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Intraspinal lipomas in infancy and childhood causing a tethered cord syndrome

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Abstract

The authors report on a series of 26 children with spina bifida occulta in combination with intraspinal lipoma and clinical signs of tethered cord syndrome. The age of the children at presentation ranged from 1 month to 12 years. The typical signs and symptoms consisted of skin lesions in the lumbar-sacral region, neurogenic foot deformities, and bladder and bowel disturbances.

The diagnosis was confirmed by neuroradiological investigations including lumbar myelography, computerized tomography, and only recently magnetic resonance imaging. Indications for surgery were based on the radiological evidence of intraspinal lipoma and tethered cord and especially on the clinical signs of neurological deterioration.

The results of the operative treatment are presented and the role of surgery before onset of symptoms as well as during the symptom-free interval is discussed.

Keywords: Intraspinal lipoma, spina bifida occulta, tethered cord syndrome.

1 Introduction

RECKLINGHAUSEN was the first to publish a report of a spinal lipoma causing the fixation of the spinal cord [23]. In 1886 he described a lipomatous tumor reaching from the subcutanous area down into the intradural space which had grown into the tethered conus medullaris at the level of the second vertebra of the sacrum.
Further case reports were published by BRICKNER in 1918 [5] and JAMES and LASSMANN [12] in 1960.

During embryonal development the differential growth of the vertebral column and the spinal cord leads to a relative cranial ascent of the cord and to the formation of the cauda equina. After the 25th week of the embryogenesis the conus reaches the L3 level where it remains until the time of birth. During the first five years of life the conus migrates upwards to the interspace L1–2.

This cranial ascent can be disturbed by many factors. A filum terminale lower than the L2 level and with a diameter of more than 2 mm is considered pathological [1, 7, 9, 10, 17, 20, 24]. Fixation of the cord or the filum terminale at the dura and the subcutanous tissue is the most common cause of a malascensus or tethered cord syndrome. Another possible cause of malascensus is intraspinal lipoma. These lipomas are thought to develop from normal fat cells which are included in the neural tube during its development [9, 13, 17, 24].

In cases of severe neurological deficits there should be no difficulties in the diagnosis of dysrhaphic disorders. But the normal or uncomplicated case can easily be overlooked in spite of subcutanous lipomas or excess hair growth at the lumbar sacral area already present at birth.

The tension of the cord causes weakness and motor imbalance of the foot and leg. These can result in foot deformities and/or scoliosis and consequent bladder disturbances which can be a major symptom.

2 Patients

26 infants and children suffering from dysrhaphism and a tethered cord syndrome were operated on at the Hannover Medical School from 1973–1986. Not included are newborns who had to be operated on for myelomenigoceles or dermal pits over the lumbosacral spine. At the time of diagnosis the patients age ranged from 1 month to 12 years.
16 patients (Group 1) were under the age of 2 years.
10 patients (Group 2) were over the age of 2 years.
The number of female patients surpassed the number of the male patients by a ratio of 8 : 5 which is in accordance to the literature [1, 2, 3, 7, 8, 11, 15, 16, 19, 20, 22].

3 Signs and symptoms

Skin lesions like flame nevus, hyperpigmentation, hypertrichosis, or subcutanous lipomas over the lumbosacral spine were the most common cause for admission. In 12 out of 16 patients of Group 1 these signs were found at birth. 8 out of 10 patients of Group 2 also presented with similar skin lesions at birth. The reason for postponing diagnostic procedures despite these obvious dermatomogical signs could not be elucidated. The most frequent neurological deficits were the absence of tendon reflexes (20 of our patients in both groups), followed by disturbed bladder and bowel functions together with sensory disturbances in this area. In 6 out of 10 patients in Group 2 this dysfunction was only secondary.

Weakness and motor deficits were distributed equally in both groups. These usually consisted of a

