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CASE REPORT

Enlarging teratoma syndrome

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Abstract

Introduction Teratomas are derived from all three germ layers and make up 3% of all childhood tumors. They are histologically classified as mature or immature. We present a case that was operated on when 30 days old for a sacrococcygeal mature teratoma and then showed long segment involvement in the thoracolumbar region 9 months after the surgery. The MRI (magnetic resonance imaging) showed a mass starting at the thoracal 4 level and extending to the lumbar 3 level with significant spinal cord compression in the extradural space.

Result The laminae between thoracal 4 and lumbar 3 levels were removed en bloc at a single surgical session and laminoplasty was performed after tumor resection. We also removed the tumor growing into the extrapleural space at the thoracal 5, 6, and 9, 10, 11, 12 levels using the costotransversectomy procedure.

Conclusions We emphasize with this case that mature teratomas can show aggressive growth following surgery and that the development of spinal deformities can be prevented with laminoplasty.

Keywords Mature teratoma · Epıdural tumor · Pediatric spine surgery

Introduction

Teratomas are tumors that are derived from all three germ layers and include ectodermal, mesodermal, and endodermal components. They are histologically classified as mature or immature depending on the differentiation of their cellular components [7]. Teratomas are derived from pluripotent cells and can therefore include hair, teeth, fat, intestinal mucosa, cartilage, bone or skin. They usually have a midline or paraxial location in children and are mostly paracoccygeal [2]. Teratomas make up 3% of all childhood tumors [16]. Extensive epidural teratomas are quite rare. Our case was a child with an extensive epidural teratoma that had extended to the peripleural area and along the spinal foramina and involved the vertebral corpuses. MRI images revealed significant spinal cord compression starting at the thoracal 4 level and extending to the lumbar 3 level and causing intraspinal extradural paraplegia. The lamina was lifted en bloc and costotransversectomy was performed for removal of the epidural and peripleural tumor.

Case presentation

The patient whose sacrococcygeal teratoma was noticed in fetal ultrasonography at the 25th gestational week and who was born at the 39th week of pregnancy with a birth weight of 3700 g underwent surgery 30 days after birth for the swelling in the sacrococcygeal region. Pathology evaluation of the mass removed from the surgical area revealed mature teratoma. Following this pathology evaluation, the patient was referred to the Pediatric Hematology-Oncology Department and chemotherapy was started. However, the patient developed bilateral leg weakness following 4 cycles of treatment and was referred to our clinic. The preoperative blood alpha-fetoprotein level was 66. The patient was brought back for increasing weakness of both legs in the last 4 months and physical examination revealed complete flaccid paraplegia under the T5 vertebral level. Spinal MR showed an intraspinal extradural space mass at the thoracal and lumbar levels, between T4 and L3 (Fig. 1). This mass showed bilateral intraspinal foramina involvement at multiple levels of the thoracal region together with extrapleural and corpus involvement at T5, T6, T11, T12 (Fig. 2). The spinal cord was significantly compressed.

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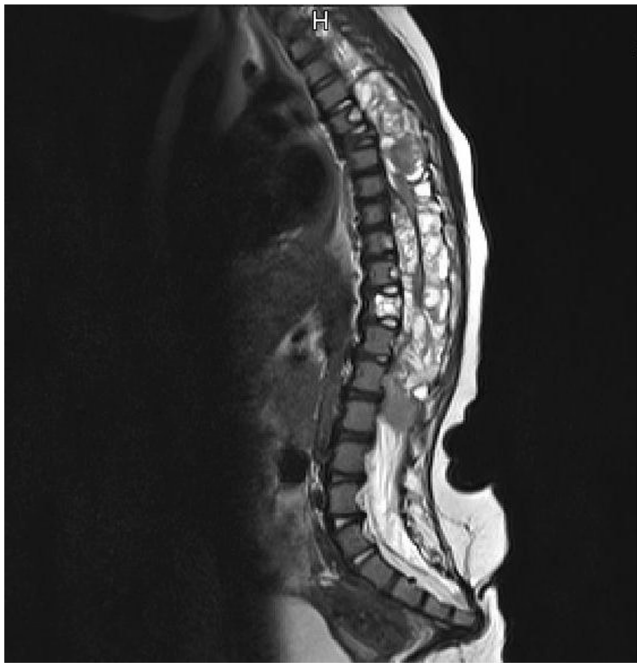

Fig. 1 Intraspinal extradural mass lesion extending from the thoracal 4 level to the lumbar 3 level

