Congenital Spinal Lipoma: analyzing the perplexed nomenclature and our management

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ABSTRACT

Objectives: In this analysis, we will investigate the epidemiological features of spinal lipomas diagnosed at a single institution.

Methods: This study was carried out by reviewing retrospectively the: patient's admission records, neuroimaging, operation records, and outpatient files and biopsy results were used to collect the data from all consecutive patients treated in single referral center. Patient data were extracted from the King Hussein Medical Center (KHMC) Primary Spinal Tumors electronic Database, between 2006 and 2016.

Results: Our cohort consisted of 33-females (55.93%) and 26-males (44.06%), mean age: 3.45 ± 1.74 years. The congenital spinal lipomas were categorized into: 40-conus lipomas (17-terminal, 15-transitional, 8-dorsal) and 19-filum lipomas, including 11-patients who had lipomyelomeningocele. The most common promoter for diagnosis was skin stigmas (46.26 %), followed by associated malformations (30.63 %), and symptoms (23.11 %). Prophylactic surgery was undertaken in selected cases. In the initially asymptomatic group, 6-patients (28.57%) had late neurological deterioration. Of the 8-patients with asymptomatic conus lipomas, 3-cases (37.50%) developed sphincter dysfunction and motor problems at long-term follow-up. In the symptomatic group, 67.50% improved, 20% remained unchanged, and 12.50 % had late neurological deterioration. None of the 6-patients with symptomatic filum lipoma deteriorated postoperatively. Postoperative complications developed in 9 patients (13.55 %): seven transient local problems, 2 definitive urological deterioration .

Conclusion: Despite the lack of knowledge regarding the precise natural history of lumbosacral lipoma, in these lesions, the chances of developing neurological deficits increase with increasing age at presentation. Management of congenital spinal lipomas is challenging. Surgery remains the standard treatment. However, literature regarding the role of prophylactic surgery is scanty.

Keywords: Spine tumors, Spinal lipoma, Extramedullary spinal tumor, Outcome.

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Introduction

Congenital spinal lipomas are the most common form of occult spinal dysraphism, constitute a heterogeneous group roughly grouped together by their mutual characteristic of being skin covered and causing tethered cord syndrome [(1,2)]. In literature diverse terms have been coined for these lesions; lumbosacral lipomas which subdivided into lipomas of conus and filum lipomas. The term lipomyelomeningocele used for meningocele, which was associated with subcutaneous lipoma [(1, 3)]. Others categorized conus lipomas into dorsal, caudal, and transitional types [(4)]. In the existing study, congenital spinal lipomas adopted for: conus lipomas, lipomyelomeningocele, and filum lipomas. The real incidence of sacral lipoma is uncertain. In the literature, the only reported data concerns the incidence of this malformation versus myelomeningocele or other occult dysraphism, which only reflects

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personal recruitment. In this analysis, the present authors aim to approach the epidemiological features and clinical characteristics of spinal lipomas diagnosed at a single institution. Special emphasis devoted to analyze the clinical manifestations, radiological findings, outcome following surgery, and also to shed light on the role of prophylactic surgery in the management of these lesions.

Methods

This study was carried out by reviewing retrospectively the: patient's admission records, neuroimaging, operation records, and outpatient files and biopsy results were used to collect the data from all consecutive patients treated in single referral center. A total of 59 consecutively treated patients with congenital spinal cord lipomas associated with spinal dysraphism, confirmed by histopathological examination were analyzed. Patient data were extracted from the King Hussein Medical Center (KHMC) Primary Spinal Tumors electronic Database, which includes approximately 414-patients treated between 2006 and 2016. Collected data included demographic features; sex, age, size of the tumor, resection extent, histological alterations, local recurrence, perioperative morbidity, and complications were documented. Preoperative radiology reports were used to confirm tumor location and size based on largest single dimension. The extent of resection was based on surgical operative notes and post-operative imaging.

Operative Details

After a detailed preoperative diagnostic evaluation, patients were managed surgically via partial or total laminectomy proximal to the marked pathology through a posterior approach. The Cardinal aim in all cases was to decompress and to release tethering, as well as to reconstruct the dural canal. Lipomyelomeningocele sac when present was delineated on all sides. Lipomyelomeningocele sac or subcutaneous lipoma was completely excised. Conus lipoma was totally or near totally excised. Conus were reconstructed after excision of conus lipoma with multiple pial sutures. Filum terminale was identified and detethered in all the patients. Filum lipoma when present was excised. Dura closure was carried out primarily in a watertight fashion (Figure 1).

