

Management of the Posterior Cerebral Fossa Ependymomas

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Abstract:

Background

Posterior fossa brain ependymomas are one of the most devastating forms of human illnesses which are more common in children. Brainstem compression, herniation and death are the risks with tumours in this critical location.

Patients and Methods:

A retrospective study including 50 patients with posterior fossa ependymoma were performed at the Neurosurgery Department in Ait IDDIR University Hospital between the period of January 2005 and December 2015. In each case, diagnosis was made clinically and confirmed radiologically and histo-pathologically. All patients received the adjuvant treatment

Results

Out of 50 patients, 30 (60%) patients were males and 20 (40%) were females. The mean age was 24 years (ranged 5 months–47 years); we identified 29 (58%) children and 21 (42%) adults, Ventriculo-peritoneal shunts were placed in all our patients, the total tumor excision was done to 35% and the partial tumor excision was done to 65%_patients.

The most common complications were as follows: Shunt malfunction : 4% , operative cavity hematoma 6%, CSF fistula 4%, deterioration of Cerebellar syndrome 6%, Cerebellar Mutism 2%, Mixed nerves palsy 4% and early post operative deaths 10%.

Tumor architecture was classified as classic (Grade II) in 35 (70%) cases and anaplastic (Grade III) in 13(26%) cases. Adjuvant treatment regimens following resection included radiation therapy only (72%) for 36 patients including children above and adults and chemotherapy only (36%) for 9 children below 4 years and 9 recurrent tumors. During follow-up period, recurrence occurred in 27% 11 patients out of 41patients .Five patients died (10%).

Conclusion

The surgical treatment of posterior fossa tumours still represents a challenge for neurosurgeons, Radical surgery with preservation of vital structures is the treatment of choice in patients with Posterior Cerebral Fossa ependymomas. Our experience shows the accepted results, complications and surgical outcome in relation to previous clinical studies.

Keywords: clippers; clinical evolution and management; primary tumors of the central nervous system; biopsy surgery for brainstem lesions; image-guided stereotactic biopsy

Introduction

Ependymoma represents the third most common childhood malignancy of the brain, accounting for approximately 10% of all pediatric brain malignancies. [1] the most common site of presentation is the posterior fossa. [1] Standard therapy involves maximal safe resection, followed by radiation therapy (RT). [2]

Patients and Methods:

Study design

A retrospective clinical cohort study of 50 patients with posterior fossa ependymoma were performed, and they underwent open surgical excision with ventriculo-peritoneal (V-P) shunt. All patients were admitted to the

Neurosurgery department in Ait IDDIR University Hospital Algiers between a period of January 2005 and December 2015.

For all the patients, the diagnosis was made after surgery. All patients received the adjuvant treatment.

Preoperative work-up

All patients underwent routine physical examination; they also received a thorough neurological evaluation. A special neuro-surgical sheet was applied in all cases included in the study to cover all suspects needed. CT as well as MRI, with and without contrast enhancement, was done for all cases before surgery as shown in (Figure 1)