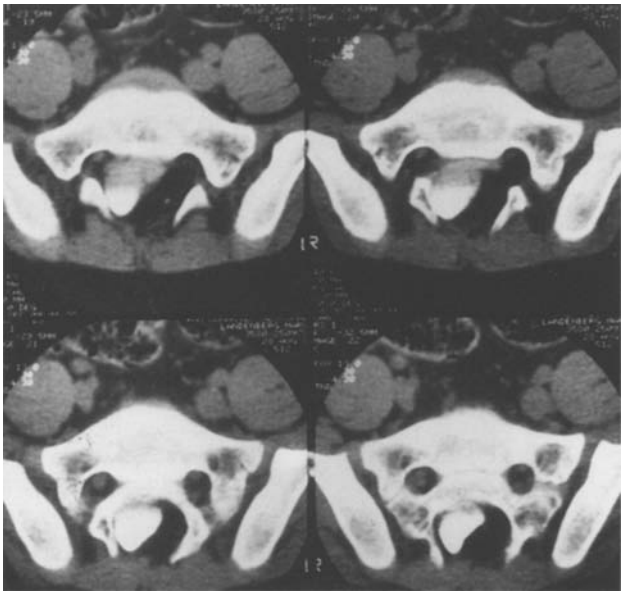

Figure 1a. Lumbar myelo-CT of level L5/S1 with dyslocation of the distal lumbar column to right side in combination with spina bifida and lipoma in an 8 year old boy.

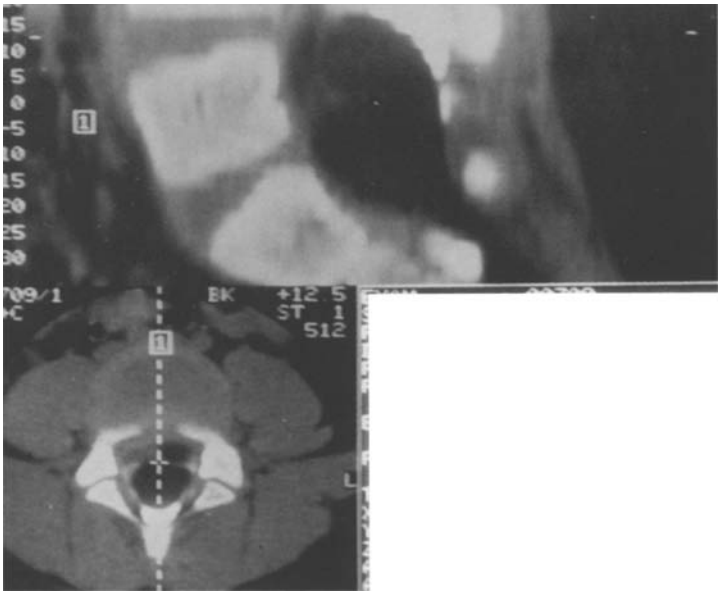

Figure 1b. Same patient: Sagittal reconstruction of the myelo-CT demonstrating the intraspinal extension of the lipoma with subcutaneous growth and tethering of the medullary conus.

weakness of the foot extensors and flexors in one or both legs. Muscle wasting was a common clinical sign. Four children presented with an asymmetry of the legs and feet. Isolated foot deformities were observed in 11 cases. These misdiagnoses led to a long lasting orthopedic treatment just as secondary bladder impairment led to urologic treatment. In the children in Group 2, misdiagnosis in 6 patients led to various treatments including cast redressing of a cavus foot deformity as well as plastic surgery of the foot tendons. In these older children, progressive neurologic deficits with bladder and bowel impairment were the symptoms which led to admission and to further diagnostic and, finally, therapeutic procedures. A summary of the symptoms is compiled in Table I.

Table I. Clinical findings in 26 patients with tethered cord syndrome and spinal lipoma

Tendon reflex failures	20
one side	9
both sides	11
bladder and bowel impairment	15
primary	9
secondary	6
foot deformities	11
motor weakness	10
sensory disturbances	10
muscle wasting	10
palpable subcutaneous lipoma	10
nevus	10
hemangioma of the skin	5
exuberant hair growth	5
back pain	1

4 Diagnostic procedures

The above-mentioned skin lesions and subcutaneous lipomas were indicative for spinal dysraphism. The diagnosis was confirmed by plain x-ray examinations which revealed anomalies of the lumbosacral arches in all of our 26 patients. In 10 cases the failed fusion of the arches were localized at the lumbar and sacral levels (Figure 1a, b). In 3 patients open lumbar arches and a sacral agenesis was demonstrated. All our children presented scoliosis as a typical sign of spinal cord tethering.

A radiological examination was performed using contrast media in 25 out of 26 surgically treated children. In 23 cases this consisted of a myelography. In two cases a computerized spinal tomography with a small amount of contrast medium was also carried out. In all of these children a conus medullaris at a low level (below L3/4) was found.

In 13 cases a megacauda was present. Nerve roots running nearly horizontal to the periphery were evaluated as a typical sign of tethered conus medullaris (Table II [16, 25]).