A wide midline skin incision from the upper thoracal region to the lower lumbar region was made. The thoracal and lumbar laminae were lifted en bloc together with the flavum using a bone cutter (Fig. 3). Once the laminae were lifted, the extradural mass was seen and paravertebral lesions at multiple peripleural levels were encountered. The peripleural mass was exposed with costotransversectomy and was removed without damaging

Fig. 2 Multiple level coronal MR sections showing the extension of the intraspinal foraminal and extrapleural mass

Fig. 3 En bloc removal of the laminae

the pleura by careful dissection. Debulking was performed at the epidural space with bipolar coagulation while the intraforaminal and vertebral corpus lesions were aspirated with an ultrasonic aspirator to perform subtotal mass excision. Following en bloc removal of the laminae, laminoplasty was performed to prevent any future deformities. The pathology result was mature teratoma. The third postoperative month magnetic resonance images showed no tumor regrowth. There was minimal rest tissue without any deformity such as kyphosis (Fig. 4). The patient was transferred to the Pediatric Hematology-Oncology Department postoperatively.

Fig. 4 Postoperative third month contrast sagittal MR imaging

Discussion

Teratomas are tumors that include elements from all three germ layers: ectodermal, mesodermal, and endodermal. They are histologically classified as mature or immature depending on the differentiation of the component [2]. Sacrococcygeal teratomas are seen once every 35,000–40,000 live birth. Despite various theories, the etiology of teratoma is unclear. The classical model states the reason as incorrect positioning of the primordial germ cells from the primitive yolk sac [8, 9, 12]. It has been said that the mesenchymal progenitor cells in the caudal cell mass that are remnants of the primitive streak and Hensen's node have a pluripotent structure and can differentiate into tissues derived from the three germ layers and result in teratoma development [8, 9]. Teratomas can cause perinatal death due to factors such as heart failure, tumor hemorrhage, and premature birth [1, 10]. Large teratomas are hypervascular enough to potentially cause fetal anemia and heart failure [10].The main prognostic factors in sacrococcygeal teratomas are the histological type and recurrence. The recurrence rate is 0 to 26% (mean 10%) in mature teratomas while it is higher at between 12 and 55% (mean 33%) in immature teratomas [17]. Chemotherapy decreases the rate of recurrence.

As regards spinal teratomas, surgical excision is needed to confirm the diagnosis and determine the histological subtype. Pure mature teratomas can be treated with total surgical excision. Teratomas with germ cell elements or malignant histological features require radiotherapy targeting the malignant cells in addition to adjuvant chemotherapy [6]. Total resection is recommended but can be difficult as the tumor is adherent to neural tissue. The difference in recurrence following subtotal and total resection is thought to be insignificant due to the slow-growing nature of mature teratomas [13]. Microscopic pieces remain even after total resection due to the adherent nature of the tumor and can increase the risk of recurrence. Chemotherapy and radiotherapy can cause proliferation in a benign piece to increase after subtotal surgical resection in some mixed teratomas. This is known as the enlarging teratoma syndrome and a second surgery may be needed for decompression [6]. When treating immature teratomas, the additional benefits and known harms of chemotherapy and radiation are not well established, especially in pediatric patients, and one must therefore be careful. Some authors support an aggressive surgical approach by avoiding adjuvant treatment until there is recurrence [15]. The results of using radiotherapy as the final solution in spinal teratomas are not satisfactory [6].

Local recurrences are generally seen after sacrococcygeal teratoma surgery but distant metastases such as to the vertebra and liver have also been reported [4].

It is rare to encounter a pathology diagnosis of mature teratoma following sacrococcygeal surgery and then detect long segment involvement in the spinal canal after chemotherapy. Tumor markers such as alpha-fetoprotein are normal or not significantly elevated in the enlarging teratoma syndrome. This idea was first proposed by Logothetis et al. [11]. The cause of the enlarging teratoma syndrome is unknown but there are two theories. The first one states that chemotherapy only destroys immature cells while the benign teratoma material survives. The second theory is that chemotherapy alters the cellular kinetics from totipotent malignant germ cell to benign mature teratoma [3, 5, 14]. The low alpha-fetoprotein levels, the administration of chemotherapy following the first sacrococcygeal teratoma surgery, and the pathology results of mature teratoma in this case resulted in the diagnosis of spinal enlarging teratoma syndrome.

We treated the case surgically with subtotal resection. We performed laminoplasty to prevent potential future spinal deformities as long segment spinal surgery had been necessary. We did not detect any spinal deformity during follow-up.

Compliance with ethical standards

Conflict of interest There are no reported conflicts of interest for all authors.

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