LUMBAR LIPOMA AND OCCULT LUMBAR DYSRAPHISM

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Lipomas in the form of a human tail in the lumbar region with occult spinal dysraphism constitute a very rare dysraphic spinal abnormality, which is seldom seen in daily practice. This clinical entity commonly presents with local changes alone, and usually lacks neurological manifestations, although life-long risk of neurological deterioration is not negligible. Surgical intervention for asymptomatic lipoma of the conus medullaris has been an area of controversy, primarily due to the scarcity of detailed studies regarding the natural history of this disorder. Nevertheless, the presence of esthetic or functional disorders warrants surgical removal of these superficial masses. We report the case of a 10-year-old boy seen initially as an infant because of a lumbar appendage that caused only esthetic concern. At that time, the child's parents refused surgical removal of the malformation. Ten years afterwards, the patient himself presented our outpatient clinic requesting the removal of his esthetic defect.

Key words: Human tail ▪ Lumbar skin appendage ▪ Lumbosacral lipoma ▪ Occult spinal dysraphism ▪ Neurosurgery

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Introduction

Lumbar spinal lipoma with the appearance of a human tail, associated with occult spinal dysraphism, constitutes a spinal abnormality, which is seldom found in daily practice. This clinical entity commonly manifests with local changes alone, and usually lacks any neurological significance. Neurological sequelae include paresis and sensory deficits in the lower limbs, neurogenic bladder and bowel disturban-

ces, pain and neuro-orthopedic syndrome as well as high risk for secondary neurological deterioration(1). Surgical intervention for asymptomatic conus medullaris lipoma represents an area of controversy, mainly due to the paucity of studies regarding to the natural history of this disorder. Nevertheless, the presence of esthetic or functional disturbances warrants the surgical removal of these superficial masses.

We describe the case of a 10-year-old boy who was seen initially as an infant due to esthetic concerns related to a lumbar spinal lipoma. The child's parents refused surgery at that time. Ten years after the boy's initial presentation, the patient presented at our outpatient clinic and requested surgical removal of the malformation due to esthetic reasons.

Case report

A newborn baby was first brought to the outpatient clinic because of a 2-cm long low-back midline appendage that was not accompanied by any other physical or neurological complaints. Surgery was scheduled several months later but it was not performed at that time due to economic reasons. In June 2006, the now 10-year-old boy consulted a neurosurgeon for removal of the lesion for cosmetic reasons. The boy's neurological examination was normal. Local examination confirmed the presence of a 15 cm long lumbar skin appendage that resembled a human tail (Figure 1).

Radiographs of the boy's lumbar spine revealed posterior defects of laminae at L5 and S1 levels (Figure 2).

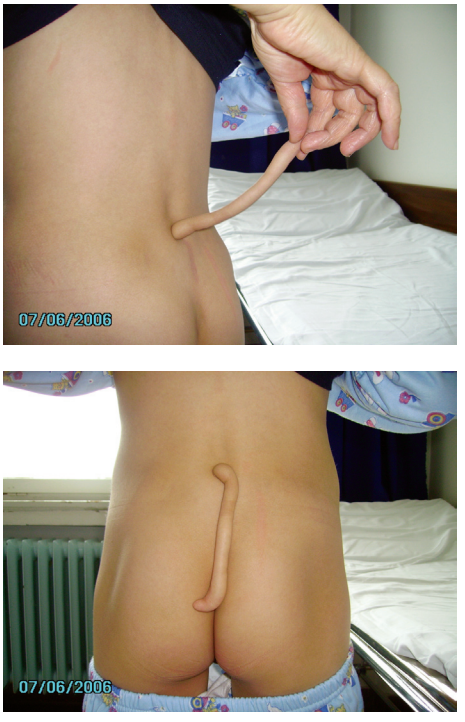

Figure 1 *Photograph of the patient showing a skin tag that resembles a tail and a subcutaneous lipoma*
Slika 1 *Fotografija pacijenta sa kožnim dodatkom koji sliči repu i potkožnom lipomu*

