Lumbar teratoma presenting intradural and extramedullary extension in a neonate

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Abstract

BACKGROUND CONTEXT: The association of teratomas and spinal malformations such as spina bifida, partial sacral agenesis, hemivertebrae, and diastematomyelia has been described in the literature. Reported cases, however, are mainly presacral or sacrococcygeal with an extremely rare presentation of intradural extension.

PURPOSE: A case of lumbar teratoma with an intradural extension and extramedullary component and the clinical outcome following surgical treatment are reported.

STUDY DESIGN/SETTING: To our knowledge, among the reported teratoma cases with an intradural extension and extramedullary component, our case has a distinguishing feature regarding the involvement of the lumbar spine. It is also the first case, showing no neurological deficit during the postoperative period.

METHODS: A full-term, female infant presented with a 30×30×10 mm lumbar mass covered with normal skin. The mass contained an irregular, bone-like, hard and mobile material accompanying cystic components. Magnetic resonance imaging revealed a total closure defect of the first and second lumbar laminae and a subcutaneous mass with intradural extension. The lesion was found to penetrate the dural sac through an extended exposure from T12 to L3. There was no firm attachment of the intradural, extramedullary component of the lesion. Total removal of the tumor was achieved.

RESULTS: The patient was discharged on day 7 without any neurological deficit or sign of hydrocephalus. The pathological examination showed a benign teratoma containing mature cartilage, muscle, adipose tissue, and glandular tissue. Follow-up at 2 years showed no recurrence or neurological deficit and a normal sphincter tone. Urodynamic evaluation was within normal limits.

CONCLUSION: Accompanying a spinal dysraphic state, the mature teratoma in our case may support the idea of a tumor actually arising from a dysraphism and growing outward to produce the mass. © 2006 Elsevier Inc. All rights reserved.

Keywords: Teratoma; Intradural extension; Lumbar, Spinal

Introduction

The clinical picture of childhood teratomas most often presents with diverse anatomic locations extending from brain to the sacral area and the gonads [1]. Reported cases are mainly presacral or sacrococcygeal teratomas [1–4]. These neoplasms are distinguished among other childhood tumors for their composition of tissue elements foreign to the organ or anatomic site of origin and biological behavior [5]. The association of teratomas with dysraphic lesions in the thoracic and lumbosacral regions is rarely described [6]. Sacrococcygeal level teratomas have been reported that encroach into the spinal foramina with associated extramedullary extension. Intradural extension of such teratomas with attachment to the spinal cord is even more infrequent [4,7–10]. This association is said to support the suggested
pathogenesis theory of disembyrogenic origin of these tumors [10].

The case presented herein is the first report of a lumbar teratoma with an intradural extension and extramedullary component.

Case report

A full-term female infant (3,100 g, head circumference = 33.4 cm) was admitted with a 30×30×10 mm lumbar mass covered with normal skin. The mass contained an irregular, bone-like, hard and mobile material with accompanying cystic components. There was no in utero testing to make a prenatal diagnosis, and the infant was born by cesarean section. The physical and neurological examination was normal, except for the mass and alpha-fetoprotein level, which was over 4,000 mg/dL.

Magnetic resonance imaging (MRI) revealed a total closure defect of the first and second lumbar laminae and a subcutaneous mass with intradural extension (Fig. 1). Surgery was performed in collaboration with a pediatric neurosurgeon when the baby was 4 days old. The patient underwent a partial T12–L3 laminectomy. The lesion was found to penetrate the dural sac through an extended exposure from L3 level up to T12. After dural opening, a reddish tumor with yellow areas was observed, which showed intradural extension and 30×30×10 mm size. There was no firm attachment of the intradural, extramedullary component of the lesion. Separation of the capsule and total removal of the tumor was possible. The dura mater was closed in a watertight manner. The histopathological diagnosis was a benign teratoma containing mature cartilage, muscle, adipose tissue, and glandular tissue. No immature or malignant cells were present (Fig. 2). The postoperative course was uneventful, and the patient was discharged on day 7 without any neurological deficit or sign of hydrocephalus.

The patient was monitored in the outpatient clinic and showed no recurrence or neurological deficit and a normal sphincter tone at 2 years of age. Urodynamic evaluation and alpha-fetoprotein levels were within normal limits.

