

# CENTRAL NEUROCYTOMAS OF UNCOMMON LOCATIONS

## Report of two cases

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**ABSTRACT** - We report two patients with central neurocytomas at an uncommon location in the brain. The first, a 58-year-old man presenting with signs and symptoms of increased intracranial pressure, had a tumor located at the pineal region. The second, a 21-year-old woman with tumor in the aqueductal region had worsening migraine-like headaches and diplopia. Both patients had obstructive hydrocephalus treated by neuroendoscopic third ventriculostomy and biopsy of the tumors. No additional treatment was done. We conclude that neurocytomas should be considered in the differential diagnosis of tumors located in the pineal and aqueductal regions.

**KEY WORDS:** central neurocytoma, aqueductal region, pineal region, third ventriculostomy.

### Localizações incomuns do neurocitoma central: relato de dois casos

**RESUMO** - Relatamos dois pacientes com neurocitoma central com localização incomum no sistema nervoso central. O primeiro, 58 anos, masculino, apresentava sinais e sintomas de hipertensão intracraniana, tinha um tumor na região da pineal. O segundo, feminino, 21 anos, tinha um tumor na região do aqueduto de Sylvius e apresentava cefaléia migranosa progressiva e diplopia. Ambos apresentavam hidrocefalia obstrutiva tratada com terceiroventriculostomia endoscópica e biópsia da lesão. Não foi feito tratamento adicional. Concluimos que os neurocitomas devem ser considerados no diagnóstico diferencial de tumores localizados na região da pineal e do aqueduto.

**PALAVRAS-CHAVE:** neurocitoma central, aqueduto de Sylvius, região pineal, ventriculostomia.

Central neurocytomas are low-grade and slow-growing primary brain tumors of neuronal origin that were firstly described by Hassoun et al. in 1982<sup>1</sup>. They comprise 0.25 to 0.5% of all primary brain tumors<sup>2</sup>. They develop predominantly in young adults and are most frequently located in the lateral ventricles near the foramen of Monro or in the septum pellucidum and into the third ventricle. Neurocytoma can also be found into the fourth ventricle<sup>3,4</sup> and spinal cord<sup>5</sup>. This tumor can spread through cerebrospinal fluid and disseminate along craniospinal structures<sup>6</sup> and organs outside central nervous system, such as the peritoneum by means of a ventriculoperitoneal shunt<sup>7</sup>. Neurocytoma localized outside the ventricular system is rarely found<sup>8</sup>. Central neurocytomas of the aqueductal and pineal regions are also rare<sup>9</sup>.

We report two patients with central neurocytomas at uncommon locations in the brain. In one patient the tumor was located in the aqueductal region and in the other the tumor laid in the pineal region.

### CASES

**Case 1** - A 58-year-old man presented with a one-year history of memory loss, gait disturbance and headache. Neurologic examination showed ataxic gait and papilledema. Computerized tomography images showed a hyperdense mass at the pineal region, with small points of calcification and moderate enhancement after contrast agent. Neither cysts nor invasion of the ventricular walls were seen. Lateral and third ventricles were dilated and showed periventricular edema, characterizing obstructive hydrocephalus. FLAIR and post-gadolinium T1-weighted MR images (Fig 1) revealed a mass with heterogeneous signal and moderate enhancement after paramagnetic agent, without

