

PRIMARY FILUM TERMINALE EPENDYMOMA

A series of 16 cases

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Abstract – Filum terminale ependymomas are slow growing tumors of the cauda equina with a high incidence in young adults. Although a complete microsurgical resection can lead to a cure, recurrence is not uncommon. Sixteen cases of filum terminale ependymomas treated at the Instituto de Neurologia de Curitiba were analyzed. Eleven patients were females and 5 males, their age ranging from 7 to 84 years. Symptoms and signs included lumbar pain (31.25%), radicular pain (56.25%) and neurological deficits (12.5%). In three cases, patients had previously undergone surgery in other hospitals. All were tested through MRI and were operated on. Two underwent a laminoplasty and 14 a laminectomy. The last 8 patients of this series had neuro-physiological monitoring during surgery. In all patients a total microsurgical resection was achieved. Histologically, 2 cases were cellular ependymomas and 14 cases myxopapillary ependymomas. There was no recurrence during a 2 to 84 month follow-up period.

KEY WORDS: ependymoma, filum terminale, spinal tumor.

Ependimoma primário de *filum terminale*: análise de uma série de 16 casos

Resumo – Os ependimomas do *filum terminale* são tumores da cauda equina de crescimento lento com maior incidência em adultos jovens. A ressecção microcirúrgica total possibilita a cura da doença, recidivas, entretanto, apresentam sérias dificuldades no tratamento. Com o objetivo de estudar os aspectos clínicos, anatomopatológicos e do tratamento, analisaram-se 16 casos de ependimomas do *filum terminale* tratados no Instituto de Neurologia de Curitiba, 11 do sexo feminino e 5 do sexo masculino, com idade entre 7 e 84 anos, que apresentavam dor lombar (31,25%), radiculopatia (56,25%) e déficits neurológicos (12,5%). Em 3 casos, os pacientes tinham sido operados em outro serviço anteriormente. Em todos os casos o diagnóstico foi confirmado pela ressonância magnética. Em 2 pacientes realizou-se laminoplastia e em 14 laminectomia. Nos últimos 8 pacientes empregou-se monitorização neurofisiológica. Em todos os casos a ressecção microcirúrgica foi total. Do ponto de vista histológico, demonstraram-se 2 casos de ependimoma celular e 14 casos mixopapilares. Não houve recidiva do tumor em um seguimento entre 2 e 84 meses.

PALAVRAS-CHAVE: ependimoma, *filum terminale*, tumor raquimedular.

Tumors of the filum terminale are rare, constituting less than 6% of all spinal tumors¹. Myxopapillary ependymomas are a distinct variant of ependymomas and were described by Kernohan in 1932². They are slow-growing gliomas that occur in young adults, located in the conus medullaris and the filum terminale. Myxopapillary ependymomas are classified as Grade I, according to the World Health Organization classification³ of central nervous system (CNS) tumors.

The clinical manifestations frequently observed are lumbago and radicular pain, which can delay diagnosis be-

cause of its low specificity. The results of surgery on cauda equina tumors depend not only on histologic findings but also on the size of the tumor, as large lesions are generally more difficult to remove without causing further neurological damage⁴. Early recognition is, therefore, fundamental for optimal management. An MRI is essential in the diagnosis and the indication of treatment.

Aiming to study the clinical characteristics, diagnosis, surgical treatment, and histology findings, 16 cases of filum terminale ependymomas surgically treated at the In-

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stituto de Neurologia de Curitiba (INC) between 1999 and 2006 were retrospectively analyzed.

METHOD

Between 1999 and 2006, 16 patients with filum terminale ependymomas were surgically treated at the Instituto de Neurologia de Curitiba. These cases were studied after having received approval from the Ethics Committee at the INC.

Of the 16 cases, 11 patients were female and 5 were male, with the age at presentation ranging between 7 and 84 years (average 41.4 years). The clinical picture that leads to the diagnosis was lumbar pain in 5 cases (31.25%), radicular pain in the lower limbs in 9 cases (56.25%), and neurological deficits in 2 cases (12.5%).

All patients were submitted to an MRI for evaluation. The lesions were found in the filum terminale in all cases, from L1 to S2 (Table).

RESULTS

All patients underwent microsurgical treatment (Figs 1, 2, 3 and 4). The surgical approach was done through a laminectomy and laminoplasty in two cases and a laminectomy in 14 cases. Starting in May 2005, all 8 patients underwent neuro-physiological monitoring. The microsurgical resection was considered radical in all patients (Fig 5). In three cases, patients had previously had operations in other hospitals and presented with recurrent or persistent tumors. There were no post-operative neurological deficits;

however, two cases were re-operated for cerebrospinal fluid (CSF) fistula with success. There was no recurrence of tumors in this series in the pursuing 2 to 84 months.