Fig 1a

Fig 1b

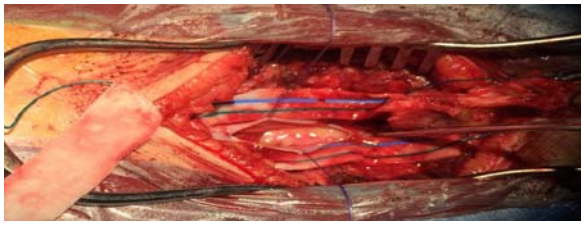
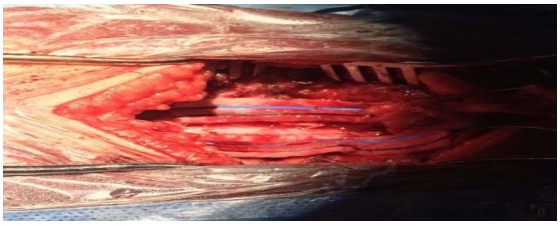


Fig 1c



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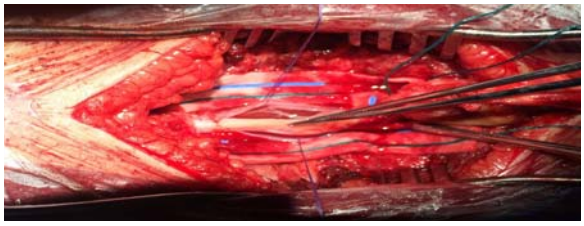


Fig 1e

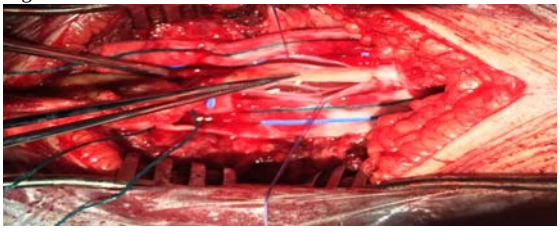


Fig 1: Intra-operative Images showing the steps of exploring and resecting a filum lipoma,(1-a. muscle dissection, 1-b. dura opening, 1-c lesion identification, 1-d. tumor removal, 1-e. dura closure)

Results

Our cohort consisted of 33-females (55.93%) and 26-males (44.06%), with ages ranging from 1-week to 12-years (mean: 3.45 ± 1.74 years).The congenital spinal lipomas were categorized into: 40-conus lipomas (17-terminal, 15-transitional, 8-dorsal) and 19-filum lipomas, including 11-patients who had lipomyelomeningocele. At the first operation, 21-patients (35.59%) were asymptomatic, and 38-patients (64.40%) presented with symptoms. The most common promoter for diagnosis was skin stigmas (46.26 %), followed by associated malformations (30.63 %), and symptoms (23.11 %). The presence of a dermal sinus tract or syrinx was the surgical indications influencer in the asymptomatic group included. Prophylactic surgery was undertaken in selected cases. The mean total follow-up for the group since the first diagnosis was 72.18 months (range: 17.20-120.30 months). In the initially asymptomatic group, 6-patients (28.57%) had late neurological deterioration. Of the 8-patients with asymptomatic conus lipomas, 3-cases (37.50%) developed sphincter dysfunction and motor problems at long-term follow-up. In the symptomatic group, 67.50% improved, 20% remained unchanged, and 12.50 % had late neurological deterioration. None of the 6-patients with symptomatic filum lipoma deteriorated postoperatively. Of the 32-patients with symptomatic conus lipomas, 65.62% improved, 15.62% remained stable, and 18.75% had late neurological deterioration. However, 74% had bladder dysfunction, 67% had neuro-orthopaedic deformity, and 45% had motor problems at long-term follow-up (Figure 2). Postoperative complications developed in 9 patients (13.55 %): seven transient local problems, 2 definitive urological deterioration.