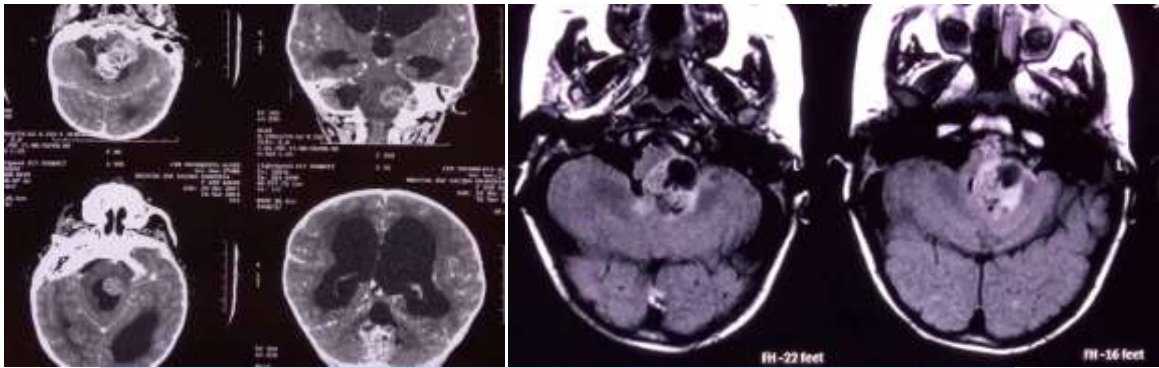


Figure 1: M.K aged 3 years, ependymoma grade II, pre-operative brain ct-scan and MRI 2012

Operative note

Surgery was performed with general anaesthesia, in prone position, using an operating microscope and microsurgical instruments in all studied cases.

In our study, all patients were operated via a midline suboccipital approach. The objective of this surgery was to complete microsurgical excision of the tumour unless that tumour was invading the brain stem or attaching important vessels or nervous tissue.

Postoperative follow-up

Early CT scan was performed to rule out early complications. Patients were followed clinically in our hospital then in outpatient neurosurgery clinic and also radiological through a period of 10 to 28 months. CT ± MRI with contrast was done at 3rd to 6th month of postoperative period then 1–2 years later. All intraoperative and postoperative complications were collected and reviewed, to detect any recurrence of the excised tumour (Figure 2-5).

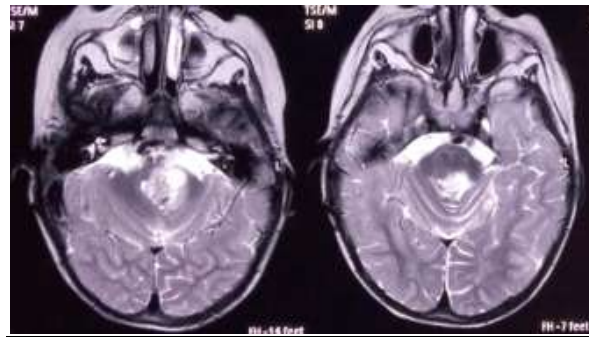


Figure 2: post-operative brain MRI 2015

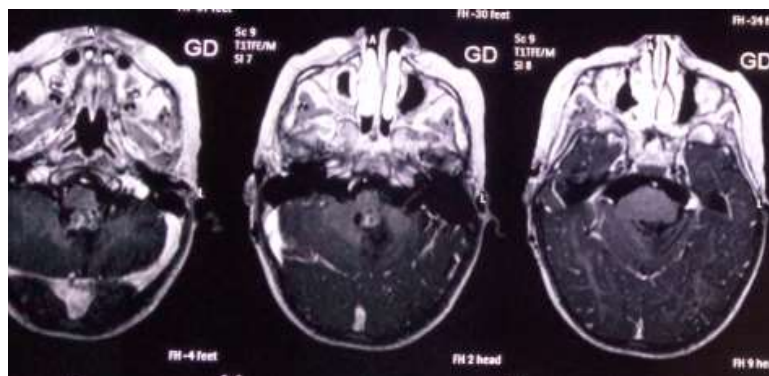


Figure 3: post-operative MRI 2016

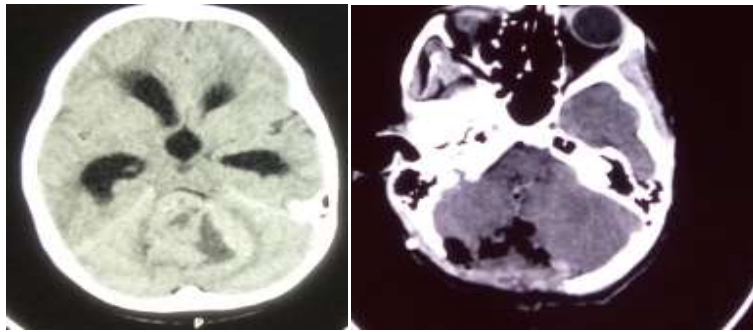


Figure 4: D.S 19 years, ependymoma grade III (pre and post operative ct-scan)