In 14 patients, the histological diagnosis was myxopapillary ependymoma, while two cases were defined as cellular ependymomas.

DISCUSSION

Ependymal tumors are lesions with a moderate cell density, originating from the neuro-epithelium that covers the cerebral ventricles and the central spinal cord canal. The new classification by the World Health Organization⁵ for tumors of the nervous system characterizes ependymal tumors as follows: 1 – ependymomas, with four subtypes – cellular, papillary, clear cells and tanycytic; 2 – anaplastic ependymoma; 3 – myxopapillary ependymoma; 4 – subependymoma.

The myxopapillary ependymoma² arises from a well-defined site within the neuro-axis and has a characteristic histological appearance. It occurs most often in the region of the cauda equina, originating from the filum terminale, although there is one report of a myxopapillary ependymoma located in the brain⁶. As for its microscopic pattern, it is described as a proliferation of columnar or cubical cells, organized in a papillary arrangement around a vascular core against a mucoid matrix. Scattered mitoses can be

Table. Data from 16 patients with filum terminale ependymoma.

Case	Sex	Age (years)	Level	Date of surgery	Pathology findings	Complications	Clinical presentation	Intra-operative monitoring
1	M	39	L2–L3	09 Nov 99	Myxopapillary	–	Lumbago	No
2	F	54	L2–L3	22 Feb 99	Myxopapillary	–	Radicular pain	No
3	F	53	L2–L3	08 Jan 01	Cellular	–	Lumbago	No
4	F	46	L3–L4	05 Jun 02	Myxopapillary	–	Radicular pain	No
5	M	40	L2–L3	16 Jul 03	Myxopapillary	CSF fistula	Radicular pain	No
6	F	45	L1–L2	05 Apr 03	Myxopapillary	–	Radicular pain	No
7	F	24	L2–L3	29 Jan 04	Myxopapillary	–	Lumbago	No
8	F	37	L4–S1	18 Jul 04	Myxopapillary	–	Lumbago	No
9	F	84	L1–L2	31 May 05	Myxopapillary	–	Radicular pain	Yes
10	F	51	L2–L3	15 Aug 05	Myxopapillary	–	Radicular pain	Yes
11	M	40	L3–L4	06 Oct 05	Cellular	–	Lumbago	Yes
12	F	60	L2–L3	13 Dec 05	Myxopapillary	–	Radicular pain	Yes
13	F	54	L1–L2	20 May 06	Myxopapillary	CSF fistula	Radicular pain	Yes
14	F	07	L1–L2 S1–S2	28 Jan 07	Myxopapillary	–	Motor deficit	Yes
15	M	08	L1–L2 L5–S1	01 Jun 07	Myxopapillary	–	Radicular pain	Yes
16	M	20	L3–L4	21 Jun 07	Myxopapillary	–	Motor deficit	Yes

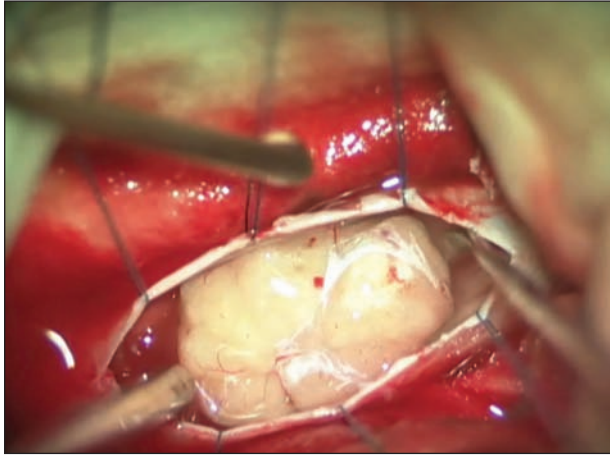


Fig 1. Surgical operating microscope view of the filum terminale ependymoma after opening of the dura mater.

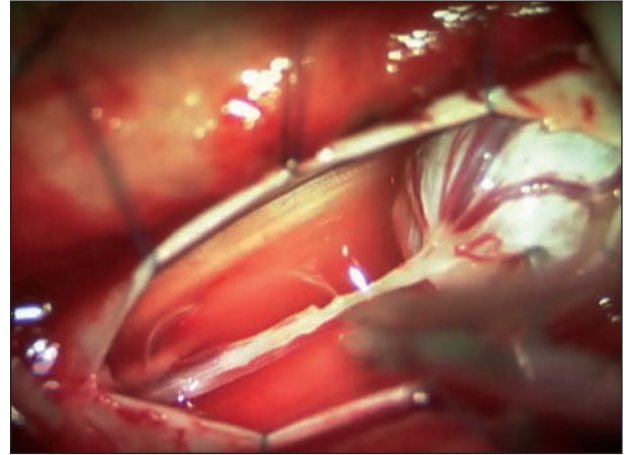


Fig 2. Magnified view depicting the filum terminale next to the tumor.

