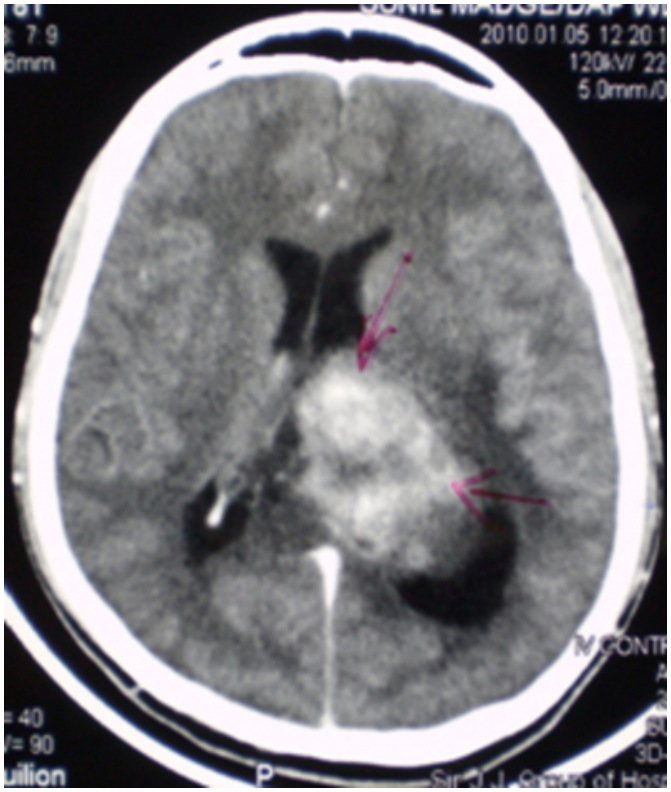


**Figure 2**

Fig.2- MRI scan with contrast showing ill defined heterogeneous lesion with minimal contrast enhancement.

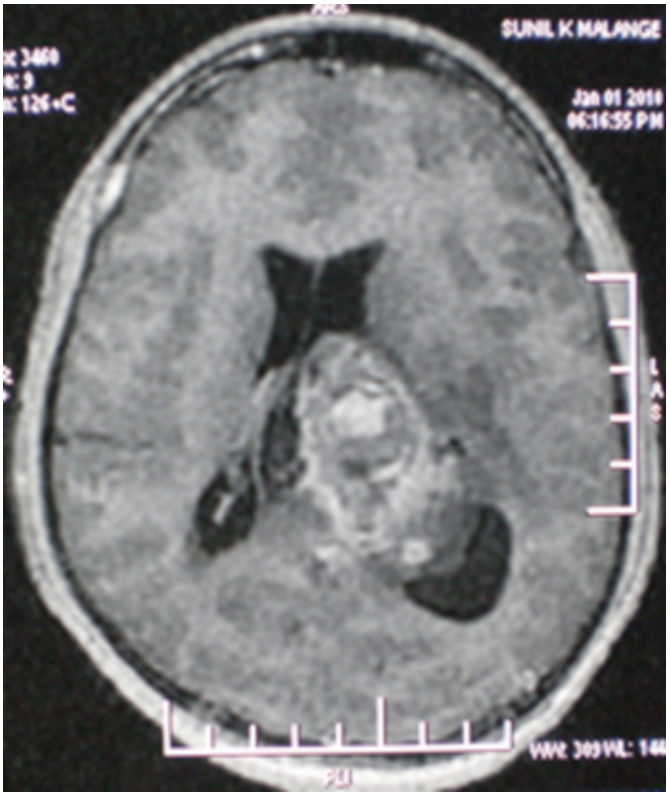


**Figure 3**

Fig. 3- Histopathological examination showing lobular pattern with clustering of tumor cells. Tumor cells were monomorphic, round to oval nuclei with bland chromatin.

