Obstructive hydrocephalus from central neurocytoma

Clinically, patients with central neurocytoma may present with obstructive hydrocephalus, most frequently from blockage of the Foramen of Monro for typically located tumors [1) 2)].

Outcome

Central neurocytoma may have a favorable prognosis, with a lower incidence of cerebrospinal fluid shunt insertion throughout its course than that for other intraventricular tumors, if total removal is achieved [3)].

Case series

Twelve patients were included in a study, with a follow-up of 24 months. Data collected included: age, sex, clinical presentation, early morbidity and mortality, radiological findings (tumor location, features, residual, recurrence, and hydrocephalus). All patients underwent surgery for total or subtotal excision through a transcortical approach. External Ventricular Drain (EVD) was inserted then removed or replaced by a shunt. Histopathology and the MIB index were used to confirm the diagnosis and guide the follow-up; adjuvant radiotherapy or Gamma Knife radiosurgery was used for residual tumor or recurrence.

The ages of the patients ranged from 14 to 48 years. Two patients died early, after total and subtotal excision, from sepsis and thalamic infarction, respectively. Six patients (60 %) had a total excision; two of them had a high MIB index and showed small recurrence at 12 months and 18 months, respectively, and received Gamma Knife radiosurgery. One of the six patients with total excision needed a shunt, and no shunt was needed in the four other patients; a subtotal excision was done for four patients (40 %). An early shunt was inserted for two of these patients, radiosurgery-controlled for one patient, while radiotherapy was used for control in the other three patients; radiotherapy control failed in one patient, who underwent a second surgery at 18 months.

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Case reports

A 58-year-old male presented with progressive decline in cognition and gait. Subsequent Magnetic Resonance Imaging revealed obstructive hydrocephalus secondary to a posterior third ventricular lesion. An endoscopic biopsy and concurrent cerebrospinal fluid diversion by ETV was performed. Pathological analysis was consistent with a CN with positivity to Synaptophysin. MIB-1 proliferation index was 1%. There was good clinical recovery, and the patient underwent adjuvant stereotactic radiosurgery 1.5 months post-surgery.

Due to the rarity of CNs arising from the third ventricle, there are only three previous reports of these approached endoscopically. Such a technique allows for good visualisation of the lesion, and therapeutic ETV to relieve obstructive hydrocephalus. This case supports this approach as a valid, minimally invasive option. Additionally, this is the first case to report the MIB-1 proliferation index, contributing to future outcome evaluation of endoscopic approaches to typical (MIB-1 < = 2%) verses atypical (MIB-1 > 2%) CNs.

Endoscopic biopsy with concurrent ETV and adjuvant stereotactic radiosurgery is a valid treatment option for deep seated isolated small posterior third ventricular CNs

This case report represents the fourth reported successful usage of endoscopic approach for a small posterior third ventricular CN and CSF diversion by an ETV. Because the lesion extended down the left periaqueduct to the superior fourth ventricle, GTR was not achievable. The patient underwent adjuvant radiosurgery of the residual lesion as recommended in previous reviews of this rare tumor. Of the four reports, this is the only case to have a documented MIB-1 proliferation index, which will contribute to future evaluation of endoscopic approaches for typical versus atypical posterior third ventricular CNs. Consideration of use of a diode laser to aid endoscopic resection was felt as an important learning point from the review of previous cases, though this is operator dependent and requires technical training [5)].

Chandshah et al. reported a case of the intraventricular lesion with bleed which was operated on an emergency basis as it caused obstructive hydrocephalus and the patient was in altered sensorium [6)].

Javedan et al. were the first to introduce this technique in a 54-year-old patient with obstructive hydrocephalus from a posterior third ventricular CN. Endoscopic biopsy of the lesion was undertaken but adherence to the surrounding tissue and vessels prevented more extensive debulking. An ETV was also performed, and the patient underwent adjuvant stereotactic radiosurgery of the residual tumor two weeks later. There was good clinical recovery, and a 25-month follow-up MRI showed a minimal decrease of the lesion, a patent ETV, and resolution of the hydrocephalus. The authors did not mention the MIB-1 proliferation index [7)].

Park et al. describe a 79-year-old female with a multilobulated 1.8 cm posterior third ventricular CN also presenting with obstructive hydrocephalus. Similarly, an ETV was performed along with an endoscopic biopsy of the lesion. Adjuvant stereotactic radiosurgery was undertaken at one-week post-surgery, and a 3-month progress MRI show a decrease in the lesion size to 1.4 cm with the patient remaining well at 8 months. It was suggested that stereotactic radiosurgery could be a suitable treatment alternative to deeply seated CNs in older patients. Unfortunately, the MIB-1 proliferation index was also not reported [8)].

In a report by Romano et al. a 37-year-old female presented with obstructive hydrocephalus and left abducent nerve palsy secondary to a posterior third ventricular CN. With the aid of a diode laser and rongeur via an endoscopic technique, GTR of the lesion was achieved and an ETV was also performed.

The patient did well postoperatively with a resolution of the diplopia. There was no adjuvant radiotherapy or radiosurgery, and subsequent post-operative MRIs up to 36 months did not show any residual or regrowth of the CN. This was the only reported case where GTR was attained, with the use of a diode laser instrumental to achieving this safely. The MIB-1 proliferation index was not mentioned [9].

A side noteworthy paper to mention is by Gomes et al. were two cases of CNs located just adjacent to the third ventricle were similarly managed endoscopically. A 58-year-old male with a pineal region lesion and a 21-year-old female with an aqueductal region lesion both presented with symptoms of obstructive hydrocephalus and underwent endoscopic biopsy along with ETV. Post-operative resolution of the hydrocephalus and good clinical outcomes were achieved, and no adjuvant radiotherapy or radiosurgery was given. Neither case had the MIB-1 proliferation index reported. They emphasized that CNs in the pineal and aqueductal regions appear similar to tectal gliomas, and can be successfully approached endoscopically [10].

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