Lateral tethering intraspinal lipoma with scoliosis

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Abstract The authors present an unusual case of an intradural lipoma at the thoracolumbar level causing lateral tethering of the spine, with complicated scoliosis. A 6-year-old girl was admitted with progressive change in posture detected by her parents. Thoracolumbar scoliosis was detected by physical examination and X-ray studies. Spinal magnetic resonance imaging revealed an intradural lipoma and right lateral tethering of the spine at the upper lumbar level. After surgical release of lateral tethering and resection of the lipoma, fusion was performed by the orthopedic surgery team. Radiological and intraoperative findings of this extremely rare case are discussed.

Keywords Intradural lipoma · Occult spinal dysraphism · Spinal scoliosis · Tethered cord syndrome

Introduction

The tethered cord syndrome (TCS) is a stretch-induced functional disorder of the spinal cord with its caudal part anchored by an inelastic structure [31]. As the spinal cord tethering can be caused by various pathologies and at any level and side of the spine, the term “tethered cord” has

broadened to include cord tethering from many different etiological conditions in the literature [3, 5]. Many theories were suggested to explain the pathophysiology of the clinical syndrome of the tethered cord but recently the most wide excepted biochemical phenomenon underlying cord traction is “traction-induced hypoxia” and neuronal membrane stretch with “loss of transmembrane ion homeostasis and electrical activity depression” [25, 29]. Surgical intervention is mainly based upon the underlying or associated pathologies with tethered cord. In filum terminale tethering cases, resection of the filum terminale in symptomatic patients is simple and efficient modality of treatment [25]. Tethered cord associated with caudal lipoma, lipomyelomeningocele and sacral myelomeningoceles is similar to that with tethered spinal cord [5, 12, 31]. Dorsal and transitional lipomas or lipomyelomeningoceles and all the myelomeningoceles require meticulous and clean dissection for cord untethering and correction of the abovementioned pathological conditions around the dorsal cord [31].

In this report, authors present an unusual case of lateral spinal cord tethering caused by intradural lipoma at the level of upper lumbar vertebra. To our best knowledge, no similar case about lateral tethering lipoma was reported in the English literature. Radiologic and clinical features and intraoperative findings are discussed.

Case report

A 6-year-old otherwise healthy girl, with normal medical history, brought to medical attention due to progressive posture abnormalities developed in a 1-year period. Physical examination revealed neither neurological signs nor skin stigmata. Spinal X-rays showed thoracolumbar

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Fig. 1 **a** Plain radiography shows thoracolumbar scoliosis and multiple vertebral structural abnormalities. **b** Coronal and **c** axial T1-weighted MRI reveal scoliosis and intradural right-sided lipomatous mass lesion (arrow)

scoliosis to the right side, with structural abnormalities of the vertebral bone in this level (Fig. 1a). Magnetic resonance imaging (MRI) revealed scoliosis and intradural right-sided lipomatous mass lesion located adjacent to spinal cord (Fig. 1b, c). Urodynamic tests were normal. Single stage surgery for releasing of the tethered cord and correction of the scoliosis was planned. After exposure of the thoracolumbar spine, laminectomy was performed at the level of lipoma. Dura was found to be expanded and

Fig. 3 Post-operative P-A (**a**) and lateral (**b**) plain radiographs

tense with extradural component of the lipoma (Fig. 2a). After opening of the dura, the spinal cord and the lipoma located at the right lateral side were exposed (Fig. 2b). The spinal cord was found tight and laterally strained by the lipoma. Lipoma was carefully dissected under the operating microscope and its pial and dural vascular attachments were coagulated and separated (Fig. 2c). After removal of the lipoma, the spinal cord was released and showed significant relaxation and the dura was sutured in water-tight fashion. Fusion with transverse process decortication and spongy bone allograft was performed by the orthopedic surgery team in the same stage (Fig. 3). Post-operative

Fig. 2 Intraoperative views of our case. **a** Dural exposure after laminectomy with the extradural component of the lipoma. **b** The intradural component of the lipoma and its relation to the dura and the spinal cord. The portion of the lipoma attached to the lateral dura and causing tethering of the spinal cord (arrow). **c** Resection of the lipoma and untethering the spinal cord

period was uneventful and the patient was mobilized with thoracolumbar brace for the first 6 weeks. The patient was discharged with no complications and neurologic deficits.