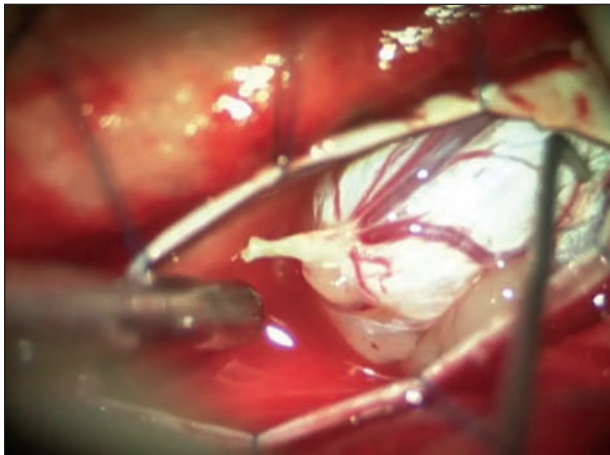


Fig 3. Image of the filum terminale being sectioned.

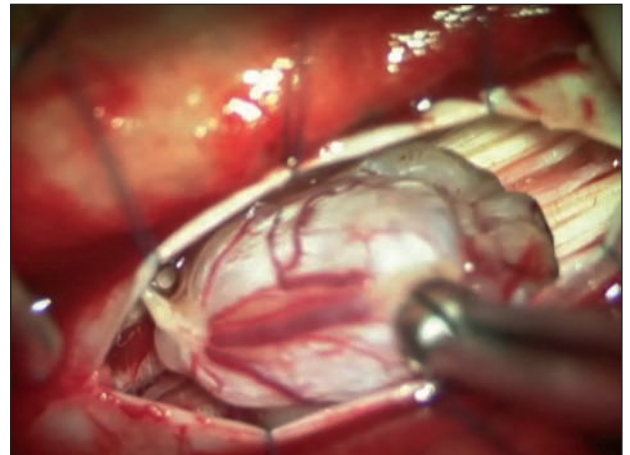


Fig 4. Image of the removal of the dissected tumor.

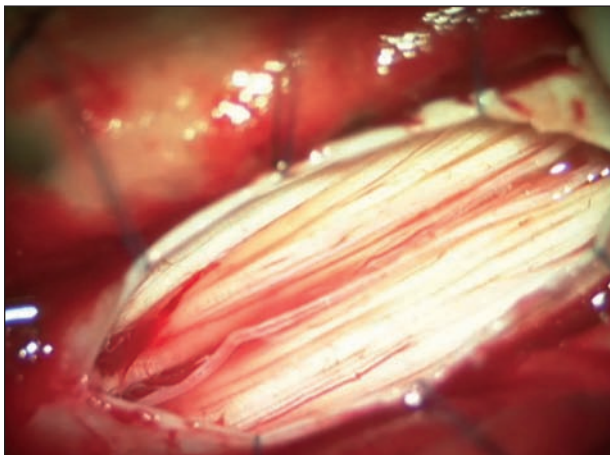


Fig 5. Image of the aspects of the cauda equina after the resection of the tumor.

observed, with the proliferative index being generally low (there is no apparent relationship with recurrence rate). The glial origin of the neoplastic cells can be confirmed by glial fibrillary acidic protein (GFAP) staining.

The differential diagnoses include papillary tumors, cordomas, mixed chordomas and mesotheliomas.

From a clinical point of view, lumbago and radicular pain in the lower limbs, were the main symptoms that lead to further investigation in our series. MRI, without a doubt, is the method of choice for the evaluation of these lesions⁷ (Figs 6A and 6B). In more than 50% of the cases of the present series, the most common symptom was radicular pain. Lumbar tumors cannot be adequately diagnosed with computerized myelography or myelography. In the MRI, these tumors almost always enhance after contrast media administration. Certain cases present with associated lipomas of the filum terminale, where T1-weighted images demonstrate the hyper-intensive characteristic of fat^{8,9}.

Microsurgery is the treatment of choice for filum terminale ependymomas, however, in some cases adjuvant treatments are employed.

The best adjuvant treatment of ependymomas has been the cause of intense discussion in literature. Until the

