ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXIV | No. 4 December 2020

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DOI: 10.33962/roneuro-2020-085

Romanian Neurosurgery (2020) XXXIV (4): pp. 504-508
DOI: 10.33962/roneuro-2020-085
www.journals.lapub.co.uk/index.php/roneurosurgery

The two stages surgery in the management of central neurocytoma. Case series of 10 patients

W. Bennabi, A. Khelifa, Y. Felissi, L. Houari, A. Morsli

Department of Neurosurgery. El Oued University Hospital, Algiers, Algeria

Abstract

Central neurocytoma is an uncommon benign tumour of the central nervous system. The intraventricular location close to the Monro foramina and the attachment to septum pellucidum are characteristics for diagnosis. The encasement of vascular structures represents a surgical challenge. We report a series of 10 cases of central neurocytoma operated at our department 06 of those was operated in two stages intentionally in order to avoid post-operative complications and to obtain complete removal.

Keywords
central neurocytoma, ventricular tumour, immunohistochemical markers

Introduction

Central neurocytoma (CN) is an extremely rare benign tumour of central nervous system accounting for only 0.1 to 0.5% of brain tumours [1]; although described for the first time by Hassoun et al in 1982 [2]; since then, we dispose poor data about theses tumours in the literature [1]. The intraventricular location is the main site of development of these lesions [1, 3]. The brain MRI is an important tool for diagnosis; however, the certainty is obtained by histological examination; in which the differential diagnosis is made with oligodendroglioma and ependymoma. Surgery represents the only effective option to deal with this kind of lesions, with usually good results, associated with conventional radiation or radiosurgery in case of atypical finding.

Materials and methods

We report a retrospective study of 10 cases of central neurocytoma treated in our department over a period extending from 2003 to 2019. All patients underwent computerized tomography (CT) scans and magnetic resonance imaging (MRI) on T1, T2, FLAIR, and T1 injected sequences.

Results

The study includes 10 patients aged between 22-60 years old, with average age of 30.5 years. The sex ratio was 3F/2M.

Corresponding author:
Walid Bennabi

Department of Neurosurgery. El Oued University Hospital, Algiers, Algeria

walidneurosurgeon@gmail.com

ISSN online 2344-4959
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First published
December 2020 by
London Academic Publishing
www.lapub.co.uk

Clinical presentation

Increasing intracranial pressure (ICP) symptoms were the main clinical presentation observed in six patients, associated to visual disorder in seven patients; other symptoms were less frequent: seizures were found in two patients, cognitive disturbance in sthree patients, and decreased consciousness in one patient.

Imaging

The computerized tomography (CT scan) showed in the majority of cases heterogeneous lobulated lesion, located frequently in the frontal horn of the lateral ventricle adherent to the septum pellucidum and frequently invading this one, the lesion was associated in most cases with biventricular dilation. The magnetic resonance imaging (MRI) showed the characteristic aspect of this lesion: isointense on T1 weighted imaging, iso or hyperintense on T2 weighted imaging with heterogeneous enhancement due to the cystic and calcic components after gadolinium injection; showing a "soap bubble" characteristic aspect.

Treatment

Hydrocephalus management

Six patients consulted with hydrocephalus treated with different modalities: ventriculo-peritonealshunt (VP-shunt) was used in two patients; endoscopic third ventriculostomy (ETV) was performed in one patient; Rickham reservoir was placed in one patient (**Table 1**).

Table 1: Hydrocephalus managements

Method	Number of patients
VP-shunt	2
ETV	1
Rickham reservoir	2
Direct approach	1

VP-shunt: ventriculo-peritoneal shunt, ETV: endoscopic third ventriculostomy

Management of the lesion

All patients were operated with open approaches, the trans-frontal trans-ventricular approach was used in all patients combined to the trans-callosal approach in three patients. Six patients were operated intentionally in two stages. A complete resection was obtained in six (06) patients and subtotal resection in four (04) patients. The resection was limited in the remaining four (04) cases because of the infiltration of the trigone and encasement of the major venous vessels.