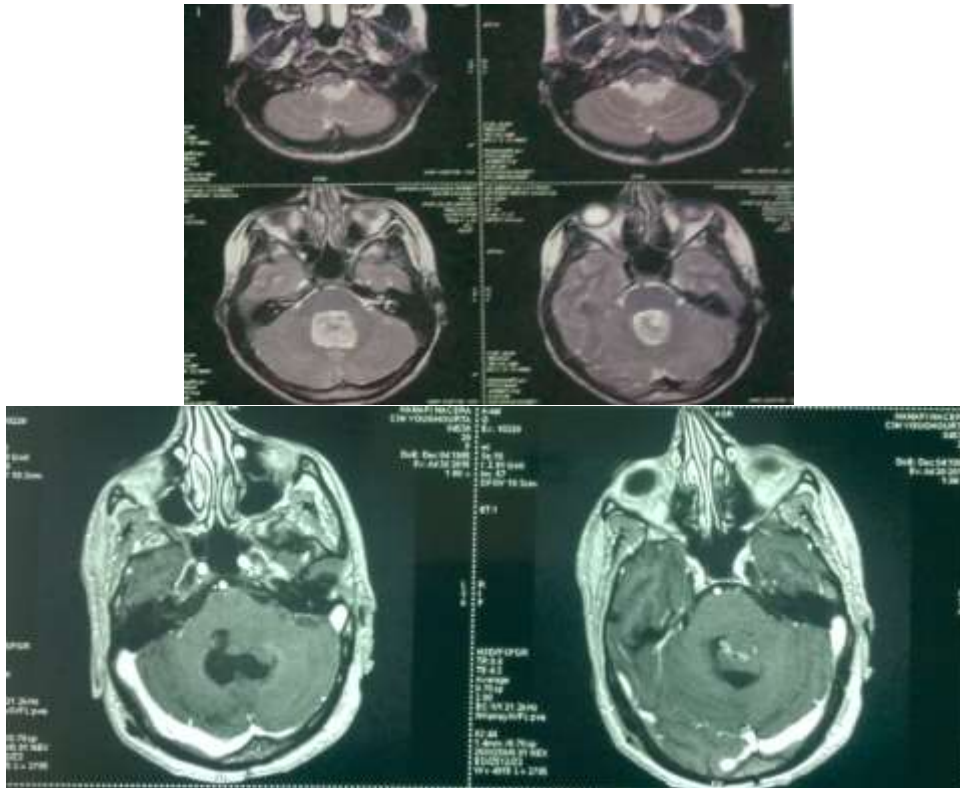


Figure 5: same patient, pre and post operative brain MRI

Results:

Between January 2005 and December 2015, 50 patients with posterior fossa ependymoma were operated, 30 (60%) patients were males and 20 (40%) were females. The mean age was 24 years (ranged 5 months –47 years). A male and pediatric predominance were observed in our series. Clinical manifestations of ependymoma vary widely according to the location. The most frequent clinical manifestations found were Intracranial hypertension syndrome (87%) and static and kinetic cerebellar syndrome (10%). The imagery had shown that 92% of these tumors originated from the floor of the 4th ventricle and only 8% were extended to the cerebello-

pontine angle cistern via foramen of Lushka and towards the cisterna magna via foramen of Magendie

For the Surgical outcome of this study: the Ventriculo-peritoneal shunts were placed in all our patients, the total tumor excision was done to 35% and the partial tumor excision was done to 65% patients.

In our study, the most common complications were as follows: Shunt malfunction : 4% (2 patients), operative cavity hematoma 3 cases (6%), CSF fistula 2 cases (4%), deterioration of Cerebellar syndrome 3 cases (6%), Cerebellar Mutism 1case (2%), Mixed nerves palsy2 cases (4%) and early post operative deaths 5 cases (10%) (Figure 6) .

