Article

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Diagnosis of, surgical technique for and treatment results from medullary lipomas associated with spinal dysraphism

Experience with 38 patients

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ABSTRACT

Objective: To observe whether microsurgical removal of medullary lipomas and untethering of the medulla is a safe and efficient procedure. **Method:** A retrospective study was carried out on 38 patients with medullary lipomas associated with spinal dysraphism who underwent operations between January 1986 and January 2008, at the Neurosurgery Department of the Federal Hospital for State Public Servants, in Rio de Janeiro. **Results:** No deaths occurred in this series, and there was no worsening of motor or bladder function among the patients. Seven individuals presented improvements in their motor deficit. Nine patients presented improvements in bladder function. Three individuals with trophic lesions achieved wound healing. **Conclusion:** Microsurgical removal of medullary lipomas associated with spinal dysraphism proved to be a safe procedure without deaths and with a low morbidity rate, and several patients achieved improvements in their neurological symptoms.

Key words: spinal dysraphism, medullary lipoma of the conus, lumbosacral lipomas, lipomyelomeningoceles, anchored medulla, microsurgery.

Diagnóstico, técnica cirúrgica e resultados nos lipomas medulares associados ao disrafismo vertebral: experiência com 38 pacientes

RESUMO

Objetivo: Observar se a remoção microcirúrgica dos lipomas medulares e a liberação da medula da tração exercida pelo lipoma é um procedimento seguro e eficaz. **Método:** Realizamos estudo retrospectivo de 38 pacientes com lipomas medulares associados ao disrafismo espinhal operados entre janeiro de 1986 a dezembro de 2009 no Serviço de Neurocirurgia do Hospital Federal dos Servidores do Estado do Rio de Janeiro. **Resultados:** Nessa série não ocorreu nenhum óbito, ou piora da função motora ou vesical em nenhum paciente. Observamos melhora do défice motor em 7 pacientes. Nove pacientes apresentaram melhora da função vesical. Três indivíduos com lesões tróficas apresentaram cicatrização das suas feridas. **Conclusão:** A remoção microcirúrgica dos lipomas medulares associados ao disrafismo espinhal se mostrou segura, sem nenhum óbito, com baixa morbidade e com melhora dos sintomas neurológicos em vários pacientes.

Palavras-chave: disrafismo espinhal, lipoma de cone medular, lipoma lombo-sacro, lipomielomeningocele, medula ancorada, microcirurgia.

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Several names have been used to describe medullary lipomas of the conus associated with spina bifida: lipomyelomeningoceles, lipomyeloceles, medullary lipoma of the conus or of the filum terminale and congenital lumbosacral lipoma. Lipomyelomeningocele is often used as a general term for all lumbosacral lipomas, but these should be distinguished from intramedullary lipoma without spinal dysraphism[1-11]. Over the last few years, several neurosurgeons have reported their experiences with the treatment of lipomyelomeningoceles, in the literature[1-16]. Nevertheless, little attention has been paid to this subject within Brazilian settings, which motivated us to review and present the cases of this pathological condition treated by our medical team, and to assess the safety and efficiency of microsurgical removal of medullary lipomas and the degree of relief from the traction that the lipoma exerts on the medulla.

METHOD

A retrospective study was carried out on 38 consecutive patients with medullary lipomas associated with spinal dysraphism who underwent surgery between January 1986 and January 2008, at the Neurosurgery Department of the Federal Hospital for State Public Servants, in Rio de Janeiro. Radiological examinations, patient records, surgical descriptions and, when available, filming of the surgery, were reviewed, thereby creating a database from which information pertinent to the present study was gathered.

