

# Polyneuretic paraparesis revealing an intramedullary ependymoma

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## ABSTRACT

Spinal tumors are rare; intramedullary tumors are uncommon among these lesions, and occur in only 10% of cases in adults. Ependymoma is the most frequent histological type (40-60%). We report the case of a 22-year-old girl, presenting with local back pain, ascendant paresthesia, a progressive flask paraparesis, and a vesical globe. The clinical examination concluded in a mild motor weakness with areflexia of both patellar and Achilles tendons. The diagnosis of intramedullary tumor was made on MRI. The radiological and the macroscopic aspects evoked an ependymoma; the diagnosis was histologically confirmed after surgery (myxopapillary ependymoma) with a favorable evolution.

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Medullary tumors are uncommon lesions and occur less frequently in children. Intramedullary localization is seen in only 10% of cases in adults; ependymoma and astrocytoma are the most frequent histological types (approximately 70%).<sup>1-3</sup> The clinical presentation is aspecific, 3 syndromes could be more or less associated; they comprise local, radicular, and central signs.<sup>1,2</sup> Magnetic resonance imaging (MRI) has a major importance in the diagnostic approach of spinal cord tumors; it helps the characterization of the lesions and guides the therapeutic attitude.<sup>2</sup> Ependymomas are considered as benign tumors, but their slow evolution causes a delay in diagnosis and there is an important functional risk for the legs, essentially. The treatment is based on microsurgery.<sup>4</sup> This is an observation of a female patient presenting with a progressive flask paraparesis, and for whom the diagnosis of intramedullary tumor has been made on MRI.

**Case Report.** A 21-year-old girl, with the antecedent of a sciatic pain evolving for one year,

presented with a 2-month history of local back pain, tingling sensations initially noticed in the feet, and subsequently in the thighs. At this time, she also complained of numbness along the legs and difficulties in walking accompanied by a pelvic pain. These symptoms became progressively more intense and prompted her to seek medical care. Neurological examination revealed a mild motor weakness of the lower extremities (3/5 on muscular testing) associated with a hypoesthesia in the same distribution, areflexia of both patellar and Achilles tendons and a bilateral indifferent plantar reflex. Somatic examination found a pelvic mass. The biology was normal except a moderate inflammatory biologic syndrome (Sedimentation's speed: 35/65). Cerebrospinal fluid examination disclosed a yellow colored liquid; the proteins were high (2.72 g/l), the cytology examination revealed less than one white element per ml. The electromyogram revealed polyradicular denervation signs of the lower extremities. Somatosensory evoked potentials showed bilateral delayed latencies. An abdomino pelvic echography showed a

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**Figure 1** - A slightly hypo intense intramedullary oblong lesion extending from T12-L2 on T1 (a), hyper intense on T2 (b), enhancing after contrast injection (c). The tumor extends into the pedicles (d).

vesical globe necessitating the setting of a catheter. Regarding the association of central and peripheral signs, a MRI was carried out. A T1-weighted MRI showed enlargement of the spinal cord, caused by a slightly hypo intense intramedullary oblong lesion extending from T11 to L2 (**Figure 1a**); T2-weighted MRI showed a hyper intense intramedullary lesion with hemosiderin deposits (**Figure 1b**). The lesions had inhomogeneous contrast enhancement after Gadolinium's injection (**Figures 1c & 1d**). The patient was operated upon; it was a well-demarcated tumor with some hemorrhage, easily released and developing at the expense of the conus medullaris and the filum

terminale. Histological examination concluded in a myxopapillary ependymoma. The evolution was characterized by the regression of the local back pain and the tingling sensations, and by the persistence of the same motor deficit. The urinary disturbance evolved favorably, and the catheter was removed 9 days after the operation.

**Discussion.** Intramedullary tumors are uncommon lesions; they compose only 2-4% of all central nervous system tumors, 10% of spinal cord tumors and approximately 15% of intradural tumors in adults.<sup>1-3</sup> They are more frequent in children and occur

most often in the cervical spinal cord.<sup>3</sup> Ependymomas (50%) and astrocytomas (20%) are the most frequent histological types in the intramedullary localizations. Hemangioblastoma is the third most frequent intramedullary tumor.<sup>2</sup> Other histological types could exist: ependymoblastomas, mixed gliomas, subependymomas, schwannomas, and lipomas. Ependymoma is the most frequent intramedullary tumor. It occurs more frequently in the cervical spinal cord. An intramedullary mass developing in the conus medullaris and the filum terminale has more chance to be an ependymoma, especially a myxopapillary one. In fact, myxopapillary ependymomas occur almost exclusively in that site (95% of cases).<sup>2</sup> Ependymomas appear in the 4th or the 5th decades, with a median age of 35 years. There is no sexual predilection.<sup>1,2</sup>

The clinical presentation is aspecific including local back pain, sometimes an irradiating pain. This symptom is frequent and usually reveals the disease. It could be associated with paresthesia and sensory disturbances.<sup>1,2</sup> Motor weakness and sphincter dysfunction appear late in the evolution of the tumor, delaying the diagnosis, they are seen in only 25% of patients at presentation.<sup>2</sup> We present the case of a 21-year-old female patient presenting with a sub acute motor weakness of the legs associated with ascendant paresthesia. This presentation had initially evoked a Guillain Barré syndrome, but the association of peripheral and central signs (urinary disturbance) brought us to perform a MRI.

The MRI is of great interest in the imaging of intramedullary spinal cord tumors; it has greatly facilitated the diagnosis and the topographic classification of these lesions. It can give some predictive elements of the histological type, and thereby helps the therapeutic attitude.<sup>1,2,4</sup> In our case, the radiological aspect was a heterogeneous lesion hypo intense T1, hyper intense T2 and enhancing after Gadolinium injection at the site of development, and evoked an ependymoma.

The treatment of ependymoma is essentially surgical. Kyoshima et al<sup>5</sup> report that a complete microsurgical resection is sufficient to guarantee total tumor control with a very low risk of recurrence (approximately 5%), and that way, adjunctive radiotherapy seems to be unnecessary.<sup>4,5</sup> However, this opinion is not agreed by upon by all researchers. Kochbati et al,<sup>6</sup> emphasize the interest of post operative radiotherapy to assure a long term favorable prognosis.

In fact, they found no significant difference between patients treated by adjunctive radiotherapy after a simple biopsy or after a subtotal resection.<sup>6</sup> Recently, the interest of chemotherapy has been discussed in ependymoma.<sup>5</sup> Our patient was operated upon, and the macroscopic appearance of the tumor was a red sharply circumscribed lesion, hemorrhagic in places, with an interface between the tumor and the safe spinal cord. This aspect was in favor of an ependymoma.<sup>4,5</sup> A gross total resection was performed. Anatomopathological examination confirmed the diagnosis. Our patient had no radiotherapy after surgery.

Ependymoma have a good prognosis because of their limited infiltrative potential, and long-term tumor control can usually be achieved with gross total resection. Functional prognosis depends essentially on the degree of the neurological deficit before surgery.<sup>4</sup> In our case, the motor deficit persisted, but the urinary problems disappeared within several days after surgery.

In conclusion, the interest of this observation is to present an uncommon clinical presentation of intramedullary tumors. This diagnosis should be considered in paraparesis with signs of a central lesion, whatever the installation mode and the clinical presentation. Surgery alone could be sufficient when a complete resection is performed; adjunctive radiotherapy should be discussed in case of incomplete surgery or high grade ependymoma.

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