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## A Rare Case of Extra-Ventricular Supra-Sellar Neurocytoma – A Diagnostic Conundrum

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## **A Rare Case of Extra-Ventricular Supra-Sellar Neurocytoma – A Diagnostic Conundrum**

### **Background**

The brain and its associated structures, not unlike other parts of the body, are often the primary source or sites of metastases for many tumors. Commonly seen tumors here are Glioblastomas, Meningiomas, Craniopharyngiomas, Pituitary adenomas etc.

Neurocytomas are a relatively rare group of neuronal neoplasms, first described by Hassoun et al in 1982 as an intra-ventricular tumor, with similar microscopic features as that of oligodendroglioma [1]. These tumors are not known to be very aggressive and they are estimated to constitute only 0.25-0.5% of all central nervous system tumors.

Extra-Ventricular Neurocytoma (EVN) is an extremely rare variant and became a separate class of pathology affecting the CNS in 2007 and was described as a Grade II tumor by WHO [2]. It was identified by Ferreol et al in 1989 [3]. Grossly, the tumor is firm and smooth with extensive blood supply and fibrous tissue. Histologically, it is similar to central neurocytomas. EVN's are so rare, that by 2015, there were less than 100 cases described in literature [4]. These tumors may be seen in the cerebral hemispheres, brainstem, spinal cord etc.

We report a case of an extra-ventricular neurocytoma of the supra-sellar region, a rare tumor, that has been seen only 12 times before, therefore, making this, probably the 13<sup>th</sup> case ever reported in medical literature [5].

### **Objective**

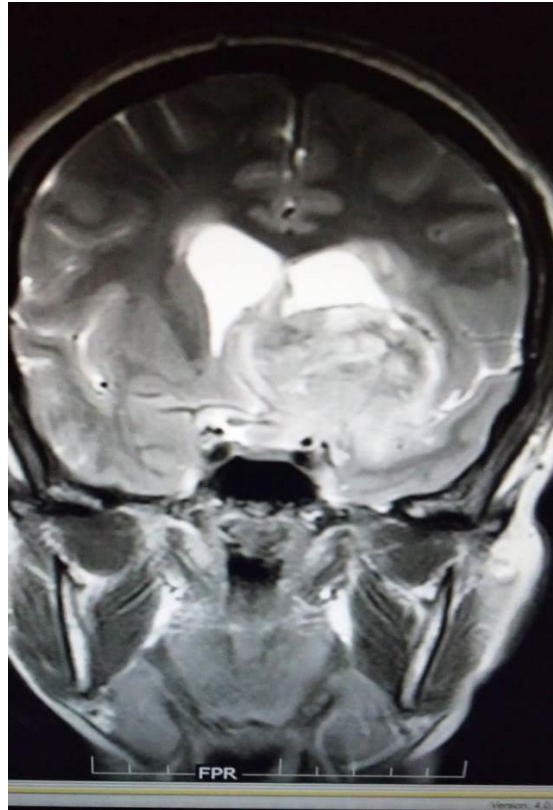
We describe a case of an extremely rare CNS tumor that is often misdiagnosed. We also comment on the diagnostic modalities and management.

### **Case report**

A 45-year-old woman presented with severe headache for two months and vomiting for 3 days with otherwise normal physical findings.

**MRI:** In the Supra-tentorial region, a well-defined, lobulated, solid-cystic, extra axial suprasellar mass measuring 4.1 x 4.5 x 5.2 cm was seen with foci of blooming on grant sequence. Marked peri-lesional flair and hyper-intense edema was noted. The same can be seen in figure 1. IV contrast showed heterogenous enhancement. Mass extended up to peri-mesencephalic and pre-pontine cistern posteriorly whilst abutting basilar and left Posterior Cerebral artery and extended inferiorly to the pituitary fossa and superiorly to the Corpus Callosum. Compression of foramen

of Munroe with right ventricular hydrocephalus was noted. MR spectroscopy showed glutamate peaks with reduced NAA (N-Acetyl Aspartate).



**Figure 1**

**(A well-defined, lobulated, solid-cystic, extra axial suprasellar mass in the supra-tentorial region)**

Course: A Pterional craniotomy and decompression surgery was performed and post-op patient was extubated. She was stable and moving all limbs. However, condition deteriorated by night and CT scan was ordered. Scan revealed residual tumor with edema and hemorrhagic bleed. Patient was re-intubated and ventilated. Worsening of status warranted re-exploration surgery. Glasgow Coma Scale remained at E<sub>1</sub>V<sub>1</sub>M<sub>1</sub>. Unfortunately, patient passed away on post-op day 3.