Surgery

All the patients underwent surgery and the same microsurgery technique was used, following these steps: General anesthesia was induced, with endotracheal intubation, and the patient was positioned in ventral decubitus, resting on the thoracolumbar support. A 4.5× loupe and coaxial lighting were used for the initial steps of the procedure. With a scalpel, a rectilinear midline incision was made, starting one vertebra above and extending to one vertebra below the subcutaneous fat mass, and proceeding around the cutaneous stigma. Throughout the procedure, careful homeostasis was performed with bipolar forceps under saline irrigation. Circumferential dissection was performed around the lipoma and/or the lipofibromatous talus. The penetration of the lipoma into the thoracolumbar fascia was viewed (Fig 2B). The fascia was opened along the midline with a scalpel and the paravertebral muscles were carefully disinserted and laterally retracted using a periosteal elevator. Exposure was maintained using autostatic retractors. The lamina and the spinous processes of the upper and lower vertebrae were viewed and the penetration of the lipoma or lipomatous talus into the osseous failure was observed. Laminectomy of the upper vertebra was usually performed. The normal condition of the dura mater and penetration of the lipofibromatous

Fig 1. Cutaneous stigmata. [A] Dimple, lipoma and hemangioma; [B] Dimple and vestigial tail.

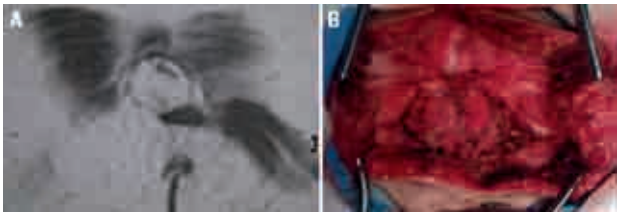

Fig 2. [A] T1-weighted MRI showing penetration of the lipoma into the lumbar fascia. [B] Surgical view of the same patient, revealing lipoma penetrating the lumbar fascia.

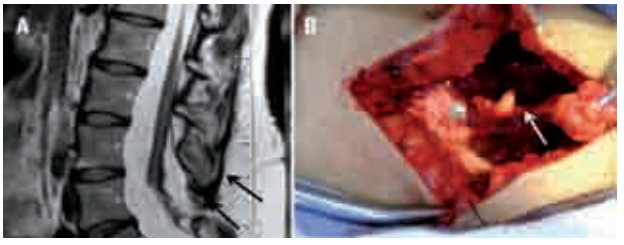

Fig 3. [A] T2-weighted MRI showing caudal lipoma with lipofibromatous talus penetrating the dura mater and adhering to the medullary conus, which was located in an abnormally low position (arrows). [B] Surgical view of a subcutaneous lipoma and lipofibromatous talus penetrating the dura mater (arrows).

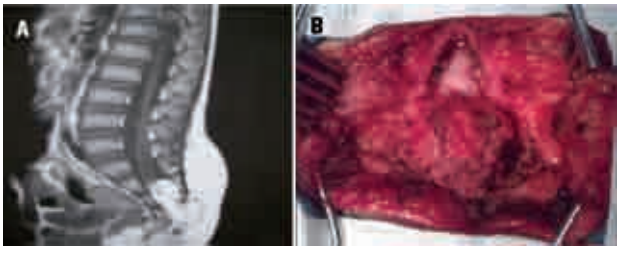

Fig 4. [A] T1-weighted MRI showing caudal lipoma penetrating the dura mater, with adherence to the medullary conus, which was located in an abnormally low position, and syringomyelia of the conus. [B] Surgical photograph showing a large caudal lipoma penetrating the dura mater.

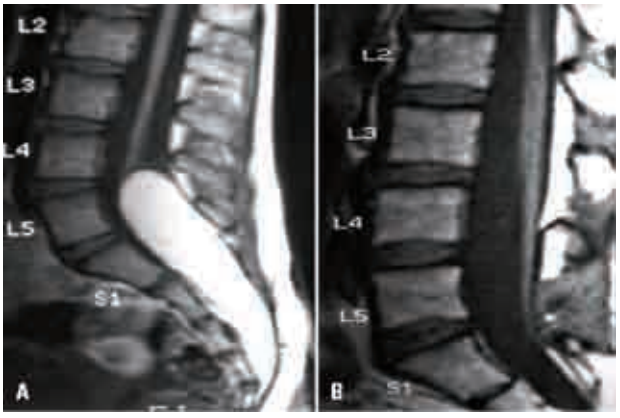

Fig 5. [A] Sagittal preoperative T1-weighted MRI showing large lipoma tethering the spinal cord. [B] Sagittal postoperative T1-weighted MRI demonstrating total removal of the lipoma.