Table 2: Surgical approaches

Patients	Approach		Outcome and Complications
	1st stage	2nd stage	
Patient 1	TFTV	TFTV	Recidivism
Patient 2	TFTV	TFTV	Worsening of the cognitive disturbance
Patient 3	TFTV	None	None
Patient 4	TFTV	Transcallosal	None
Patient 5	TFTV	TFTV	None
Patient 6	TFTV	None	None
Patient 7	TFTV	Transcallosal	None
Patient 8	TFTV	Transcallosal	None
Patient 9	TFTV	None	None
Patient 10	TFTV	None	None

TFTV: trans-frontal trans-ventricular

Pathological findings

The diagnosis of CN was based on microscopic examination and several immunohistochemistry markers such as synaptophysin, GFAP, Olig2, NSE, chromatogramine, Ps100. Patients with Ki-67 antigen (Mib1) ≥8% was considered as atypical CN. This finding was observed in three patients.

Adjuvant therapy

Conventional radiotherapy was used for the three patients with atypical CN whereas surgery was preferred in recurrence with low grade lesion. We didn't experience stereotactic radiosurgery in our series as well as chemotherapy.

Mortality and morbidity

We noted that one patient presented a worsening of his cognitive disturbance (memory loss) while we didn't observe any death in our series. Although we reoperated one patient for recidivism three years later. (table2)