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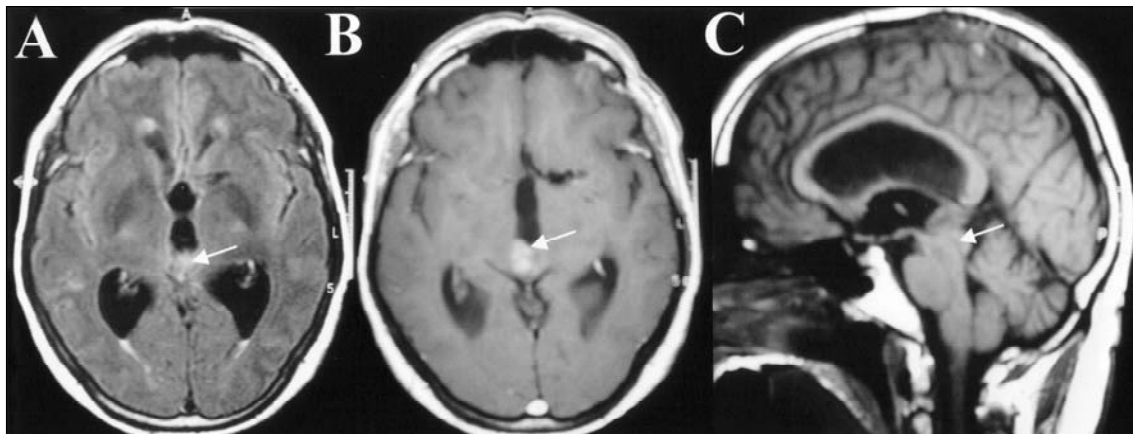
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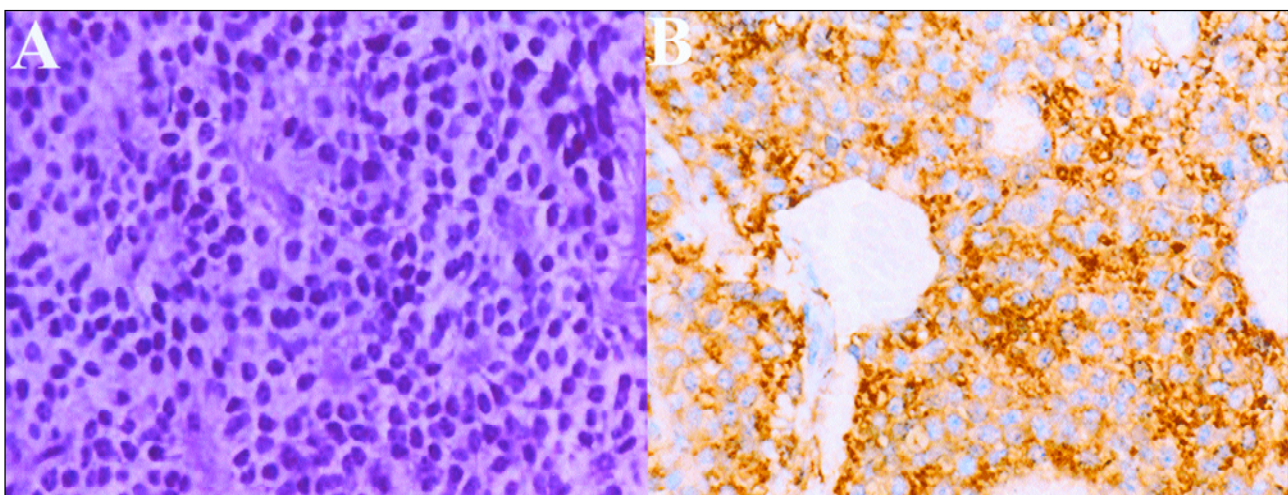
edema surrounding the tumor. Sagittal T1-weighted image showed total occlusion of the aqueduct of Sylvius by the mass. There was a bulging in the posterior wall of the third ventricle caused by the mass. The patient successfully underwent neuroendoscopic biopsy and third ventriculostomy. Light microscopy (Fig 2) disclosed a neoplasm composed of uniformly small, rounded cells. Mitotic figures, necrosis, and atypia were not observed. Immunostains were positive for synaptophysin, confirming the diagnosis of central neurocytoma. After surgery the patient showed total remission of the symptoms. He did not want to be re-operated or undergo radiosurgery. He is being regularly followed by the neurosurgical team and has no sign of tumor growth. An MRI study performed one year after the diagnosis showed no tumor growth.