talus were identified (Figs 3B and 4B). At this moment, a surgical microscope was introduced and the remaining surgical procedures were performed under magnification that varied from 10 to 16×. The dura mater was sectioned along the midline, while bypassing the lipoma or lipofibromatous talus. The free border of the dura mater was sutured to the paravertebral musculature using 4.0 thread. The adhesion of the lipofibromatous talus to the medullary conus, and the relationship with the nerve radices was identified. Using microsurgery scissors, the lipoma was resected as close as possible to the medullary conus, and as much adipose tissue as possible was removed. However, no attempt was made to resect the intramedullary portion of the lipoma, thus leaving the small remainder of the lipoma freely gliding within the canal sac. The arachnoid bands were also sectioned, but care was taken to lyse this adhesion without damaging functional nerve roots. The filum terminale was identified and sectioned, with the aim of relieving the medullary conus. We only used intraoperative electrical stimulation to locate functional neural elements in difficult situations. In all the cases, a decrease in axial tension was observed, while some cases presented cranial displacement of the conus. The viable radices of the cauda equine were placed in an anterior position and were either straightly aligned or had the outlet angle inverted. It was essential to preserve them. The dura mater was sutured hermetically using 4.0 thread. Duraplasty with fascia or using artificial dura mater was commonly performed to prevent straightening of the dural sac diameter and adhesions to the medulla. The Valsalva maneuver was performed with the aim of detecting whether any liquor fistula was present, and such cases were sutured. Biological glue was used on the suturing area with the aim of preventing liquor fistula. The paravertebral musculature and of the lumbar fascia were then closed in planes, using absorbable thread. The skin was sutured in a tension-free manner and the stitches were separated with monofilament thread. During the postoperative period, the patient was kept in the ventral decubitus position and in the Trendelenburg position from 3 to 5 days, to prevent any appearance of a liquor fistula. Prophylactic antibiotics were prescribed for 24 hours.

RESULTS

This was a retrospective study, and therefore it had inherent biases and drawbacks. We did not develop any special protocol for analyzing these patients. Nonetheless, in addition to the examinations performed in the neurosurgery department, the pediatric patients were routinely examined before and after the operation within the pediatric neurology and urology services and the adults, within the neurology department.

There were no deaths in the current series. The 38 patients included 14 men and 24 women, of ages ranging from 4 months to 37 years (average of 14.5 years).

The most frequent sign observed was subcutaneous lumbar lipoma, which was found in 31 cases (81.5%). Genitourinary malformations such as cloaca and duplication of the genitalia were detected in nine cases (23.6%) and a dimple was seen in seven cases (18.4%). Previous surgical scars, vestigial tails and angiomas were other signs observed in this series (Figs 1 and 2; Table 1).

The most frequent symptom was urinary incontinence, which was found in 21 patients (55.2%), followed by motor deficit in 13 (34.2%) and constipation in nine (23.6%). There were only three asymptomatic patients (Table 2).

X-rays identified spina bifida in 29 cases. Magnetic resonance imaging (MRI) clearly showed the lipoma penetrating the rachidian canal and the dura mater, with adhesion to the medullary conus, which was located abnormally low: below L2 in 16 cases (Figs 3, 4 and 5).

In the current series, we did not observe any worsening of motor or bladder function among the patients during the early postoperative period. Seven individuals (53.8%) showed improvements in their motor deficit. Eleven patients (42.3%) showed some improvement in their bladder function, among whom four presented decreases in repetition of urinary tract infections, and seven, decreases in the residual volume. There were three patients with trophic lesions, and they presented healing of chronic ulcerations. Three patients reported transient dysesthesia and numbness in the legs immediately after their operations. Over a mean follow-up period of 7.5 years (range: 2-13 years), we achieved a symptom improvement rate of 55.2%. Stabilization of the symptoms occurred in 10 patients (26.3%) and some degree of neurological deterioration was noticed in seven individuals (18.4%). The majority of the patients achieved

a satisfactory improvement, which allowed them to return to functional life. Despite the care mentioned above, there were four cases of CSF fistula (10.5%), which were treated with prolongation of the ventral decubitus and Trendelenburg positions. Reoperation to close the fistula was only necessary for one patient (2.6%). In all the other cases, the fistulas closed spontaneously, without infectious complications.