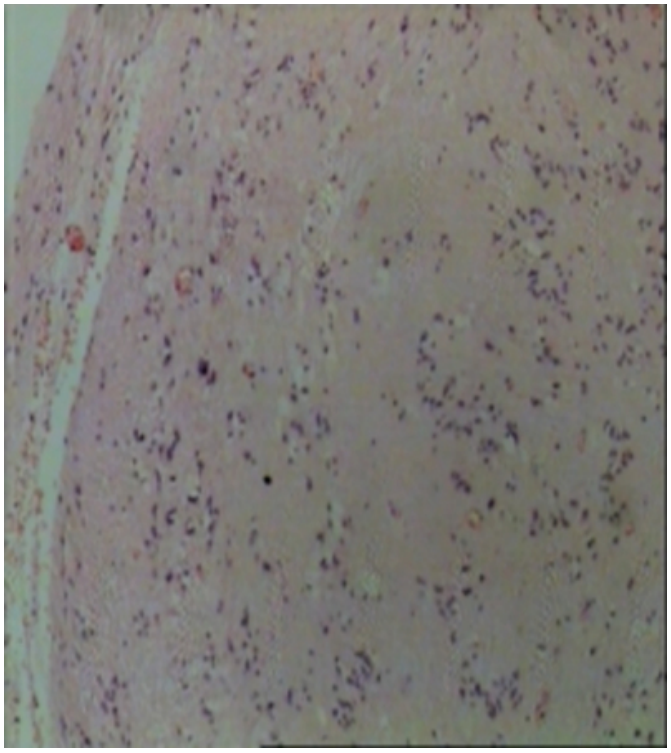
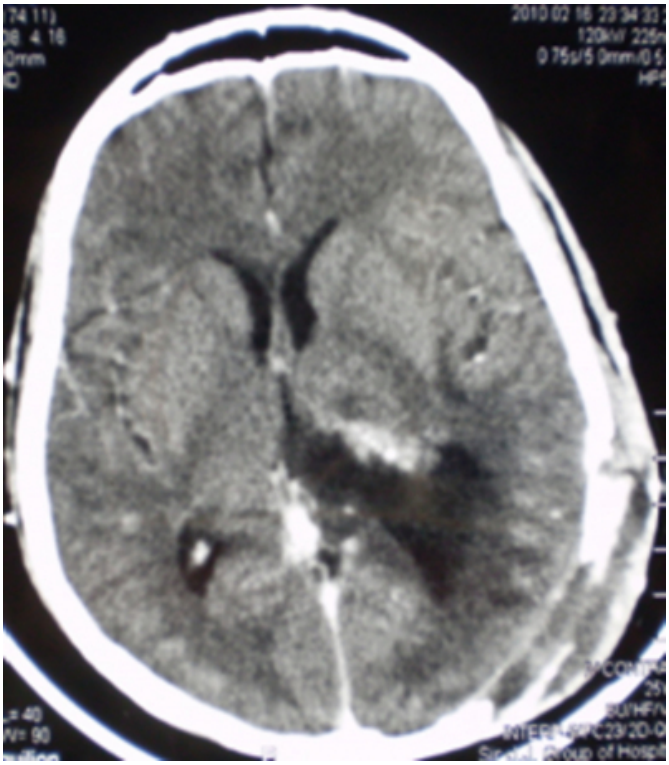


**Figure 4**

Fig.4- Post-operative CT scans showing near complete excision of lesion.



## DISCUSSION

Subependymoma is a rare, benign, usually asymptomatic, slowly growing glial tumor (WHO grade 1) with noninvasive charactestics. The incidence being 0.2-0.7% of all intracranial tumors [(1)].Ultrastructural studies indicate an origin of subependymal layer. Some authors suggest that this lesion could be more likely a hamartoma rather than a tumor because of a very low MIB-l index and lack of telomerase activity [(2)].

Symptomatic patients are usually men in the fourth and sixth decade of life and presents with features of raised intracranial pressure.

Symptomatic subependymomas are usually 3-5 cm or greater in size and are due to obstruction of CSF pathways leading to increased intracranial pressure.

The most common presenting symptoms include headache,gait ataxia,giddiness,vertigo,nausea and vomiting [(3)].

In our case, patient was a young male of 25 years and presented with headache, giddiness and occasional visual obscurations.

On CT Scans subependymomas appear as a well defined, lobulated intraventricular mass lesion that is hypodense to isodense to brain. These lesions are usually solid or solid with cystic component and shows minimal or no enhancement on contrast images[4]. Calcifications are absent in lateral ventricular ependymomas, however it can be seen in fourth ventricular lesions. Fourth ventricular lesions can also show heterogeneous enhancement on contrast images [5].

MRI Scans shows a well defined intraventricular solid or solid with a cystic component lesion which is hypo intense to isointense on T1 weighted images and hyper intense on T2 weighted images. On contrast MRI scans lesion shows minimal or no enhancement [4]. Fourth ventricular lesions can show heterogeneous enhancement on contrast enhanced T1-weighted

images [5] .

Subependymomas shows different CT and MR imaging features, depending on their anatomic location. Calcification and heterogeneous contrast enhancement are common features of fourth ventricular subependymomas.Lateral ventricular subependymomas showed a lack of calcification as well as minimal or no contrast enhancement of CT and MR images [5].

In our case also, lateral ventricular lesion had similar characteristics as described above. The neuroimaging features of subependymomas are nonspecific and cannot be used to establish the correct pre-operative diagnosis. The differential diagnosis includes central neurocytoma, choroids plexus papilloma, subependymal giant cell astrocytoma, metastasis and ependymomas.

Subependymomas are nodular tumors composed of remnants of ependymal cells in a dense glial fibrillary matrix along with hemosiderin deposits.

Calcifications and microcysts are frequently observed [3,6].These tumors are classified by WHO as Grade 1 Neoplasm. However, histological origin remains controversial. Subependymomas may actually represent a hamartoma than a neoplasm due to their bland microscopic appearance and low Ki-67 labeling index [4].

As in our case, histopathological examination was showing vaguely lobular pattern with clustering of tumor cells. Individual cells were monomorphic,round to oval nuclei with bland chromatin. Mitoses were absent. Stroma is gliofibrillary with presence of microcysts. Hemorrhage and hemosiderin were noted.

Complete surgical excision is the treatment of choice for symptomatic subependymomas to achieve cure [4]. As in our case complete excision was achieved.

The prognosis is very good after total resection of tumor, therefore post-operative radiotherapy is not recommended [7]. Post-operative radiation therapy is indicated for sub totally excised or recurrent lesions [1].

Ki-67 labeling index can be used as a predictive-proliferative marker which may indicate aggressive clinical course and recurrence [4].

## CONCLUSION

Subependymomas of lateral ventricles are rare, benign and very slow –growing tumors. Total surgical excision is the treatment of choice for symptomatic subependymomas to achieve cure. Post-operative radiation therapy is indicated in partially excised or recurrent lesions. Prognosis is very good after complete surgical excision. Tumor recurrence may be correlated with high Ki-67 index.

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