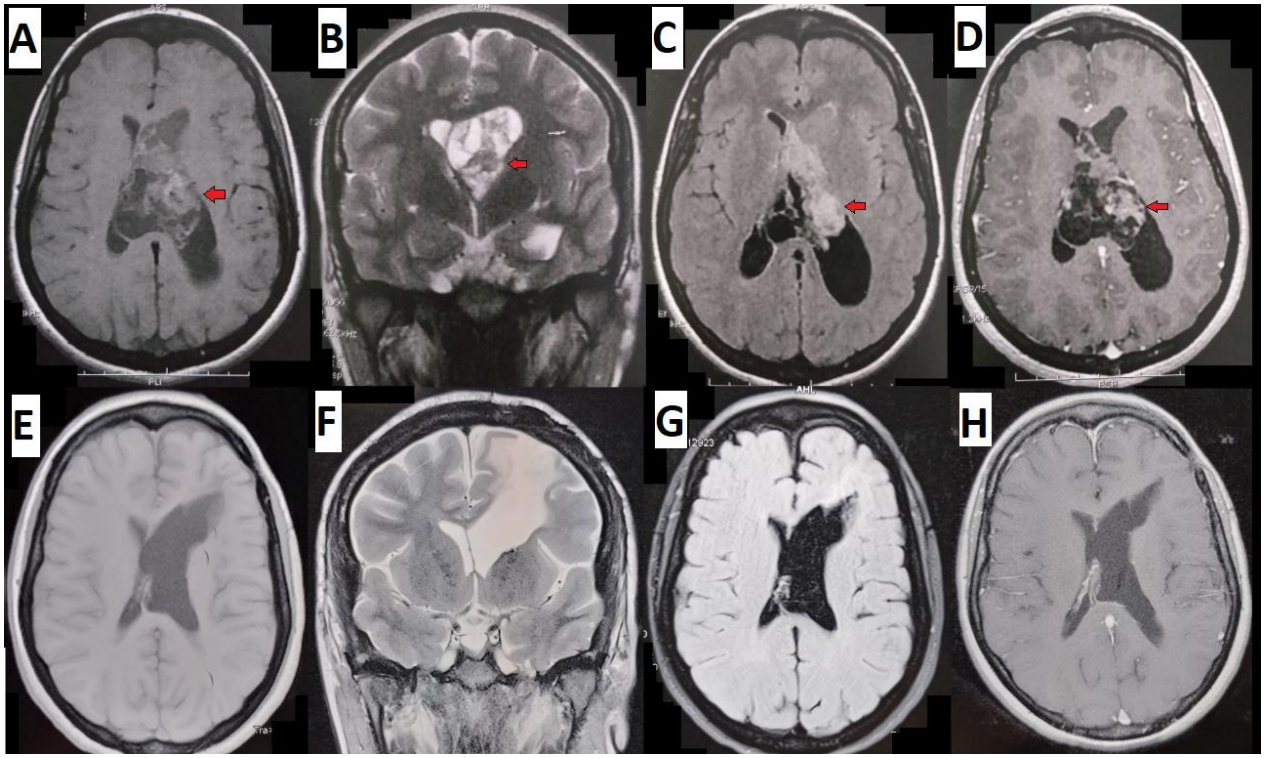

Figure 1. 30 years old female, with six months history of headache vertigo and visual disturbance with progressive worsening, at the admission the patient was conscious presenting visual acuity at 1/10 and 5/10, there was also a stage III papilledema in both eyes. Brain MRI objectified a 55 mm intraventricular process located on the left lateral horn, isointense on T1 WI, hyperintense on T2 and FLAIR sequences, with heterogeneous enhancement due to those images were defined a "soap bubble" aspect characteristic for neurocytoma (A, B, C, and D). The patient was operated and the lesion was totally removed through a left trans-frontal trans-ventricular approach. Pathological study confirmed the diagnosis of benign neurocytoma in post-operative there was a disappearance of headache and vertigo; the control imaging performed four years later showed no recidivism (E, F, G, and H). (A and E: T1 WI; B and F: T2 WI; C and G: FLAIR sequences; D and H: T1 injected images).