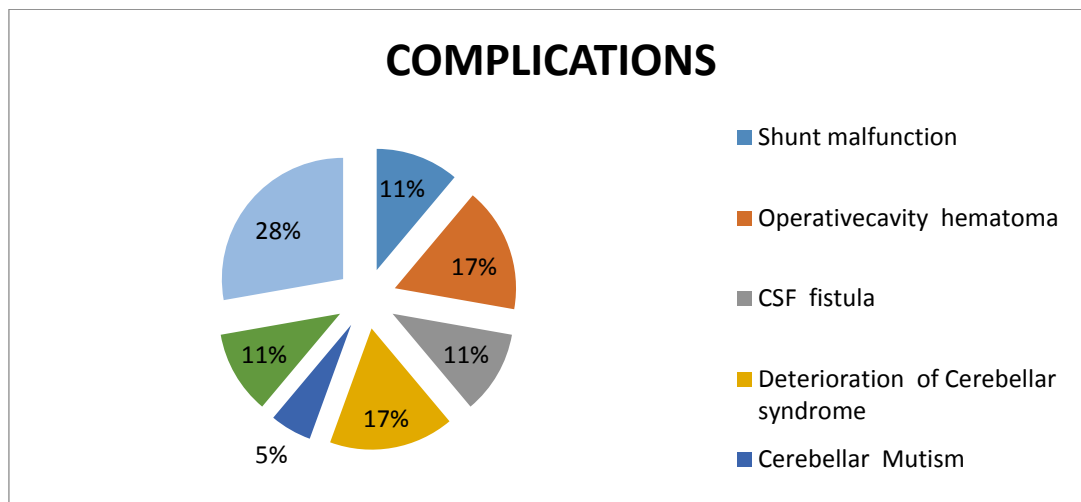


Figure 6: Post-operative complications

According to tumour pathology, two patients (4%) had Grade I, thirty-five (70%) patients had Grade II (non-anaplastic) ependymoma and thirteen (26%) had Grade III (anaplastic) pathology.

Adjuvant treatment regimens following resection included radiation therapy only (72%) for 36 patients including children above and adults and chemotherapy only (36%) for 9 children below 4 years and 9 recurrent tumors. During follow-up period, recurrence occurred in 27% 11 patients out of 41 patients. Five patients died (10%). The decision to re-operate must always be discussed before. All recurrent tumors were re-operated. Chemotherapy was often indicated after repeat surgery or in case of tumor inoperability.

According to the long term progress, the percentage of survival was as following:

at 5 years 29% (12 patients) and at 10 years 12% (7 patients).

Discussion:

According to an Australian study [1], the management of children operated of ependymoma of the posterior cerebral fossa is controversial. Radiotherapy should be delayed in children where tumor excision was total, on the contrary it is strongly indicated in those who underwent partial excision.

A European study [2] showed that the prognosis of PFC ependymomas depends on:

- 1) Surgical excision must be as complete as possible.
- 2) Histologic grade (according to WHO classification)
- 3) Tumoral volume
- 4) Indication for radiotherapy in partial excision or in case of recurrence

For the first time, WHO classification of CNS tumors has used molecular parameters and also histo-pathology to define the numerous tumoral entities [3]. This recent classification is based on the advances in molecular knowledge that guide therapeutic strategies to improve the prognosis.

According to Packer et al Children operated of Posterior Cerebral Fossa ependymomas, with total tumoral excision followed by chemotherapy and radiotherapy were completely cured. [4]

- The actual survival rate after posterior cerebral fossa ependymoma surgery is centered through close interaction of different factors: age, tumor location and histological grade of the tumor [5]
- Prognosis of the younger children is directly related to surgery and radiotherapy.

Conclusion:

Radical surgery with preservation of vital structures is the treatment of choice in patients with Posterior Cerebral Fossa ependymomas. It must be followed by chemotherapy and/or radiotherapy if it is of higher histologic grade or after subtotal excision.

Progress has been achieved in the management and prognosis of these tumors, and is attributed to the high knowledge of the clinical and biological behaviour of these lesions.

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