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The Primtif Spinal Cord Localization of Ependymoma about 2 Cases Treated by Surgery and Adjuvant Radiotherapy and Review of the Literature

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Abstract **Case Report**

Primary intramedullary ependymomas are rare tumors, characterized by a slow evolution. We report 2 cases of primary intramedullary ependymomas diagnosed following sensory-motor disorders of the 2 lower limbs, both patients underwent incomplete surgery followed by adjuvant radiotherapy and surveillance at the Mohammed VI University Hospital in Marrakech.

Keywords: Ependymoma, intra medullary ependymoma, surgery, adjuvant radiotherapy.

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INTRODUCTION

The ependymomas are rare tumors, these tumors arise from the cerebral ventricles or the ependymal canal but can also be located in the cerebral hemispheres.

Due to the different localization of ependymomas, the clinic differs according to the affected anatomical area.

The optimal management is multidisciplinary between surgeon-onco-radiotherapist.

PATIENTS AND OBSERVATION

We report 2 cases the 1st patient is a 52 years old man with a history of benign prostatic hypertrophy who presents a dorso-lumbar ependymoma.

The second patient is a 19 year old girl with no previous history of dorsolumbar ependymoma.

The 2 patients are followed at the oncology-radiotherapy department of the CHU Mohammed VI of Marrakech.

The 1st Patient: A 52 year old father of 6 children with a history of BPH who presented with chronic low back pain with sciatica complicated by sphincter disorders with sensory disorders of the two lower limbs and sadlle anaesthesia.

On MRI: Solid-cystic lesion process of the intramedullary of D12-L4 of 16*12*124mm evoking an ependymoma Figure 1.

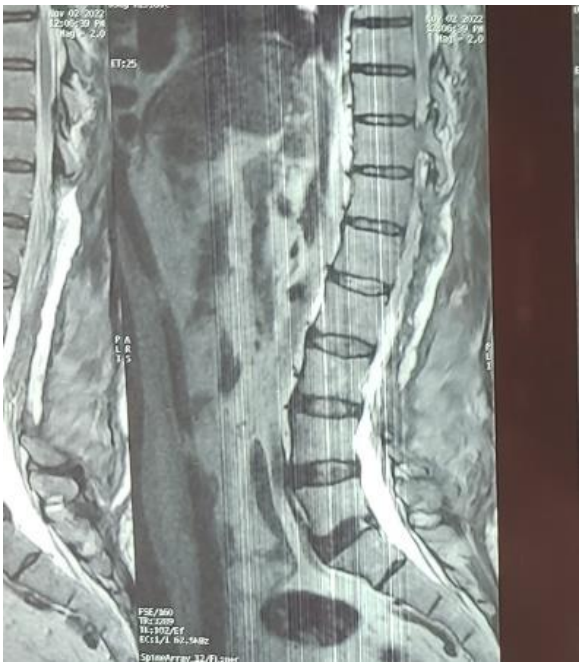


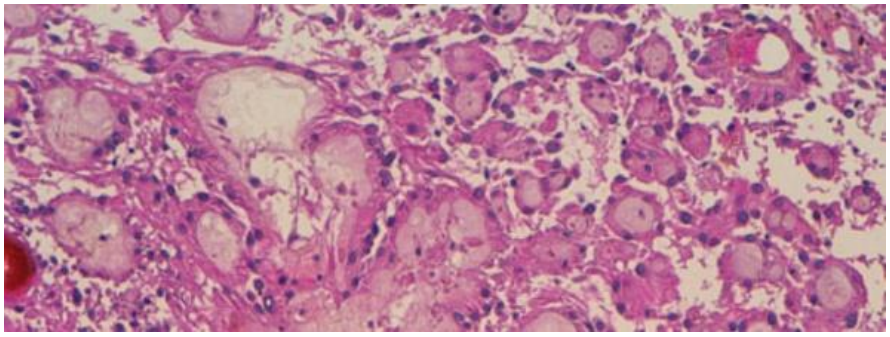
Figure 1: Spinal MRI sagittal section sequence T2 Showing solid-cystic lesion process of the intramedullary of D12-L4

Patient Operated: Incomplete surgery with morphological and immunohistochemistry appearance of a grade 2 ependymoma on anapath on neurological examination glasgow 15/15, gait examination deviated to the right, muscle strength 4/5 in both limbs, osteo-tendinous reflexes preserved, examination of the cranial pairs without abnormalities the patient presents with saddle anaesthesia and anal incontinence, the surgical wound is clean.

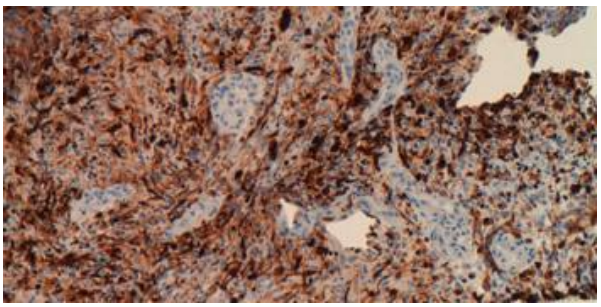
The 2nd patient is a 19 year old girl with no particular history who presented to the emergency room with a flaccid paraplegia unable to stand and walk. Abolished ROT of the 2 lower limbs with a sensory and motor deficit of the 2 lower limbs.

On MRI: Large intramedullary dorso lumbar expansive process D10-L2 of 138*20mm.