Selected case

A 21-year-old patient with lumbosacral lipoma, axial lumbar pain and urinary incontinence previously underwent two superficial interventions to reduce the lipoma at another medical institution. No improvement of the neurological condition was observed, and the lipoma regained the original size. At our department, the patient underwent microsurgical resection of the medullary lipoma, with untethering of the medulla, thus achieving better sphincter control and pain relief (Figs 4A and 4B).

DISCUSSION

These patients with lipomyelomeningoceles presented a subcutaneous lipoma in lumbosacral region, usually on the midline. Subcutaneous lipomas develop through closure defects in the thoracolumbar fascia, lamina and dura mater, with adherence to the medullary conus[1,4,5,7,10,11] (Figs 2,3,4 and 5). In a 15-mm embryo, the lengths of the medulla and spinal column are the same. From that stage onwards, the spinal column develops towards the caudal end more rapidly than the medulla does, thus causing the medulla to "ascend" towards the encephalon, inside the rachidian canal. The medullary conus usually reaches L3 during the 30th week of pregnancy and, in adult individuals, its location should not exceed the lower border of L2. If any process such as lipomyelomeningoceles keeps the medulla stuck in one position, thus preventing it from migrating, a gradual longitudinal stretching process that tethers the medulla will take place[9,14,16-20]. Chapman[21] defined three types of lipoma: dorsal, caudal and transitional. The dorsal variant attaches to the dorsal aspect of the conus. The caudal type attaches to the inferior aspect of the conus and the transitional variant is a combination of these two. Lipomas of the terminal filum are mostly fully enclosed inside the dura and seldom become symptomatic and require treatment. The great majority of our patients presented with the caudal or transitional variant. We only found three patients with dorsal lipomas and three with lipomas of the filum.

Yamada et al.[20] demonstrated experimentally that the traction exerted on the medulla causes changes to the oxidative processes of the neuron mitochondria. This might cause paraplegia or urinary incontinence. Patients with lipomyelomeningoceles are, most often, born neurologically normal. The risk of neurological and/or bladder deterioration exists at all ages, but particularly during periods of rapid growth[3,9-11,13,16]. Reviewing the literature, Pierre-Khan et al.[6] observed that between 35 and 67% of the patients developed neurological or bladder lesions when not properly treated. Other authors[3,7,13-15] have believed that the majority of the patients will present symptoms before they are four years old. The neurological and/or bladder deterioration among these patients is most commonly progressive, but it can appear abruptly during the course of certain events such as natural birth, practicing of physical exercises, sexual intercourse or accidents[1,2,6,7,10,16]. Several authors who operated on symptomatic patients have stated that very few of them achieved total regression of their deficits. It is

Table 1. Signs of 38 patients with medullary lipomas.

Signs	#Patients (%)
Lipoma	31 (81.5)
Cloaca	8 (21.0)
Dimple	7 (22.5)
Scar	7 (22.5)
Foot asymmetry	5 (13.1)
Cutaneous angioma	4 (10.5)
Wounds on the legs	2 (5.2)
Hypertrichosis	1 (2.6)
Dermal sinus	1 (2.6)
Escoliosis	1 (2.6)
Hydrocephalia	1 (2.6)
Muscle atrophy	1 (2.6)
Vestigial tail	1 (2.6)
Total	70

*Some patients presented more than one sign.

Table 2. Symptoms in 38 patients with medullary lipomas.