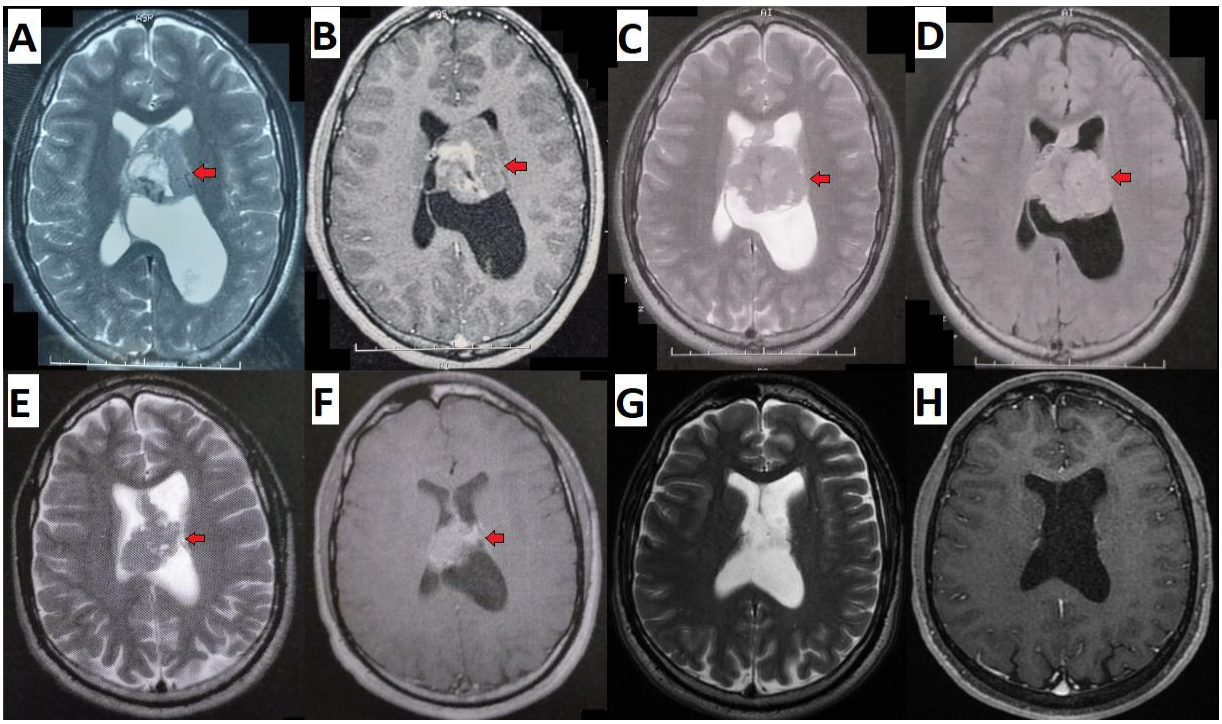

Figure 2. 22 years old male with one-month history of severe headache clinical exam at the admission found conscious patient without neurologic deficit there was no papilledema. Brain MRI objectified a process in the left ventricular frontal lateral horn measuring 33 X 45 mm with heterogeneous isosignal in T1WI, T2 WI, and FLAIR sequences with heterogeneous enhancement after gadolinium injection (A, B). The patient was operated and the lesion was subtotaly removed through a left Trans frontal Trans ventricular approach. One year later a control brain MRI objectified a re-expansion of the lesion it measured 50 X 50 X 42 mm (C, D), this time a total removal of the tumour was planned to be performed in two stages first a subtotal removed was performed through the same approach. Three years later the brain MRI objectified the persistence of the residual amount of the tumour measuring 23 X 34 mm (E, F) so the second stage of the removal was lunched and as planed the total removal obtained through the same approach. The brain MRI control images performed four later objectified no evidence of recurrence (G, H). A, C, E, and G axial T2 WI b; B, F, and H injected sequences; and C: FLAIR sequence).

Discussion

The central neurocytoma represents a very small proportion of brain tumours (0.1-0.5%), mostly in the lateral ventricle (77%),[2] with sometimes extension to the third ventricle (26%) [6] and rarely an exclusive localization in the third ventricle. As in our series it occurs frequently at the third decade with extremities ranging from (8 to 67) years old [7,8]. Most studies attest that there is no correlation between gender and incidence of central neurocytoma [5,9,10,11]; a slight female predominance was noted in our study, also higher incidences of this lesion was mentioned in some studies in Japan, Korea and India [8,12,13]. The clinical presentation is dominated by increasing intracranial pressure signs due to obstructive hydrocephalus by obliteration of the foramen of Monro associated to cognitive disturbance. Seizures, decreased consciousness and vision problems are less frequent. We consider that the cognitive disturbance is due to the close relation of the tumour to the trigone which is frequently infiltrated by the lesion (..).In radiological findings this lesion is mostly located in the anterior half of lateral ventricle appears to be isodense in computerized tomography (CT) scans associated to hyperdensities indicating calcifications which occur in up to 50% of all cases[5, 14].In magnetic resonance imaging (MRI) the central neurocytoma appears isointense on T1 weighted imaging, iso-hyperintense on T2 weighted imaging; and with heterogeneous moderate enhancements after contrast agent injection which is the classic "*soap bubble*" aspect [15]. Surgery is the only effective treatment for this kind of lesions and complete resection is associated with better rates of survival and local control, with five-year survival rate of 99% for gross total resection (GTR) and 86% for subtotal resection (STR) [19].

The trans-frontal trans- ventricular approach as well as the anterior trans-callosal approach offers the best surgical corridor to all lesions located in the frontal horn of the lateral ventricle [20]. We noted in patients operated in two stages that the resection was safer and easier comparing with those operated in single stage, this can be explained by the devascularization of the lesion in the first stage. Due to its histological similarities with other ventricular tumors such as ependymoma oroligodendroglioma the CN is frequently not easy to diagnose, the immunohistological markers are very useful in the diagnosis of this lesion and synaptophysin is the most specific one [13]. Adjuvant therapies are indicated in case of atypical, incomplete resection or inoperable recurrence neuocytoma; tow modalities are used conventional radiotherapy and stereotactic radiosurgery; with no statistically difference between both therapies. [16, 18]

Conclusion

The CN is usually a benign tumour of the central nervous system in which the surgical treatment is the only efficient option with very good results in term of survival rates and local control. Conventional radiotherapy or stereotactic radiosurgery can be considered in cases of atypical neurocytoma. The two stages surgery adopted in our department can be a good and safe strategy to deal with this kind of haemorrhagic tumours.

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