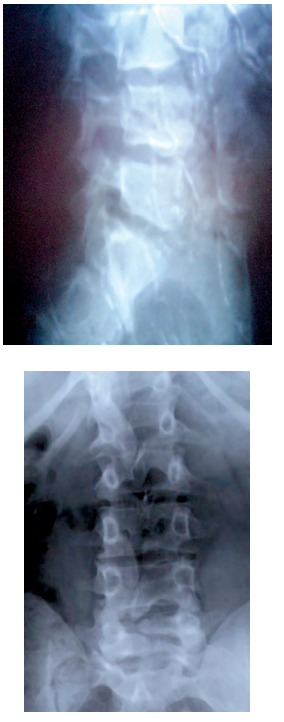

Figure 2 *Lumbar spine radiographs revealing widespread posterior defects of the laminae*
Slika 2 *Rendgenogram kralješnice sa defektom stražnjega luka kralješka*

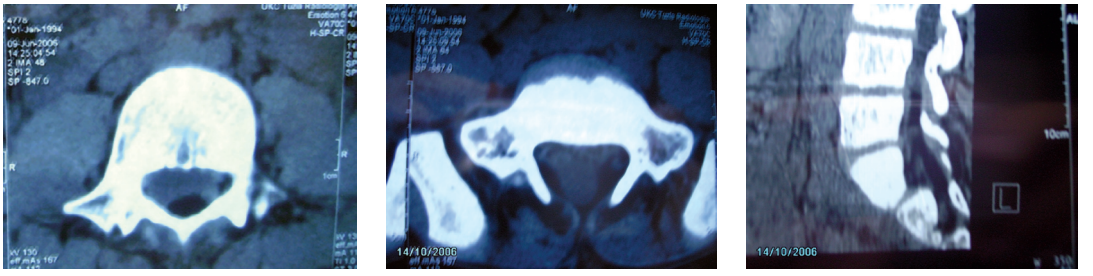

Figure 3 *CT images showing a defect of the lamina and the presence of a lumbar lipoma*
Slika 3 *CT ukazuje na defekt stražnjeg luka lumbalnog kralješka i postojanje lipoma*

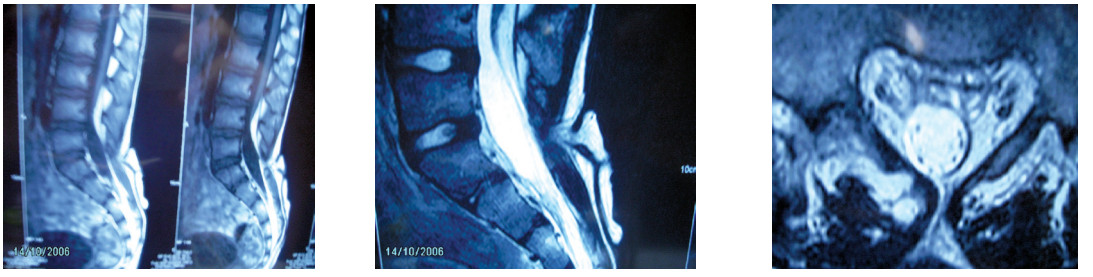

Figure 4 *MRI of the lumbar spine revealing spina bifida occulta and an intradural lipoma*
Slika 4 *MRI lumbalne kralješnice sa zatvorenom spinom bifidom i intraduralnim lipomom*

Computerized tomography scan confirmed the defect of laminae and also revealed a hypodense intra- and extra spinal mass (Figure 3). Magnetic resonance imaging (MRI) revealed an extra- and intradural lipoma around the dura mater and neural tissues within the spinal canal (Figure 4).

The patient underwent surgical resection of the extraspinal portion of the lesion and partial removal of the intraspinal extradural lipoma. Histopathological examination of the excised mass showed that it consisted primarily of fat cell deposits interlaced by numerous blood vessels. The patient's postoperative recovery was complete.