**Case 2** – A 21-year-old woman with migraine-like headaches for a few months with recent increase in frequency

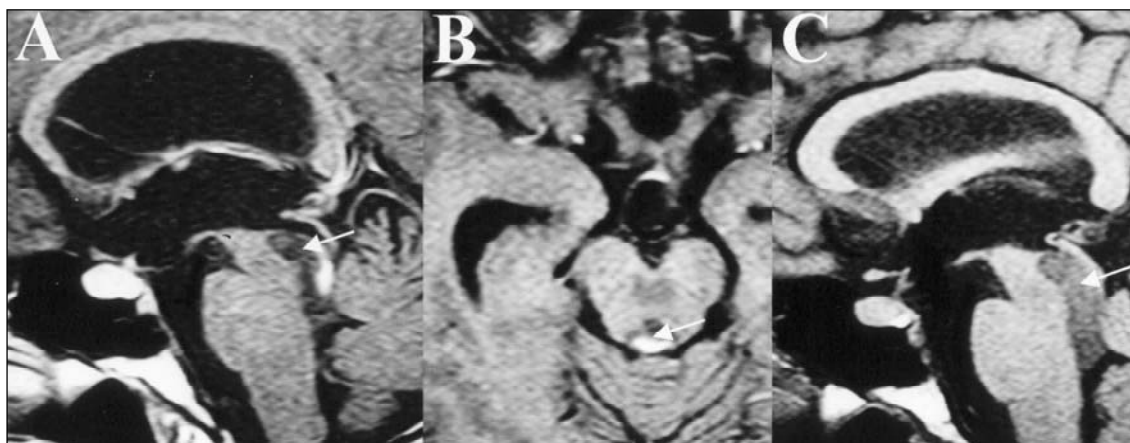
and severity, associated with diplopia. MRI images (Fig 3A and B) showed a midline mass in aqueductal region, obliterating the aqueduct of Sylvius and causing obstructive hydrocephalus. This mass showed hypointensity on T1-weighted images and hyperintensity on the T2-weighted images; heterogeneous enhancement after gadolinium was observed. She underwent neuroendoscopic biopsy of the lesion and third ventriculostomy during the same procedure. Tumor blocked direct access to the aqueduct. Hemostasis was done with irrigation and endocauterization. Hematoxylin and eosin stained sections of the tumor showed a vaguely lobular arrangement of uniform small cells separated by delicate capillary branches. Tumor cell nuclei were round, surrounded by perinuclear halos and contained single inconspicuous nucleoli. Mitoses were rare and necrosis was not detected. Synaptophysin staining was strong in cell cytoplasm. Histologic findings were diagnostic of central neurocytoma. The patient was discharged 2 days later and



*Fig 1. MRI images of the central neurocytoma. (A) Axial FLAIR magnetic resonance image showing no edema around tumor lesion. (B) Post-gadolinium magnetic resonance image showing the tumor mass at the pineal region (arrows). Tumor mass moderately enhances after gadolinium. (C) Sagittal view showing total occlusion of the aqueduct of Sylvius and signs of obstructive hydrocephalus.*



*Fig 2. Case 1 – Light microscopy view of the neurocytoma cells. (A) Hematoxylin and eosin staining showing microscopic appearance of neurocytoma. Cells are isomorphic and have scant cytoplasm. No signal of atypia can be seen. (B) Synaptic terminals of tumor cells stain positive for synaptophysin (brown).*



*Fig 3. Case 2 – MRI images of the central neurocytoma. (A-B) Magnetic resonance images obtained before surgery, showing tumor mass (arrows) in the aqueductal region leading to obstructive hydrocephalus. Enhancement after gadolinium is evident. (C) Magnetic resonance image obtained 6 months after surgery showing flow void signal at the region of the floor of the third ventricle (arrow) and no signs of hydrocephalus.*

immediately noted improvement of the symptoms. At 6 months follow-up, the patient objective neurological examination was normal. An MRI (Fig 3C) showed no signals of hydrocephalus and the presence of a flow-void in the floor of the third ventricle. The mass was similar. The patient remained neurologically intact.