Discussion

Intraspinal lipomas are responsible for 25–30% of TCS seen in children [22] and accounting for 1% of all intraspinal tumors [7, 10, 16, 26]. Finn et al. [9] suggested a practical and clinically relevant classification of spinal lipomas into three broad categories: lipomas of the conus medullaris, lipomas of the terminal filum, and “subpial” lipomas. The latter group is not unique to the lumbosacral region. Lipomas of conus medullaris are further classified according to Chapman [6] into dorsal, caudal, and transitional lipomas. Intraspinal lipomas are most commonly found in the lumbosacral area (90%) [1, 17, 23, 26]. Excluding the lumbosacral spine, the reported distribution of lipoma was thoracic in 32% of cases, cervicothoracic in 24%, and cervical in 13% [18]. In McLone et al. series [19], 62% of cases were midline, 33% left side and 5% right side. Neurologic deficits were found in 50% in children younger than 6 years old [11]. In children, urologic and orthopedic problems are more pronounced. In some series, sphincter dysfunction was found in 30–50% [2, 19, 22, 27]. After puberty, sexual dysfunction was found in 25% of the patients. CT findings of lipoma include very low density, with a value of -100 Hounsfield units [27]. MRI findings are also characteristic, being hyperintense on both T1- and T2-weighted images, with more pronounced hyperintensity with fat suppression sequences [26]. The preoperative MRI indicated that the margin of the lipoma was well demarcated from normal spinal cord; however, the margin between the lipoma and the normal spinal cord maybe indistinct. As spinal lipoma is an intramedullary tumor, total removal may injure the spinal cord leading to impairment of spinal function [26]. Clinical symptoms induced by spinal lipoma usually resulted from cord compression within the spinal canal rather than direct cord invasion by the tumor and due to secondary tethering of the spinal cord [20].

Yamada et al. [31] defined the TCS as a stretch-induced functional disorder of the spinal cord with its caudal part anchored by an inelastic structure. As the spinal cord tethering can be caused by various pathologies and at any level and side of the spine the term “tethered cord”, Bui et al. [3] had broadened this definition to include cord tethering from many different etiological conditions in the literature. The diagnosis and treatment of this member of the occult spinal dysraphism family are controversial and can be difficult. Recently, the incidence of occult spinal dysraphism and TCS increased, probably due to higher rates of incidental detection with the more wide use of MR

imaging, greater clinical awareness, and a continued broadening characterization of the disease process [25]. The true incidence, however, is unknown, and little is known of its true prevalence. Female to male ratio was 2:1 for occult spinal dysraphism in many series [6], but it is not certain for TCS.

Many theories were suggested to explain the pathophysiology of the clinical syndrome of the tethered cord but recently the most wide excepted biochemical phenomenon underlying cord traction is “traction-induced hypoxia” and neuronal membrane stretch with “loss of transmembrane ion homeostasis and electrical activity depression” [25, 29]. Changes in oxidative metabolism can be indicated by recording shifts in the reduction/oxidation (redox) ratios of the components of the mitochondrial electron transport chain. This shift demonstrates oxidative metabolism because oxygen consumption and ATP production are tightly coupled to mitochondrial electron transport [8, 24]. In cat models, shifts toward a reduction in cytochrome *a,a3* occurred as the spinal cord was stretched with weights [31].

The changes in mitochondrial redox ratios and interneuron potential changes that occurred in animal models of spinal cord traction were similar to those observed during hypoxemia and ischemia [32]. Also glucose consumption of the stretched spinal cord was found to have an essential role in the pathophysiology of the TCS. With traction, decreased glucose consumption, indicated by impaired 2-deoxyglucose metabolism, was consistent with cytochrome reduction, which correlated with the degree of the reversibility of neurological dysfunction and glucose metabolism is altered in parallel to the redox alterations in cytochrome *a,a3* [28].

Patients with symptomatic TCS can present with a wide variety of signs and symptoms in combination with cutaneous, orthopedic, spinal, anorectal, and urological abnormalities, as well as pain, with the cutaneous stigmata being the most common clinical presentation (59%) [3].