The levels of the tethered conus in myelography and CT scan were in accordance with the surgical findings in 18 out of 26 cases (about 60%). The most

Table II. Radiological findings: plain x-ray, myelography, and contrast CT examination

Failure of the arches	26
lumbar	8
sacral	8
lumbar and sacral	10
Agenesis of the sacrum	3
Megacauda	13
low level of the conus	25
Diastematomyelia (bony spur in the midline)	3

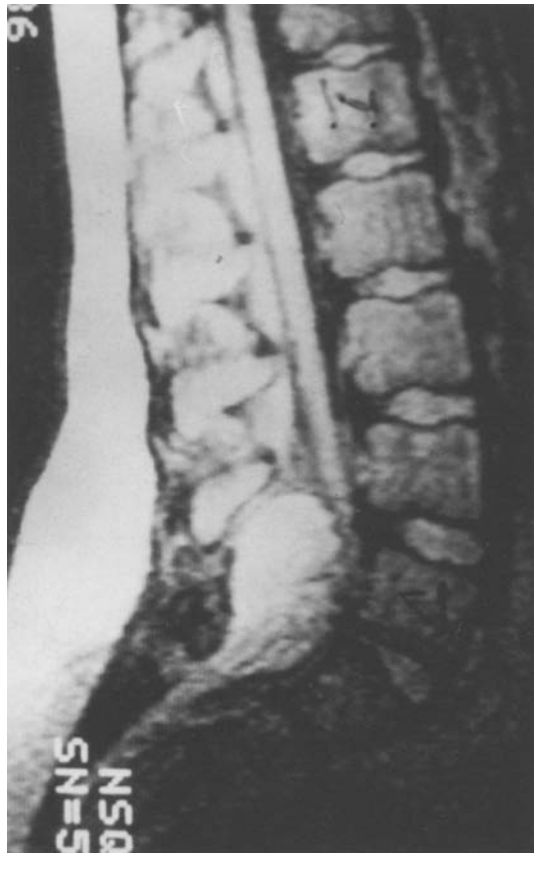

Figure 2. Lumbar magnetic resonance image of the same patient with excellent demonstration of the intraspinal lipoma at the L5 level and tethering of the conus medullaris.

often verified level of fixation of the conus to the dura and the transmigrating lipoma was found to be at L5 and S1 (15 cases).
Modern procedures such as MRI allow an excellent anatomical imaging of the structures to be operated on (Figure 2).

5 Surgical treatment

The aim of surgery is to untether the cord, to eliminate the mechanical constraint, and to release the elongated conus. The simplest way to accomplish this is usually to resect the thickened filum terminale which would release the elongated conus. Untethering is more difficult when the lipoma attachements must be dissected and a lipomatous mass excised. Because the layer between lipoma and neural tissue is poorly demarcated, even when microsurgical techniques are employed the mass of the lipoma may often only be reduced so that both the dural adhesions and the filum terminale have to be thouroughly dissected. In our clinic a CO_2 laser and CUSA are now being used for dissection and reduction of the intraspinal lipoma mass (Figure 3a, b). As a general rule we restrained from radical excision of the lipoma in order to preserve functionally important neural elements (Figure 4a, b c).

The intraoperative findings are summarized as follows:
In 21 cases we found an extra- and intradural lipoma growing into the conus and into the thickened filum terminale.

In 5 cases we found a extensive intradural lipoma as well as adhesions to the posterior dura causing the tethering of the cord.

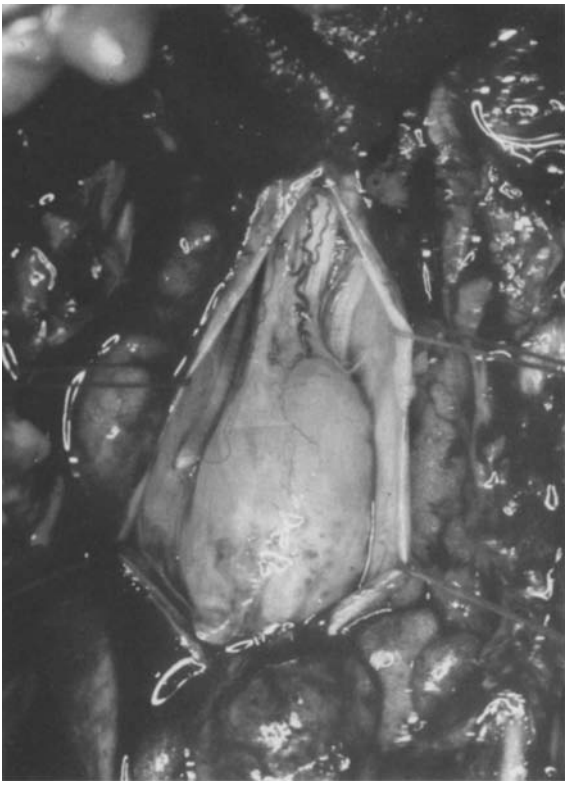

Figure 3a. Intra-operative image of a lumbar sacral lipoma in situ together with a tethered conus medullaris.