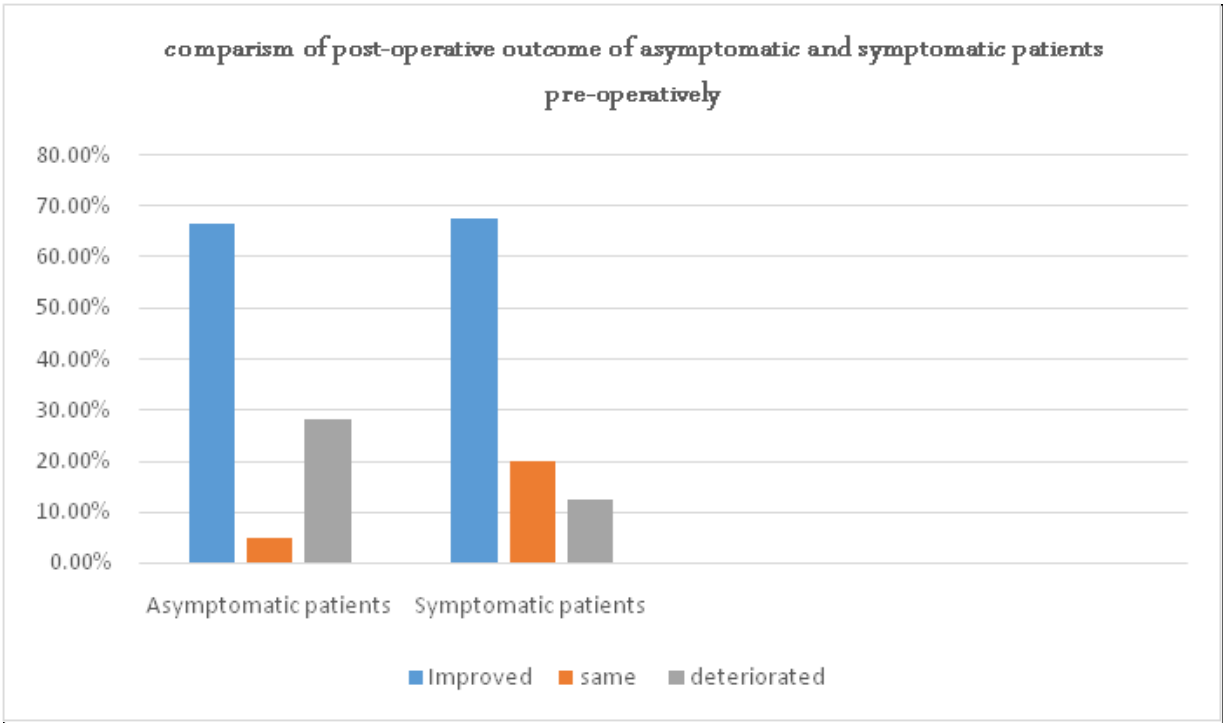


Fig 2: Diagram comparing the post-operative clinical outcome of symptomatic and asymptomatic patients

Discussion

Congenital spinal lipomas genetically complex malformations that are likely etiologically, morpho-genetically, molecularly, and genetically heterogeneous. Arise from an abnormal embryologic development [(5)]. Over the years, several theories have been proposed to explain the development of spinal lipomas, these have endured significant evolution as knowledge of normal neural development is attained [(2,4-7)]. Current theories are still only speculation, and are thus the center of persistent controversy. A contemporary theory that precisely interprets the surgical anatomy of

conus medullaris lipomas is that of McLone and Naidich[2,4,5,6]. These authors propose that an aberration disjunction, or separation of the neural tube from the surrounding ectoderm, occurs prematurely, leaving the neural plate open posteriorly and allowing the paraxial mesenchyme enters through the transient gap in the neural tube, and under the inductive influence of the neural placode, forms fat [2,7,8].The reported incidence of congenital spinal lipomas is in the range of 0.4-0.8/100.000.Reports state lipomyelomenigocele occurs in approximately one in 4000 births in the United States and females are at increased risk[1,3,9,10].In the current review we noticed slight female predominance 1.27:1, the exact incidence in our study is unknown. The initial clinical presentation of a child with lumbosacrallipoma is variable and depends largely on their age. lipoma might grow and apply mass effect on lumbosacral nerves and cause radicular symptoms. Intradural lipoma is usually manifested with neurological changes, causing compression of the conus medullaris elements. Clinical manifestations, that are associated with other stigmata, include the spinal dysraphism, skin stigmata, vertebral spine anomalies (scoliosis), extremity deformities, neurological deterioration at the level of the lower spinal cord, sphincteric dysfunction[11,12]. Usually, there are just cosmetic changes without neurological disturbances during pediatric growth. The most common promoter for diagnosis in this review was skin stigmas (46.26 %), followed by associated malformations (30.63 %), and symptoms (23.11 %). Neuroimaging evaluation is the keys to define the anatomical and pathological features of the lesion, whenever occult spinal dysraphism is assumed based on clinical presentation. Ultrasonography is a useful tool to apply even parentally, on an infant suspected of having lipomyelomeningocele[13]. Plain x-ray films almost equivalently show abnormal findings although interpretation is difficult due to lack of ossification. Spina bifida (dorsal midline fusion defects) and a widened spinal canal are the most common findings encountered. Computerized tomography scanning provides an excellent resolution of the anomaly; however, this modality is invasive, and requires exposure to radiation. Nowadays, magnetic resonance imaging (MRI) has evolved to become the gold standard imaging modality for dysraphic conditions. Lipomatous tissue demonstrates a high signal on T 1 -weighted MRI mages and T 2 – weighted MR images (Figure 3). Contrast material administrations not necessary [14,15]. In the current study, once infants presented with obvious skin tag or developed neuro-manifestations related to his condition we conducted MRI. While it is broadly accepted that congenital spinallipomas are anatomically stable lesions, the growth of extra- and intraspinal lipomas is documented[16]. Lumbosacral lipomas are not neoplasms, thus the surgical goal is not total removal of the lipoma, but rather the protection of neurological function and the prevention of delayed neurological decline attributed to a tethered cord [17]. Surgical planning in a patient with a newly diagnosed lumbosacral lipoma, is a sophisticated process; we should consider the type of lipoma and whether the patient is symptomatic [17,18]. In all symptomatic patients and in patients with asymptomatic filar lipomas with lower-lying coni, the decision to operate is straightforward. However, the decision to operate in asymptomatic patients with conus lipomas is controversial. Some surgeons promote, regardless of symptoms, to do prophylactic surgery for all patients, while others propose that surgical option to be withheld until symptoms develop, because conus lipomas, especially transitional-type lipoma and lipo-myelomeningocele, have relatively high surgical morbidity.[17-20]. In this analysis, the surgical management motivator in the asymptomatic group was the presence of skin stigmata. Prophylactic surgery was undertaken in selected cases. Results in asymptomatic group, 6-patients had late neurological deterioration. In the symptomatic group, 67.50% improved, 20% remained unchanged, and 12.50 % had late neurological deterioration. None of the 6-patients with symptomatic filum lipoma deteriorated postoperatively. This study revealed that filum and conus lipomas have similar clinical manifestations, but diverge in