Discussion

Lipomas of the spine are among the most fascinating lesions encountered by pediatric neurosurgeons comprehending. Terminology related to spinal lipomas may be a daunting task, due to the fact that distinguishing these lesions from accumulations of fat in the vertebral canal is rather confusing. Lipomas of the lumbar spine constitute a very rare occurrence and cause symptoms related to their mass effect and compression radiculopathy. Likewise, lipomas of the conus medullaris are the most common form of fatty masses in the spine and can be broadly classified into different forms. These lesions are a manifestation of an occult spinal dysraphism and a frequent cause of the tethered cord syndrome (2, 3). Spinal dysraphism may exist in an open form (spina bifida aperta), as well as in a closed form, (spina bifida occulta). Symptoms may appear after hypoxic damage to the conus medullaris (4). Improvement in spinal cord blood flow has been noted after surgical decompression of the spinal cord. Clinical manifestations en-

compass a vast array of symptoms and signs, including, among others, cutaneous stigmata, vertebral anomalies, orthopedic abnormalities (scoliosis and extremity abnormalities), neurological deterioration at the level of the lower spinal cord including bowel and bladder dysfunction, and anorectal malformations (5, 6). Cutaneous marks without neurological symptoms are the most common form of presentation during the pediatric age.

Occult spinal dysraphism with lipoma sometimes lacks neurological changes. Most often, lipomas are localized extraspinal and they seldom exhibit intra-dural and/or intraspinal involvement. Extraspinal and extradural forms usually present without neurological changes. It is possible for lipoma to grow and compress the lumbosacral nerve roots, thereby causing radicular symptoms. Intradural lipomas usually present with neurological changes, due to compression of the conus medullaris.

Diagnosis of these abnormalities is most readily made in the setting of clinical findings supplemented with neuroimaging studies. Clinical evidence of a subcutaneous lipoma, including skin changes, orthopedic anomalies, vertebral abnormalities, and associated anorectal malformations suggests spina bifida. CT scanning, MRI, and plain radiographs assist to the process of localizing the level of the conus medullaris and to identify specific lumbar spine changes. Neurological and urological status may be further investigated with electromyography, cystometry, and evoked potentials.

Surgery for lumbar lipoma with spina bifida occulta is an area of controversy, primarily due to the paucity of detailed studies on the natural history of this disorder (7, 8, 9). Many authors advocate early prophylactic surgery to prevent deterioration (7, 10, 11). Others argue that, in their experience, prophylactic untethering may not prevent some deterioration and, because the natural history of asymptomatic conus medullaris lipoma is not clearly known, prophylactic untethering may not be warranted (9). Most authors propose early prophylactic surgery in patients with asymptomatic lipoma of the conus medullaris, due to the low rate of neurological deterioration (3-4%) that results from surgery and because of the better results of operations performed on asymptomatic patients as compared to the surgical outcome in symptomatic patients (7, 10, 11). If indicated, surgical intervention for lipoma that involves the spinal cord or conus medullaris includes intraoperative identification of the tumor (lipoma), release of the spinal cord, and reconstruction to as normal an anatomy as possible. A carbon dioxide laser has been found to be useful for debulking and dissecting intradural lipomas and for reducing blood loss. The surgical goal of these untethering operations is to stabilize neurological function and to prevent any further deterioration. Surgery-related complications include standard anesthesia-related risks, neurological deterioration, cerebrospinal fluid leakage, and meningitis.

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Sažetak

LUMBALNI LIPOM I OKULTNI DIZRAFIZAM

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Spinalni lipom u križi sa okultnim spinalnim disrafizmom je vrlo rijetka anomalija kralješnice. Ove promjene obično se manifestiraju lokalnim promjenama na koži, bez neuroloških smetnji, iako dugorčni rizik za razvoj neurološkog deficita nije zanemariv. Liječenje ovih promjena, koje su najčešće asimptomatske, je još uvijek nedovoljno definirano zbog nejasnog nastanka ovih promjena i porijekla tkiva koje se u ovim promjenama nalazi. Razlog za hirurško zbrinjavanje su kozmetske smetnje i funkcionalni poremećaji. Opisujemo kliničke smetnje desetogodišnjeg dječaka kome je dijagnosticiran spinalni lipom po rođenju. Roditelji dječaka po rođenjju, protivno savjetu ljekara, nisu željeli hirurško liječenje. Deset godina nakon toga pacijent na lični zahtjev, iz estetskih razloga, sam je zahtijevao pregled ljekara neurohirurga, nakon čega je i načinjeno operativno liječenje.

Ključne riječi: ljudski rep ▪ lumbalni kožni dodaci ▪ lumbosakralni lipom ▪ zatvoreni spinalni rascjep ▪ neurohirurgija

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