

## Original Article

# Extent of resection and postoperative functional declination of Klekamp's type A intramedullary tumors in adult patients

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## Abstract

**Background:** The most commonly primary intramedullary spinal cord tumors (ISCT) in adults are the noninfiltrative lesions, corresponding to Klekamp's type A classification. There are few reports exclusively considering this type of lesions, their resectability and postoperative functional declination risk, and to our knowledge, none from Latin America. This led us to evaluate our results to provide information that might contribute to the decision making process in our region.

**Methods:** A retrospective observational study was conducted comprising a cohort of 21 adults having primary Klekamp's type A ISCT. Diagnosis was made by magnetic resonance imaging (MRI), along with diffusion tensor/tractography in the last 7 cases. Preoperative functional status was assessed using the McCormick's modified scale (mMs), which was also used for the postoperative assessment within postoperative 90 days period. MRI was used to confirm the extent of resection.

**Results:** Radical resection was obtained in 20/21 cases. The postoperative functional status was stable in 42.8% of the cases, and in 57.4% was even better than in the preoperative period. Temporary declination was observed in 2 cases in the early postoperative period. There were 2 cases with complications; one patient had cerebrospinal fluid fistula with meningitis, which was conservatively resolved, and another patient died from pulmonary embolism.

**Conclusion:** Although the number of patients in this series does not allow to conclude from a statistical point of view, the outcomes showed that the modern surgery of Klekamp's type A ISCT permits a complete resection with low functional declination risk.

**Key Words:** Intramedullary tumor, spinal cord tumor, surgical results

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## INTRODUCTION

The incidence of primary spinal cord tumors both extra and intramedullary is 74 in 100,000 people per year. Among them, only purely intramedullary spinal cord tumors (ISCT) amount to 30%.<sup>[3,13]</sup> This low rate together with the fact that they are more frequent in children than in adults is the reason why there is less information regarding the latter age group.<sup>[4-6,10]</sup>

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ISCTs in adults usually appear during the productive stages of life, they manifest deceptively, and although their growth is often slow, their progress is unvaryingly constant, generally leading to serious and incapacitating neurological deficits unless they receive treatment.<sup>[3]</sup>

According to Klekamp's classification, there are three types of ISCTs, namely, type A, noninfiltrative; type B, infiltrative; and type C. Taking into account the literature, approximately 70% correspond to type A and are represented by the ependymoma as the most common tumor, followed by a heterogeneous group of other lesions such as malignant neuroepithelial tumors, cavernomas, hemangioblastomas, dermoid, and epidermoid cysts.<sup>[9,16]</sup> In most cases, the expectation of cure is usually present when the resection is radical, without requiring later oncological treatment. Published literature, mostly in the pediatrics field, show that the better the preoperative status is, the higher the expectation of neurological preservation becomes.<sup>[1,2,5,6,8-12,14]</sup>

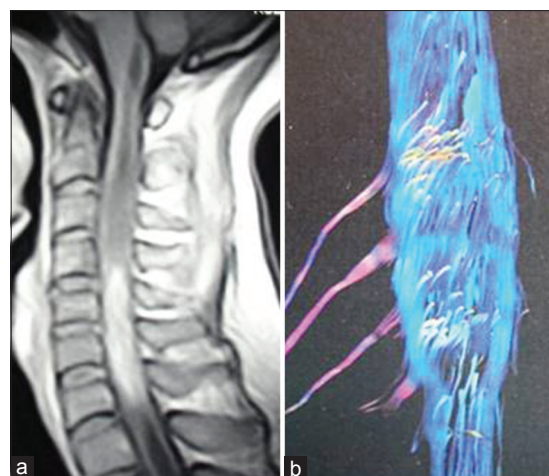
There are few reports considering exclusively the Klekamp's type A ISCT, their resectability and postoperative functional declination risk, and to our knowledge, none from Latin America. This led us to evaluate our results to provide information that might contribute to the decision-making process in our region.

## MATERIALS AND METHODS

A retrospective study was conducted comprising a cohort of 21 adult patients having lesions corresponding to type A in Klekamp's classification, who were treated from 1998 to 2015 by the senior author (ATR). The lesions were diagnosed through gadolinium magnetic resonance imaging (Gd MRI) and through MR diffusion tensor/tractography in the last 7 cases [Figure 1]. The preoperative functional status was assessed using McCormick's modified scale (mMs), which was also used for the postoperative assessment within postoperative 90 days period. The extent of resection was defined through Gd MRI.

The surgical approach was by laminectomy and laminoplasty on the last 7 patients. Spinal cord surgery was assisted by monitoring evoked potentials. Identification of the posterior raphe to perform the myelotomy was carried out using the middle point of the virtual line that extends between the areas of input of the right and left dorsal roots in addition to the visualization of the incoming vessels to the posterior raphe. Once the lesion had been exposed, a frozen biopsy was performed, which was later complemented by routine techniques of histopathology, immunohistochemistry, and/or molecular biology.