## DISCUSSION

Central neurocytomas are more frequent from 20 to 40 years of age (about 70% of the cases). It is extremely rare in children. Signs and symptoms are caused by increased intracranial pressure due to cerebrospinal fluid block. The majority of central neurocytomas are benign. Approximately 25% of these rare central nervous system tumors are more aggressive, with an MIB-1 labeling index  $>2\%$  or atypical histologic features, and are classified as atypical neurocytomas. Macroscopically, neurocytomas appear as cystic, well circumscribed, and frequently calcified masses attached to the septum pellucidum and walls of the lateral ventricles, occluding the foramen of Monro. Tumor cells are stained for synaptophysin and neuron specific enolase<sup>10</sup>. On light microscopy they are similar to oligodendrogliomas and need to be distinguished from these glial tumors by synaptophysin reaction.

Around 60% of the central neurocytomas show genetic alterations, the most frequent being chromosomal gains (2p and 10q / 18q and 13q). MIB-1 reaction correlated with the degree of tumor recurrence (values higher than 2% indicate 63% of recurrence). At electronic microscopy, there are signals of neuronal differentiation, such as cytoplasmic processes, microtubules, dense granules, synaptic knobs, synaptic vesi-

cles and tight junctions. According to some authors, surgery followed by radiotherapy in case of incomplete resection is the most effective treatment<sup>11-14</sup>.

Based on image studies, the differential diagnosis of masses located in the ventricular system depends on their exact location and on the age of the patient. Oligodendrogliomas, subependymal giant cell astrocytoma, ependymoma, low-grade (pilocytic) astrocytomas and also neurocytomas comprise the most part of the tumors located in the lateral ventricle in young adults. The typical central neurocytoma is located in the supratentorial ventricular system (lateral ventricles in 77% and third ventricle in 21% of the cases). The majority of them are located in the anterior half of the lateral ventricle. Hassoun and colleagues showed this to be the case in 98 of the 127 cases studied<sup>2</sup>. Some series reported that the left lateral ventricle is more frequently involved<sup>2</sup>. Tumor extension into the third ventricle occurs in 26% of central neurocytomas. Isolated involvement of third or fourth ventricle and even extra-ventricular sites are rarely reported<sup>3,4,15,16,17</sup>. There are also rare cases of central neurocytomas with craniospinal and extra central nervous system dissemination<sup>6,7</sup>.

In this paper we show two patients with central neurocytoma at locations different from those most frequently reported in the brain. One man had a tumor in the pineal region and a woman with a tumor in the roof of the midbrain. Before surgery both patients had symptoms and signs of increased intracranial pressure. Obstructive hydrocephalus was seen in both patients by means of image studies. They both were treated the same way and had good outcomes

with total remission of the symptoms, although no additional therapy has been done up to now. Image studies one year (for the man) and 6 months (for the woman) after surgery showed no signal of tumor growth.

Central neurocytomas located at the pineal and aqueductal regions resemble tectal gliomas in, at least, three aspects. First, they both seem to be very slow growing tumors that can be managed conservatively<sup>18</sup>. Second, the clinical signs and symptoms are very similar in both types of tumors and are characterized by increased intracranial pressure due to obstructive hydrocephalus<sup>19</sup>. Third, in both tumors, endoscopic third ventriculostomy can result in excellent control of hydrocephalus<sup>20</sup>. However, longer follow-up in larger studies is required to determine if such an indolent behavior is kept after longer periods.

We conclude that central neurocytomas should be considered in the differential diagnoses of masses located at the pineal region and aqueductal region. Neuroendoscopic third ventriculostomy was effective in the treatment of obstructive hydrocephalus in these patients. There was no tumor growth although no additional treatment was done. If such tumor locations are related to a more indolent behavior than the intra-ventricular location is not known and needs further investigations.

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