Neuroimaging is used in clinically suspected cases of TCS. Ultrasonography is a useful tool in young infants with ability to obtain a dynamic view without having to submit a young child to irradiation or sedation. Plain X-ray films can help in identifying associated bone abnormalities such as lamina defects and scoliosis. Nowadays, MRI is the best modality for visualizing spinal cord and filum terminale pathologies, level of tethering and lipomatous lesions. Sagittal T1- and T2-weighted images are best for localizing the level of the conus, whereas T1-weighted axial MR images are better for identifying fat within the terminal filum and for measuring the diameter of the filum. Computed tomography scanning or CT myelography can be used if MR imaging is not available, with good bone window and definition of the level of conus medullaris, but not used routinely due its invasiveness and difficulty to perform upon young infants.

The main goals in surgical treatment of TCS are to improve or stabilize deficits in the symptomatic patient and to prevent future deficits in the asymptomatic patient. Many neurosurgeons prefer to intervene asymptomatic patients with a low-lying conus and a fatty filum (with or without occult spinal dysraphism) to prevent future deterioration [3].

Surgery in these patients can be controversial because of the broad spectrum of pathological involvement [13, 21]. Observation is advocated for symptomatic patients with normally positioned conus, no occult spinal dysraphism, and normal filum and search aggressively for other causes for the symptoms and for asymptomatic patients with a normally positioned conus and normal filum but who have multiple occult spinal dysraphism [3]. Because the pattern of deterioration seemed unpredictable and symptoms in many patients were not reversed by surgery, many surgeons recommended that surgery be undertaken in infancy prior to the development of symptoms [4, 14, 15]. The rationalities were that cord tethering and the intramedullary lipomatous mass were the principal causes of deterioration that untethering and lipoma debulking could be achieved without significant acute or long-term functional impairment, and that re-tethering and subsequent deterioration could be prevented.

Surgical intervention is mainly based upon the underlying or associated pathologies with tethered cord. In the first category, the spinal cord dysfunction associated with caudal lipoma, lipomyelomeningocele and sacral myelomeningoceles is similar to that with tethered spinal cord [31]. In the case of lipomeningomyelocele, untethering surgery consists of clearly detaching conus tip from the fibroadipose sac wall [12, 31]. In filum terminale tethering cases, resection of the filum terminale in symptomatic patients is simple and efficient modality of treatment [3].

In the second category, dorsal and transitional lipomas or lipomyelomeningoceles and all the myelomeningoceles require very careful and clean dissection for cord untethering and correction of the other pathologies [31]. If the main consideration of the surgery is the untethering procedure, neurological improvements are predictable [30]. In the first group of the third category, filum resection for prevention of the progressive scoliosis and lumbosacral lordosis is still controversial. In the second group, prophylactic surgery is not recommended by some, because various complications are anticipated [8] and the procedure is not always simple. In patients with scoliosis and tethered cord, it is preferred to perform the untethering at the time of the scoliosis repair. This can prevent the child from undergoing two operations and general anesthesia. The untethering is done before the fusion, and an effort is made to keep the two incisions separate [3].

Our case presents an unusual case of intradural lipoma. To our best knowledge, it is the first case of intradural

lipoma causing lateral tethering of the spinal cord at the level of upper lumbar vertebra, and no similar case was found in the English literature. The patient complaints were back pain and progressive posture abnormalities due to developed scoliosis which was not responsive to conservative treatment. There were no neurologic deficits and skin stigmata. In our case, there were concomitant congenital deformities of the vertebral bodies. These deformities along with the lipoma may be the result of mesodermal disorganization during the primary neurulation leading to two separate unrelated pathologies. However, the intraoperative finding of the tight spinal cord at the site of the lipoma which showed laxation after resecting the lipoma is more suggestive that the vertebrae were under lateral straining effect. Even in previous presence of the congenital deformities of the vertebral bodies, we believe that the straining of the tethering lipoma—noticed intraoperatively—had the important effect of the progression of the scoliosis during growth of the patients.

Conflict of interest statement None of the authors has any potential conflict of interest.

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