In the postoperative period, patients received prophylaxis for deep venous thrombosis (DVT) through pneumatic



**Figure 1: Illustrative case (n = 19). (a) Example of magnetic resonance (MR) of intramedullary ependymoma C4–C7 with associated syringomyelia; (b) interrelationship with MR tractography**

compression on the lower limbs, elastic stockings, and/or low molecular weight heparin. Early physical therapy was also prescribed.

## RESULTS

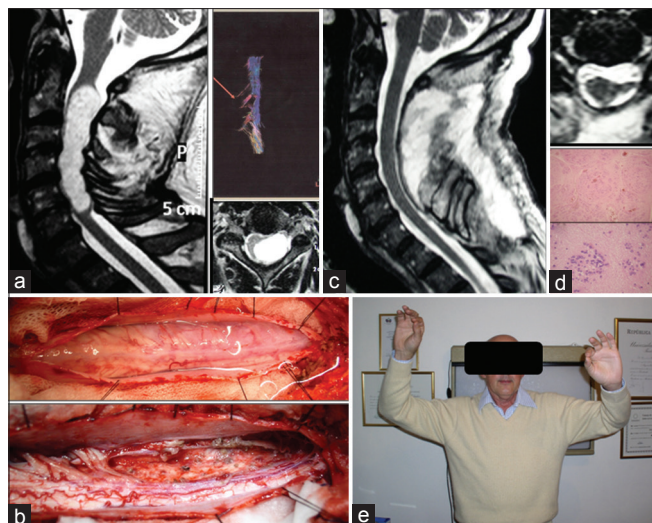
Patients' median age was 39.8 years (range: 16–62). The locations of the lesions were: Cervical: 11; dorsal: 6; and conus: 4 cases. In the preoperative period, 5 patients had mMs = 1; 10 cases mMs = 2; 4 cases mMs = 3; 1 case mMs = 4; and 1 case mMs = 5. In summary, 19 cases had mMs >3 and 2 cases mMs <4.

Total resection was performed in 20 cases (95.2%) [Figure 2]. Subtotal resection occurred in only 1 patient who had a D11-tanycytic ependymoma, in whom a small minimal remnant persisted, which was observed during a period of 14 years without progression [Figure 3].

The intraoperative frozen biopsy permitted the differentiation of infiltrative lesions from noninfiltrative ones in all cases. Later on, pathological analysis confirmed 5 ependymomas, 1 tanycytic ependymoma, 4 hemangioblastomas, 1 ganglioglioma, 1 subependymoma, 1 intramedullary schwannoma, 1 epidermoid tumor, 3 arachnoid cysts, and 4 vascular malformations.

Preoperatively, 8 cases had secondary syringomyelia that was resolved in all of the cases through surgical excision of the masses without additional treatment.

There were two complications. One patient presented cerebrospinal fluid (CSF) fistula with meningitis, which was conservatively resolved with antibiotic therapy, acetazolamide, and CSF external lumbar drainage for 5 days. Another patient, who was discharged, died from pulmonary thromboembolism on the postoperative 20<sup>th</sup> day.



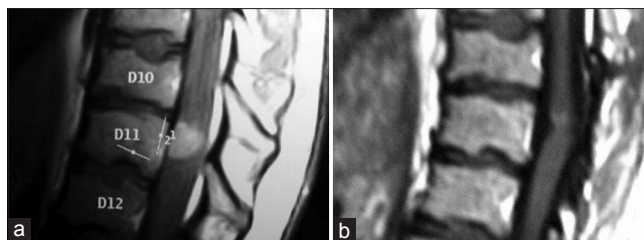
**Figure 2: Illustrative Case (n = 15). (a) Magnetic resonance (MR) axial, sagittal planes, and tractography; (b) surgical images; (c) postoperative MR of axial and sagittal planes showing radical resection; (d) pathology demonstrating subependymoma; (e) patient photo at postoperative 30<sup>th</sup> day**

The assessment of the functional status within the postoperative 90 days showed a stable functional condition in 9 cases (42.8%) and a better condition in 12 cases (57.1%). A temporary decline was observed in 2 cases with recovery before 90 days had passed [Table 1]. Four patients had persistent dysesthesias that required symptomatic medication with pregabalin.

Only 1 patient received postoperative radiotherapy and chemotherapy according to the characteristics of the histopathology; a ganglioglioma with areas of malignancy in the glial sector.