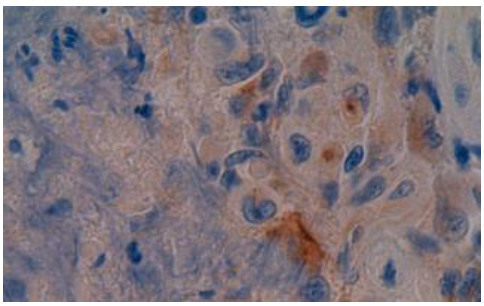
Patient operated on fragmented surgery and at the anapath it is a morphological aspect and IHC in favor of myxopapillary ependymoma, anti GFAP+, anti EMA + anti vimentine +, anti ps100+ anti ki 67<



Histological appearance of myxopapillary ependymoma



Anti EMA +



anti GFAP+

On current neurological examination glascow 15/15, gait examination normal, muscle strength 4/5 on 2 limbs, with no sensory disturbance, the patient holds the barré and mangazzini test, osteo-tendinous reflexes preserved, examination of cranial pairs without aomalies.

The 2 patients received an adjuvant radiotherapy between 45 in 25 to of 1.8gy and simple radiodermatitis type complications were observed with a good clinical-radiological response, reduction in size with disappearance of the initial symptoms after a 6 months follow-up.

DISCUSSION

Ependymomas occur preferentially around the age of fifty and are more frequent in men than in women [7, 8].

In 1954, Greenwood described a surgical technique for total removal of intramedullary ependymomas [1]. The tumors were successfully removed in 10 patients, 8 of whom were alive with no evidence of disease at 4 months to 21 years later [2].

As a result, many neurosurgeons have advocated radical excision as the standard treatment for spinal cord ependymomas. Certainly, complete en bloc resection is the treatment of choice if the lesion is small and well encapsulated [3].

In our case the, 2 patients had the tumor removed en bloc and none developed local recurrence. However, not all ependymomas can be removed en bloc. Cellular ependymomas often involve a long segment of spinal cord, and the upper and lower poles of the intramedullary ependymomas are usually indistinct [3, 4].

Sometimes no clear-cut tissue plane can be identified, especially with high grade infiltrative ependymomas. Furthermore, myxopapillary ependymomas often intermingle with the spinal nerve roots and are firmly attached to filum terminale [5, 6].

Consequently, it can be difficult to remove the tumor en bloc without sacrificing normal neurologic function. The term "complete" vs. "incomplete" or "total" vs. "subtotal" resection is commonly used in the literature [2, 3, 7, 8].

If the tumor has been removed en bloc, it is considered to have been completely resected. However, if the tumor has been removed in a piecemeal fashion, the determination of "complete" versus "incomplete" is often unclear. In general, the description of "complete" or "incomplete" is based on surgeon's subjective assessment. Shaw rt ul. reported that 3 of 8 patients thought to have had the tumor completely resected developed local recurrence [9].

This was presumably caused by microscopic residual disease in the tumor bed. Because of the ambiguity associated with the terms "complete" and "incomplete". Piecemeal resection seems to carry a significant risk of both residual disease in the surgical bed and drop metastases to the thecal sac.

Local recurrence in the irradiated volume represents the main form of therapeutic failure in primary intramedullary ependymomas. This mode of therapeutic failure is reported in several retrospective studies. Six of the 7 recurrences occurred in the irradiated volume in the series of Shaw *et al.*, [10] of 22 primary intramedullary ependymomas. Local recurrences in the irradiated volume represent the main form of therapeutic failure in primary intramedullary ependymomas. This mode of therapeutic failure is reported in several retrospective studies. Six of the 7 recurrences occurred in the irradiated volume in the series of Shaw *et al.*, of 22 primary intramedullary ependymomas [10].

For Waldron *et al.*, [12], 9 of the 11 recurrences were in the irradiated volume. Despite the apparent slow evolution of primary intramedullary ependymomas, the majority of recurrences occur within three years after treatment.

Waldron *et al.*, [13] reported a recurrence in 18.6% of cases (11/59), occurring in a mean time of 2 years (extremes of 1 month and 16 years). There was a correlation between these recurrences and the histological grade; the tumors were moderately differentiated or undifferentiated in 82% (extremes of 1 month and 16 years).

Radiation therapy provides long recurrence-free survival in primary intramedullary ependymomas after simple biopsy or subtotal excision [14-17].

The dose-response relationship has not been widely studied in the literature. In the majority of series, the number of patients was small and the dose was close to 45 Gy [16, 18].

For Waldron *et al.*, [19], a dose of 50 Gy in 25 fractions and 5 weeks is associated with good results without evidence of postradial myelitis.

O'Sullivan *et al.*, [20] did not find a significant dose-response relationship for intramedullary tumors; the radiation doses for the 31 intramedullary tumors studied in children ranged from 20 Gy to 56 Gy.

Dimarco *et al.*, [15], in a review of 252 intramedullary ependymomas, reported 4 intracranial relapses among 7 patients with primary anaplastic intramedullary ependymoma.

Taking into account these data and the potential toxicity of extensive irradiation of the cerebrospinal axis, we recommend localized irradiation of differentiated primary intramedullary ependymomas. The treatment of anaplastic forms should be individualized and irradiation of the cerebrospinal axis seems justified. The contribution of maximal surgery in primary intramedullary ependymoma is controversial; "conservative" surgery respecting neurological function and associated with radiotherapy of the tumor bed could ensure acceptable functional and carcinological results [14-17].

CONCLUSION

1. Radiation therapy is effective in controlling ependymomas of the spinal cord. A dose of 4500-5000 cGy appears to be adequate to achieve local tumor control.
2. Postoperative radiation therapy should be used if the tumor has been incompletely resected or if it has been removed in a piecemeal fashion.
3. If the tumor has been removed in a piecemeal fashion. thecal sac irradiation is necessary. 4. Myxopapillary ependymomas and high grade cellular ependymomas appear to be more likely to recur in the thecal sac. However, no big difference is detected in local recurrence.

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