Figure 3b. Intra-operative image of the same patient following total exstirpation of the lipoma and untethering of the conus. The thickened filum terminale is well demonstrated.

Table III. Intraoperative findings

intra- and extradural lipoma	21
intradural lipoma	5
thickened filum terminale	16*
normal filum terminale	1
occult medullary plate	6
diastematomyelia	4
diplomyelia	2
torquated cord	2
cysts	7
megacauda	13

* diameter > 2 mm)

In 6 cases we found an medullary plate directly beneath the skin and partly attached to the lipoma. In 7 cases we found cysts in the conus medullaris. In 4 cases we found a diastematomyelia with bony spur and duplication of the dura.

Excision of the bony spur as well as lysis of the surrounding adhesions were carried out. Diplomyelia and torquated spinal cord were each found twice. Our findings are compiled in Table III.

6 Results

In our 26 patients there were no mortalities and no post-operative complications. Postoperative neurological deterioration did not occur in any patient. 7 cases improved. In 3 cases only the bladder and bowel impairment disappeared, whereas in 2 older children subjective amelioration of symptoms was reported. Preoperative muscle atrophies, disturbances of tendon reflexes, and foot deformities could not be ameliorated.

7 Discussion

The disturbed ascent of the spinal cord within the vertebral canal, which can be caused by lipomyelomeningoceles, a thickened filum terminale or ab-

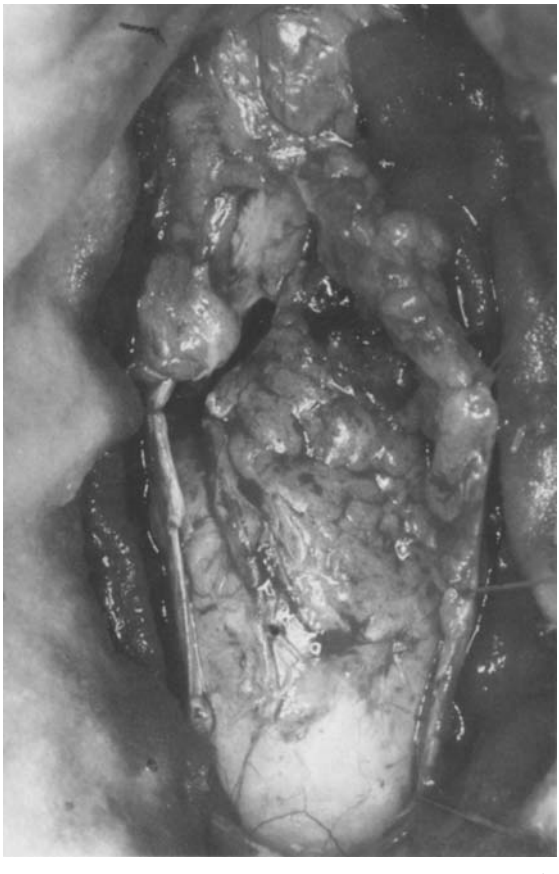

Figure 4a. Typical intraoperative image of a lumbar sacral lipoma.

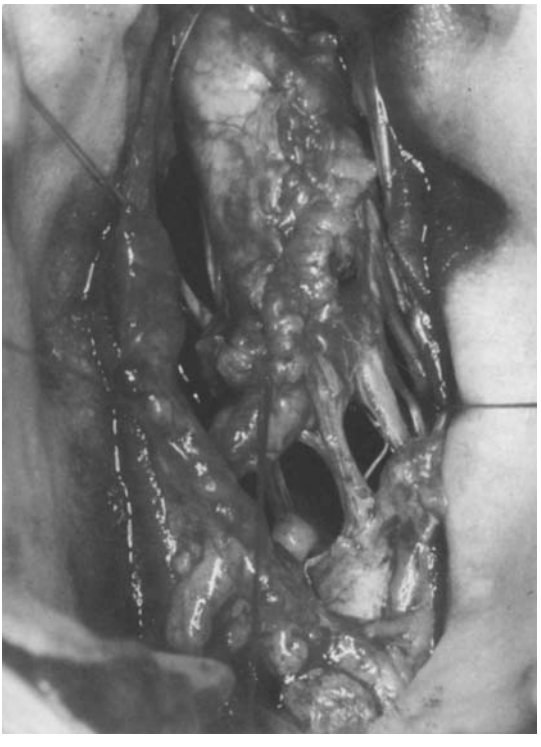

Figure 4b. Intraoperative image of the same patient following partial exstirpation of the lipoma and untethering of the nerve roots.