## DISCUSSION

The reported predictors of good outcome of the primary ISCTs are the preoperative functional status, histology of the tumor, and extent of the surgical resection.<sup>[2,6,7,10-12,14,15]</sup> The only variable that we as surgeons could influence is the last one, which is related to the infiltrative versus noninfiltrative nature of the ISCTs. In our case series, total resection was obtained in 95.2%. Despite the retrospective characteristic of our work, we believe that the recognition of infiltrative versus noninfiltrative lesions, according to Klekamp's classification, could be predicted with great certainty by thorough analysis of preoperative images, which is particularly improved in recent years by the input of diffusion tensor and tractography that showed displacement of ascending and descending spinal cord bundles with respect to the ISCTs. This is the main factor to consider in the preoperative planning of the extent of resection. Intraoperatively, the contribution of the frozen biopsy



**Figure 3: Illustrative case of subtotal resection (n = 11); tanyctic ependymoma. (a) preoperative MR showing the hyperdense mass at T11 level; (b) postoperative MR demonstrating the residual lesion.**

helps to identify lesions corresponding to noninfiltrative type as Klekamp's type A.

In regards to the results of the functional status reported in the literature, the improvement could be expected in 30–40% of the cases, stability in 50–60%, and decline in 5–10%.<sup>[1,8,10,11,14]</sup> We did not observe permanent functional deterioration in our case series; on the contrary, a high degree of improvement was achieved. Dysesthesias were present in patients who already had it in the preoperative assessment, and in all of the cases, there was improvement through pregabalin in personalized doses. The slightly better functional outcome obtained in our group of patients is probably related to the clear identification of the boundaries of lesional tissue despite the fact that the lesions were extensive. During surgery, the information given by the neurophysiological monitoring of sensory and motor evoked potentials represented a decisive factor to preserve functional neural tissue, thus favoring the maneuvers of dissection and limiting the use of bipolar coagulation during the resection process.

With regards to the surgical complications that are reported in the literature, these complications vary between 3.8 and 17%, and in most cases are CSF fistulas sometimes complicated with meningitis.<sup>[6,11,12]</sup> In our group of patients, we had 1 case with CSF fistula with meningitis, which was resolved with antibiotic therapy and conservative management of acetazolamide and CSF external lumbar drainage for 5 days. Another complication to prevent in adult patients is DVT for which prophylaxis through pneumatic compression on the lower limbs, elastic stockings, and low weight heparin, as well as ambulation and physical therapy are set up in early stages; however, even if this prevention is done, thromboembolism might occur.

Although the present study does not allow to reach statistical conclusions, we can conclude that every effort should be made to identify the Klekamp's Type A ISCTs during preoperative planning, and confirmed later on during surgery by frozen biopsies because these data determine the resectability of the lesion, encouraging us to perform radical surgeries with a low risk of functional declination.

**Table 1: Basal data and preoperative and postoperative functional assessment by Mc Cormick scale\***

Case	Age	Sex	Spinal lesional level	Histopathology	Preop McS	90 days Postop McS	Extent of resection	Complications
1	16	M	C7-D1	Arachnoidal Cyst	4	1	Total	
2	36	F	D3-7	Hemangioblastoma	3	1	Total	
3	59	F	Cono	Ependymoma	1	1	Total	
4	46	F	D5-6	Hemangioblastoma	3	1	Total	
5	19	F	C7-D1	Cavernoma	2	1	Total	
6	20	F	D9-11	Hemangioblastoma	5	5	Total	
7	42	F	D11-12	Arachnoidal Cyst	2	1	Total	
8	24	M	D11	Tanycytic Ependymoma	2	2	Subtotal	
9	62	M	C3-4	Hemangioblastoma	2	1	Total	
10	18	F	Cono	Epidermoid tumor	1	1	Total	
11	54	F	D8-9	Intramedullary schwannoma	2	2	Total	Dead
12	37	F	D3-4	Ependymoma	2	1	Total	
13	45	M	C4-7	Ganglioglioma	2	2	Total	
14	50	F	C4-5	Thrombosed AVM	3	2	Total	
15	54	M	C2-6	Subependymoma	2	1	Total	
16	57	M	C5-D1	Ependymoma	3	2	Total	
17	27	F	C6-7	Cavernoma	2	1	Total	
18	42	F	Cono	Cavernoma	1	1	Total	
19	45	F	C4-7	Ependymoma	1	1	Total	
20	40	F	C1-3	Neuroenteric Cyst	1	1	Total	
21	50	M	C3-7	Ependymoma	2	1	Total	CSF Fistula + meningitis

M: Male, F: Female, McC: Mc Cormick score, Preop: Preoperative, Postop: Postoperative. \*McCormick PC, Torres R, Post KD, Stein BM: Intramedullary ependymoma of the spinal cord. *J Neurosurg* 72:523–532, 1990

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## Conflicts of interest

There are no conflicts of interest.

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