Histology: Section shows a tumor with monomorphic cells interspersed by thin-walled blood vessels. Individual cells have round to oval nuclei with coarse chromatin with fine neuropil matrix. Occasional cells with eccentric nuclei and moderately eosinophilic cytoplasm seen. 3-4 mitoses per 10 high power field seen. No necrosis noticed. Ki 67 index was 8%. Cells stained positive for Synaptophysin. Features confirmed to be **Extra Ventricular Neurocytoma, WHO grade II.**

## Discussion

Neurocytomas, which are typically central and seen commonly arising from the septum pellucidum and lateral ventricles, are a rare occurrence. These tumors have been seen in different ages, but the age of onset is usually between 40 and 60 years. Extra-ventricular neurocytomas are significantly rarer, with only about a hundred cases described so far. Asians are more commonly affected and the mean age of onset is 34 years, quite a bit lower, compared to central neurocytomas. EVNs are mostly seen in the frontal lobe and secondly in the parietal lobe. They have also been found in the cerebellum, brain stem, sella, thalamus, spinal cord etc., as well as some regions outside the nervous system like in the abdomen and pelvis.

Since the sellar region does not have neuronal cells, it is believed that the tumor cells of origin reached here as a result of abnormal migration during embryogenesis. The common symptoms include hypopsia, headache and vision defects. Endocrinal disturbances (particularly Pituitary) and involvement of cranial nerves, especially, CNIII and CNVI may also be seen.

Currently, diagnosis relies heavily on histo-pathological examination and scans. Special stains like GFAP, NeuN, SYN etc. have been used. Out of these, SYN has shown the highest sensitivity. Extensive scans and high clinical acumen are needed in order to differentiate EVN from other tumors of the region. Table 1 shows how easily these tumors are misdiagnosed.

For treatment, total resection is the common consensus. There are two approaches, craniotomy or endoscopic transsphenoidal approach. Total resection is only possible in 13% cases. Use of radio-therapy is controversial.

|              |    |      |                  |                   |    |     |     |                 |    |
|--------------|----|------|------------------|-------------------|----|-----|-----|-----------------|----|
| Peng P et al | 56 | Male | Visual impairing | Pituitary adenoma | No | Yes | Yes | Total resection | No |
|--------------|----|------|------------------|-------------------|----|-----|-----|-----------------|----|

|                                    |  |    |        |                    |                   |     |  |     |     |                   |     |
|------------------------------------|--|----|--------|--------------------|-------------------|-----|--|-----|-----|-------------------|-----|
| <b>Feature</b>                     |  |    |        |                    |                   |     |  |     |     |                   |     |
| <b>Age</b>                         |  | 64 | Female | Visual impairing   | Pituitary adenoma |     |  | Yes | Yes | Partial resection | No  |
| <b>Sex</b>                         |  | 48 | Male   | Visual impairing   | Pituitary adenoma | No  |  | Yes | Yes | Partial resection | Yes |
| <b>Symptoms</b>                    |  | 23 | Female | Visual impairing   | Pituitary adenoma | No  |  | Yes | Yes | Partial resection |     |
| <b>Initial diagnosis</b>           |  | 40 | Male   | Blurring of vision | Pituitary adenoma | Yes |  | Yes |     |                   |     |
| <b>Cystic component</b>            |  | 27 | Male   | Visual impairing   | Macro-adenoma     | No  |  | Yes | Yes | Total resection   | No  |
| <b>Cavernous sinus involvement</b> |  | 25 | Male   | Visual impairing   | Meningio-ma       | Yes |  | Yes | Yes | Partial resection | No  |
| <b>Synaptophysin Staining</b>      |  | 58 | Female | Visual impairing   | Pituitary adenoma | No  |  |     | Yes | Total resection   |     |
| <b>Treatment</b>                   |  | 46 | Female | Visual impairing   | Meningio-ma       | Yes |  | Yes |     | Partial resection | No  |
| <b>Recurrence</b>                  |  |    |        |                    |                   |     |  |     |     |                   |     |

TABLE 1 – Analysis of some previously confirmed cases of EVNSR [5]

## References

- Hassoun, J., Gambarelli, D., Grisoli, F. *et al.* Central neurocytoma. *Acta Neuropathol* **56**, 151–156 (1982). <https://doi.org/10.1007/BF00690587>
- Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, Scheithauer BW and Kleihues P. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol* 2007; 114: 97-109.

3. Ferreol E, Sawaya R, de Courten-Myers GM. Primary cerebral neuroblastoma (neurocytoma) in adults. *J Neurooncol* 1989;7:121–28
4. Kane AJ, Sughrue ME, Rutkowski MJ, Aranda D, Mills SA, Lehil M, Fang S and Parsa AT. Atypia predicting prognosis for intracranial extraventricular neurocytomas. *J Neurosurg* 2012; 116: 349-354.
5. Nery B, Bernardes Filho F, Costa RAF, et al. Neurocytoma mimicking macroadenoma. *Surg Neurol Int.* 2019;10:8. Published 2019 Jan 21. doi:10.4103/sni.sni\_387\_18