their result following surgery. Filum lipomas are tend to be more 'benign', for which surgery is safe and effective. Lipomas of conus are more demanding to cope. Prophylactic surgery might provide some protection from future neurological deterioration in asymptomatic patients. When symptomatic, conus lipoma surgery is effective in preventing further worsening. Improvement in neurological function can occur, but few patients return to normal neurological function, and pre-existing sphincter dysfunction is not considerably improved by surgical intervention.

Fig 3: Magnetic Resonance Imaging: (3-a): T-1 weighted sagittal cut, (3-b): T-2 weighted sagittal cut, (3-c): axial cuts, showing a filum lipoma.

Fig 3a

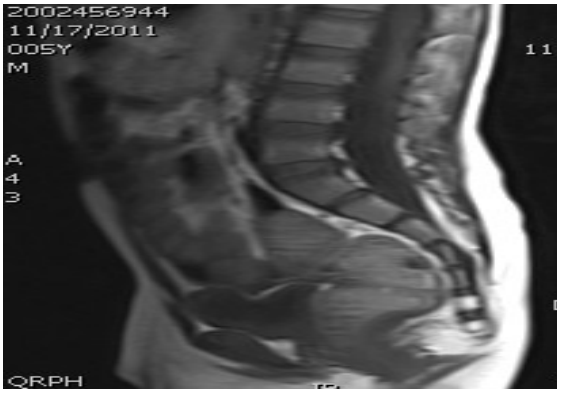


Fig 3b

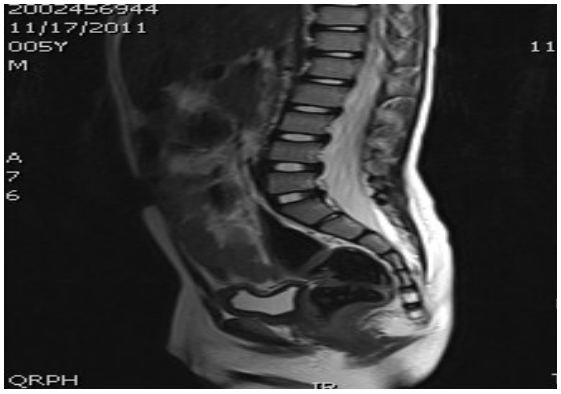
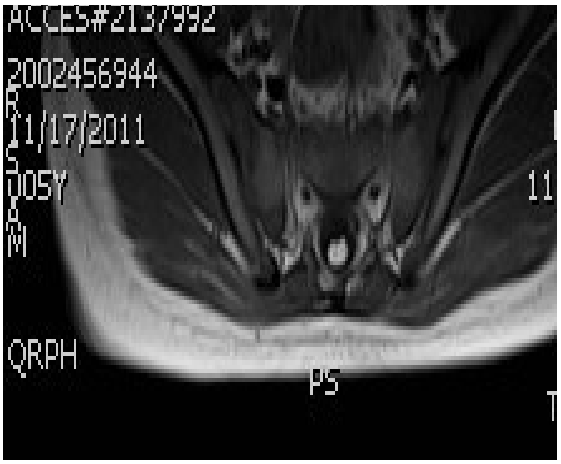


Fig 3c



Conclusion: Despite the lack of knowledge regarding the precise natural history of lumbosacral lipoma, in these lesions, the chances of developing neurological deficits increase with increasing age at presentation. Management of congenital spinal lipomas is challenging. Surgery remains the standard treatment. Spinal lipomas can cause progressive neurological deficits irrespective of spinal untethering surgery. However, literature regarding the role of prophylactic surgery is scanty.

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