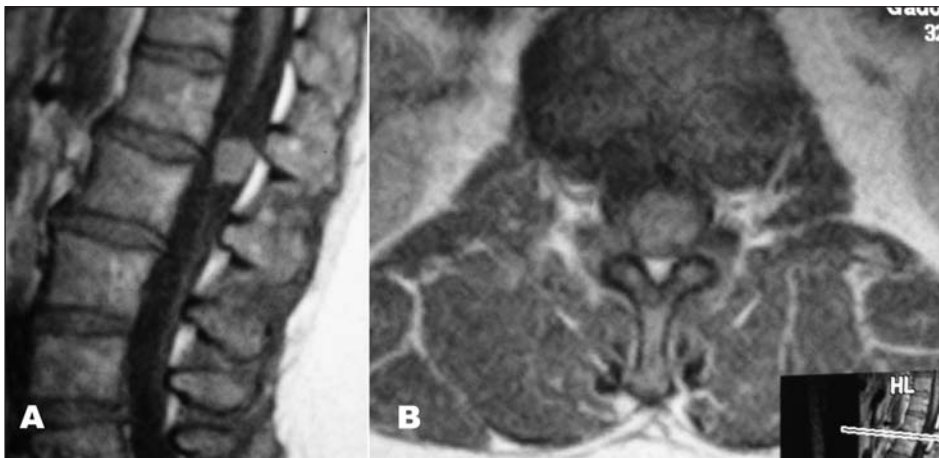


Fig 6. T1 weighted MRI of the lumbosacral spine demonstrating the tumor nodule with homogeneous enhancement of contrast in the region of the filum terminale ((A) sagittal; (B) axial).

mid of 1990s, there was no consensus on the indication of radiotherapy and even less of chemotherapy¹⁰. Only now, after the completion of long follow-up studies, could certain conclusions be drawn. Radiotherapy should be reserved only for those cases in which the tumor has not been totally resected. In small and medium sized tumors when a tumor capsule is kept intact, the lesion is less likely to disseminate through the neuro-axis¹¹. In large tumors, whether through manipulation or a partial resection, radiotherapy has been indicated as the treatment of choice¹².

Recent articles show that radiotherapy reduces the progress of the illness, improving the overall survival rates^{10,11,13}. Initially, a dose of 50 to 60 Gy is used on the tumor bed. Radiotherapy in the neuro-axis was reserved for cases in which a diagnosis of dissemination already existed prior to the surgery or as rescue therapy after conventional radiotherapy in cases of late spreading¹¹.

Chemotherapy is also a matter of discussion. However, with the selection of specific patients within defined parameters, the earlier diagnosis of CNS spreading and higher local control rates of the disease, the optimization of the treatment has been made possible. The drug of choice is etoposide at 50 mg/m², which has a good level of tolerance and is easy to administer. Nonetheless, several schemes have been tested without any one demonstrating superiority^{14,15}. The current indications for chemotherapy are the neuro-axis dissemination and as rescue therapy for recurrence. An increase in life expectancy and the quality of life has been reported¹⁴⁻¹⁶.

Although initially developed for the prevention of cervical deformities in children¹⁷, the use of laminotomy with replacement of the posterior elements of the vertebra removed during surgery can be useful in many cases. The theoretical advantages described by some authors^{18,19} are: restoration of bone protection in the spinal canal, better esthetic results, a lesser risk of the formation of epidural fibroses²⁰, which can be related to pain, and support for

the function of the intra-spinal musculature. Adequate fusion of the spine seems to occur with a greater probability in the thoracolumbar region¹⁸. Although there is no Class I evidence that a laminotomy prevents the occurrence of a deformity, we believe that the thoracolumbar region may theoretically benefit from this technique due to its transitional characteristics and higher risk for late onset deformities. The situations in which we prefer laminotomy to laminectomy are: children and young adults; when more than two levels of laminectomy are necessary and when there is an existing degeneration of the affected segments.

The objective of the microsurgery in filum terminale ependymomas is a total resection. The intra-operative neuro-physiological monitoring is a valuable tool that optimizes the procedure. Depending on their size, the ependymomas can involve both the nerves of the cauda equina and the conus medullaris. Further, the tumor causes local inflammation and arachnoiditis of the nerves and medullar tissue, making it imperative to dissect tumor from these structures with maximum care. As such, two types of monitoring can be employed: the direct stimulation of the roots and the monitoring of somato-sensory evoked potentials (SSEP's) of the lower limbs. The protocol of nerve stimulation used in our unit involves bipolar stimulation, varying from 0.2 to 2mA in intensity, a frequency of 60 Hz, the duration of each pulse being 1.5ms, delivered through the MSC-02B stimulator (Micromar, Brazil). This protocol avoids the spreading of electrical activity through the CSF, reducing the unwanted stimulation of remote roots and nerves.

The involvement of the conus medullaris is a good indication for the SSEP's. The anterior tibial nerve area is stimulated and the evoked potential is registered from the somato-sensitive cortex area (Cz' according to the international 10-20 assembly system) or preferably at the epidural space of the thoracic (D8-D10) region. The monitor used is the MK-15 (Amplaid, Italy), with latency refer-

ence of 20 ms and 38-40 ms for the lower thoracic region and contra-lateral cerebral cortex respectively.

The radical excision of the tumor through microsurgery techniques is what makes the cure possible for this disease and this should be the goal of the initial surgery, since a second approach can be hazardous and less likely to result in a total resection.

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