Discussion

Spinal teratoma is an extremely rare entity and usually exhibits a benign histopathological profile. Only a few series have been described in the literature regarding the association of teratomas and spinal malformations such as spina bifida, partial sacral agenesis, hemivertebrae, and split cord malformations. Reported cases, however, were mainly presacral or sacrococcygeal with an extremely rare presentation of intradural extension, and patients were mostly young adults and males [9–13].

Less frequent is the presence of pediatric sacrococcygeal teratomas showing intraspinal extension as noted by Gross et al., who reported two cases in their series covering 40 sacrococcygeal teratomas [14]. Donnellan and Swenson reported an additional three cases with intraspinal extension among 54 sacrococcygeal teratomas [3]. Aschcraft and Holder reported spinal involvement in hereditary presacral teratoma [2].

Teratomas are composed of remnants from all three primitive germ layers and are classified into three groups, mature, immature, and malignant teratomas. Benign lesions are mainly composed of mature elements such as cartilage, glands, muscles, and adipose tissue as described in our case. The traditional view for the origin of teratomas is that early in embryogenesis, primordial germ cells become misplaced because of an alteration during cell migration. The pathogenesis of an intraspinal teratoma, however, remains controversial. In his written comment about Ribeiro et al.’s article, Leslie Sutton states that the pleuripotential embryonic caudal cell mass of the spinal cord might differentiate along various cell lines if it persists as part of dysraphism [4]. Accompanying a spinal dysraphic state, the mature teratoma in our case may support the idea of a tumor actually arising from a dysraphism and growing outward to produce the mass.

Intradural spinal cord tumors are categorized as intramedullary or extramedullary. Intradural extension of a teratoma presenting with attachment to the spinal cord,
however, is extremely rare. Only two cases were reported presenting with intradural extension and attachment to filum terminale. The initial one described by Teal et al. [8] was an immature teratoma with intradural extension, and the subsequent one was a mature teratoma [7]. Both of these cases were reported to have no extramedullary components.

Ribeiro et al. reported a sacrococcygeal teratoma in a neonate with intradural and extramedullary extension [4]. It was reported as the first case of teratoma that also had an extramedullary component ascending up to the dorsal level. To our knowledge, our case has a distinguishing feature regarding the involvement of the lumbar spine among the reported pediatric teratoma cases with an intradural extension and extramedullary component. These tumors may be asymptomatic until adulthood and then become a problem, as has been the case in the reports of Fernandez-Cornejo et al. and Arai et al. [9,10].

Monajati et al. pointed out the accuracy of preoperative diagnostic MRI in spinal tumors [15]. In this study they adequately demonstrated all three main components of a benign lumbar intradural teratoma. On the contrary, MRI or computed tomographic scan may not always accomplish a definite diagnosis [10].

Treatment for intramedullary tumors is radical surgery, because it not only constitutes a prognostic factor but also is diagnostic in definitive approach. Extensive laminectomies are said to cause spinal instability in 30–80% of pediatric cases, and a concomitant laminoplasty is advocated [10]. Poeze et al. [16] have reported better results in adults with regard to improvement after surgical treatment of intramedullary teratomas, whereas only a quarter of pediatric cases showed improvement. We did not observe any spinal instability after laminectomy in our case. The patient is still young and there may still be the possibility of deformity as she grows.

It is also the first case reported showing no neurological deficit during the postoperative period. The fact that the diagnosis of this infant was made on the basis of the presence of a mass and there was no preoperative neurologic deficit may explain the favorable result. The presence of preoperative deficit may mean that the neurological structures are susceptible to postoperative compromise because of preexisting nerve problems. The timing of our diagnosis and surgical approach may also have influenced a rather satisfactory result.

The differential diagnosis of a congenital mass in the subcutaneous space behind the spine in the lumbar or lumbosacral region should include lipoma, meningocele, hemangioma, and teratoma. MRI has proved to be useful in the diagnosis of these lesions. The choice of treatment deserves collaborative work among pediatric surgeons and pediatric neurosurgeons. It is certain that such cases merit special emphasis on preserving the neural elements.

References
One Hundred Forty-five Years Ago in Spine

In 1860, Friedrich Goll [1] published a treatise on anatomy that included description of what has been called “Goll’s Column”—the posterior column of the spinal cord.

Reference