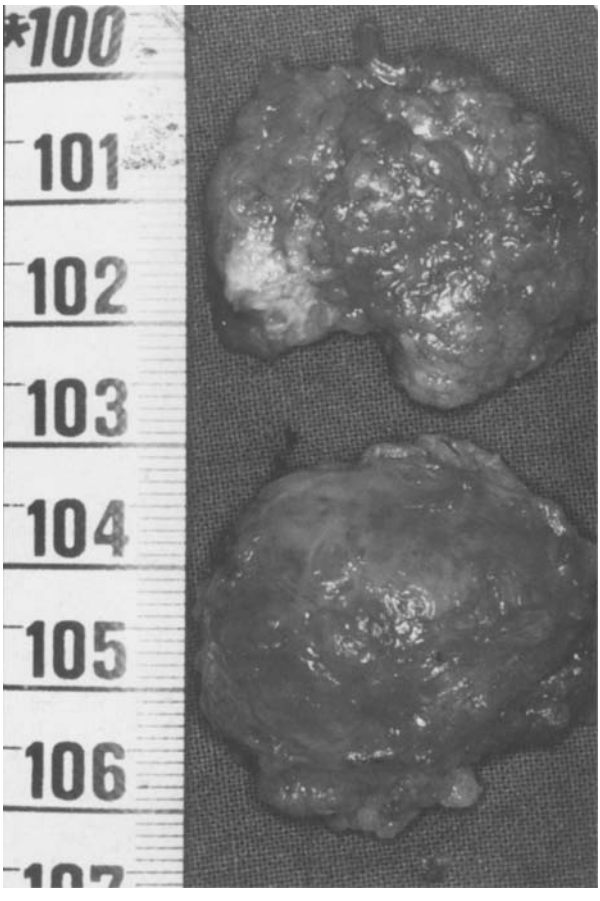

Figure 4c. Operative specimen of the exstirpated lipoma.

normalities like diastematomyelia, is a severe disease accompanied by progressing neurological deficits as well as deformities of the lower limbs and scoliosis. These signs and symptoms worsen with the age of the patient [1, 2, 7, 8, 9, 10, 16, 18, 20]. In addition, the already tethered spinal cord is much more vulnerable to mechanical alterations and injuries to the vertebral column. Children beyond the age of 2 demonstrated the highest rate of progressive neurological deterioration. This is in accordance to the experience reported by other authors [7, 11, 15, 19, 21]. The treatment of choice in diagnosed tethered cord syndrome is neurosurgical intervention. This is the opinion of all authors, especially as the operation using microsurgical techniques no longer involves severe complications. Various opinions have been published concerning prophylactic surgical treatment, i.e. surgical untethering of the cord in patients who show no evidence of neurological deficits. Some authors advocate a wait and see attitude and monitoring of the neurological status [4, 6]. Others tend to prophylactic surgical treatment in order to avoid the development of neurologic disturbances and deficits [12, 18]. According to our experience, as soon as dysfunction has occurred no return to normal can be expected from the operation. Moreover, foot deformities cannot be ameliorated even though one case of amelioration of a cavus foot deformity has been reported [14]. The results achieved in our prophylactically operated patients (Table IV) as well as the results reported by 7 other authors are encouraging. Considering that the rate of complications is almost negligible, we advocate the prophylactic early operation even though malformation of the nerves and their rootlets may hinder amelioration after surgery [3].

In our opinion early operation will protect symptom-free patients against the development of neurological deficits. Follow-ups of these patients show that 60% develop symptoms of a tethered cord during adolescence [1, 2, 9, 12, 13]. In contrast, operative and post-operative complications only occur in 0–8% of patients [3, 7, 11, 21]. These facts support our preference for early operation.

Table IV. Results of surgical treatment (review of 8 reports)

Author	Year	Number of operated patients	Deterioration	Amelioration	unchanged
Lassmann und James[15]	1967	22	2	5	15
Andersson[2]	1975	21	0	0	21
Mc Lone et al.[21]	1983	42	2	0	40
Chapman und Davis[8]	1983	17	0	0	12
Hoffmann et al.[11]	1985	62	10	8	44
Pierre-Kahn et al.[22]	1985	55	2	26	27
Bakker-Niezzen et al.[3]	1985	25	7	1	17
Stolke et al.	1986	26	0	7	19

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