Symptoms	# Patients (%)
Urinary incontinence	21 (55.2)
Paraparesia	10 (26.3)
Constipation	9 (23.6)
Pain	7 (18.4)
Urinary retention	5 (13.1)
Hypoesthesia	4 (10.5)
Monoparesis	3 (7.8)
Saddle block anesthesia	3 (7.8)
Total	62

*Some patients presented more than one symptom

harder to achieve recovery of bla dder function than of motor function[2-6,11,13]. These characteristics were also observed throughout our clinical experience.

In this present group, we achieved a symptom stabilization or improvement rate of 81.5%. The aim of surgical treatment is to release the medulla from the traction exerted by the lipoma and, in some cases, decompression of the medulla relieves the mass effect. Therefore, cosmetic operations that do not untether the medulla are a risk factor for delayed neurological deterioration[3,4,5,7,9,10,13-15]. This phenomenon occurred in the cases of six patients who had undergone previous surgery at other institutions.

Recently, the treatment of these lesions has generated much controversy. Some physicians have advocated surgical treatment for all patients regardless of symptoms, while others have proposed that surgery should be withheld until symptoms develop[10].

Kulkarni et al.[12] found that the incidence and patterns of neurological deterioration seemed to be very similar, regardless of whether or not early surgery had been performed. These results suggest that conservative treatment of asymptomatic patients is a reasonable option. They observed that, during the follow-up period, deterioration was experienced by 33% of the patients who had been conservatively treated, and 46% of those who had undergone prophylactic surgery.

On the other hand, Pang et al.[14,15] concluded that total and near-total resection of lipomas and complete reconstruction of the neural placode produced a much better long-term result, with greater likelihood that it would be progression-free, than seen in cases of partial resection and non-surgical treatment. With total resection, preoperative complications can be successfully treated with low surgical morbidity and a high yield of agreeable long-term postoperative results.

In the present series, all the symptomatic patients underwent the microsurgical technique for total/near total lipoma resection, untethering of the medullary cone, lysis of adhesions and sectioning of the filum terminale, thereby releasing the medulla from any traction, followed by duraplasty to avoid reanchoring. Some neurosurgeons have suggested that intraoperative monitoring should be used, such as rectal EMG, urethral EMG, continuous EMG, evoked potential and other types. To date, there has not been any proof that such methods are effective for avoiding injury. In the initial cases of this series, we used intraoperative electrical stimulation to locate functional neural elements, but with progressively increasing experience within our group, we began to use neural stimulation only for difficult situations.

The extent of surgical resection and untethering of the conus was established taking into consideration the surgeon's opinion and the postoperative images (MRI and/or CT scans). Over the period (22 years) covered by the present study, there has been remarkable progress in imaging techniques. With the first patients of this series, assessment was made based on CT scans, which do not allow precise measurement of the tumor residues. In eight cases, postoperative MRI was available for assessments, and we observed near total removal (>90%) in five cases (Figs 5A and 5B), and partial resection in the other three patients. Radiological evaluation of the untethering is always difficult to make. We were only able to established ascending of the conus in two individuals with certainty.

Kulkarni et al.[12] suggested that it may be difficult to set up close observation of patients in our environment, due to the long distances that usually exist between the patient's home and the hospital, thus making neurological surveillance more difficult. In our series, there were even some patients who came from indigenous tribes. Therefore, management remains an open question[10].

For symptomatic patients, we recommend microsurgical treatment for all patients, regardless of the symptoms, in order to protect the neurological function and delay any neurological decline. Our experience with asymptomatic patients only consists of three cases, and it is difficult to reach any conclusion with such a small sample.

We can conclude that medullary lipomas of the conus associated with spinal dysraphism are a significant cause of neurological dysfunction. Early diagnostic procedures should be followed up by pediatricians, urologists or plastic surgeons, thus helping to select the appropriate treatment. Microsurgical treatment can be expected to achieve total/near total resection of the lipoma with relief of the traction on the medulla.

Our experience described above, on 38 cases operated on using microsurgical techniques, showed that this is a safe method, with no deaths, low morbidity and satisfactory results, comparable to those found in the international literature. For asymptomatic patients, the treatment should be decided on an individual basis.

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