CASE REPORT

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Sacrococcygeal Teratoma with Split Spinal Cord Malformation

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The incidence of diastematomyelia associated with teratoma is extremely rare. We present a case of sacrococcygeal teratoma in a neonate with split spinal cord malformation(SSCM). Magnetic resonance imaging(MRI) showed a heterogenous mass lesion with cyst in the sacrococcygeal region and multiple spinal anormalies (diastematomyelia, tethered cord, hydromyelia, and hemivertebrae). The mature teratoma was confirmed on histopathological examination. In SSCMs, the potential for coexisting congenital anomalies at separate levels of the spinal cord must be considered in radiological investigations.

KEY WORDS : Teratoma · Split spinal cord malformation · Multiple spinal anomalies.

Introduction

A teratoma contains recognizable mature or immature elements representative of more than one germ layer. Teratomas have their origin in totipotential cells, which are normally present in the ovary and testes and are sometimes abnormally present in sequestered midline embryonic rests[14]. Although sacrococcygeal teratomas(SCT) are rare, they are the commonest tumors in the neonatal period[21].

Diastematomyelia, a complete or imcomplete sagittal division of the neural axis into halves, is usually accompanied by a number of other malformations including cutaneous changes, skeletal, especially vertebral anomalies, hydromyelia, meningocele or myelomeningocele. And it is defined as split spinal cord malformation(SSCM)[5,8,22]. However, SCT associated with diastematomyelia is extremely rare. We report a rare case of SCT with multiple spinal anomalies incluing diastematomyelia, tethed cord, hydromyelia, hemivertebrae with review of the literature.

Case Report

A newborn girl was admitted to our hospital because of a large midline dorsal mass. She was delivered at 38-weeks' gestation to a 26-year-old philippine mother who had had a previous successful pregnancy. Delivery was by cesarean section. Birth weight was 2.7kg. Her appearance was consistent with that of an infant delivered at 38 weeks. The head, heart, peripheral pulses, and abdomen were normal. She moved all four extremities spontaneously. The external genitalia were normal. There was an approximately 6×5×3cm mass on the dorsal midline involving sacrococcygeal area (Fig. 1). Magnetic resonance imaging(MRI) showed a heterogenous mass with cystic and solid component (Fig. 2), low-lying tethered cord (Fig. 3A), hydromyelia (Fig. 3B), and diastematomyelia (Fig. 3C).

On the 5th day of life, surgical removal of the mass was carried out. Intraoperatively, the mass was composed of solid

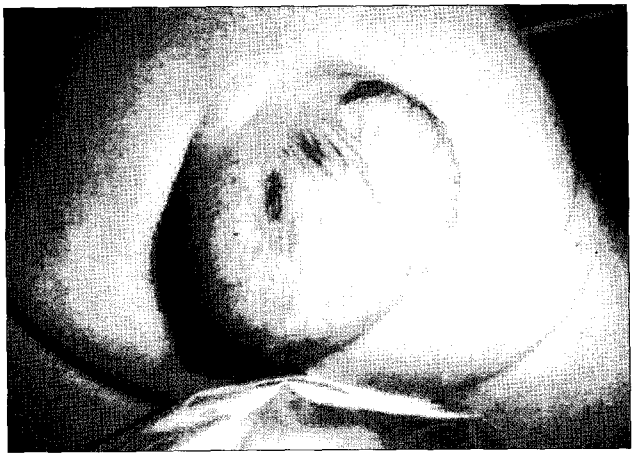

Fig. 1. The photograph demonstrates a large, predominantly solid mass at sacrococcygeal area.

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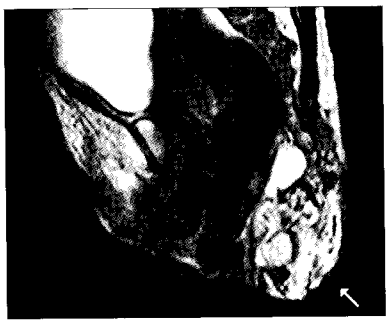

Fig. 2. Spinal magnetic resonance image shows heterogenous mass (arrows) with solid and cystic component in the sacrococcygeal area and the mass appears to be heterogenous signal intensity on sagittal T2-weighted image.

portions mostly. The mature teratoma was confirmed on histopathological examination that showed the tumor made up of complex tissue with a mixture of intestinal epithelium, squamous epithelium, immature cartilage, and bone (Fig. 4). The postoperative course was uneventful, and she was discharged after 25 days without neurological deficit. And she has no neurologic deficit now. Posto-perative follow up MRI after 7 months showed a postoperative cystic change in previous operation site (Fig. 5) and no interval changes in other anomalies including low-lying tethered cord, hydromyelia, and diastematomyelia.

Discussion

The SCTs are rare tumors as is evidenced by the small number of patients collected over a period of several decades in the most series[21]. The sacrococcygeal area is the most frequent site of teratoma in infancy, occuring in 1 of 35,000-40,000 live birth[1,21]. There is a female predominance; most series report a female-to-male ratio of 3-4:1[1,2]. But, Wakhlu et al.[23] reported that the male-to-female ratio was nearly equal, in contrast to the universal female preponderance reported[1,2,12].

The treatment of teratomas is primarily surgical, regardless of the size of the mass, or the young age, even prematurity, of the host. The operation should be instituted as soon as possible[12]. Most children born with SCT have an excellent prognosis after early surgical resection[7]. Prompt diagnosis and resection are critical because the risk of malignancy increases with delayed excision. Altman's classification of the tumor depends on the extent and degree of histological differentiation[2]. A postnatally useful clinical grading scheme was proposed by the American Academy of Pediatrics Surgical Section(AAPSS) (Table 1)[2,7]. Our case belongs to the type I.

The SCT is thought to develop from totipotential cells in the area of Hensen's node[7,17]. The finding of intact motor function in this neonate with split notochord also brings to mind the possibility of diplomyelia, a term that is rarely used because it applies to true doubling of the spinal cord[17]. However, the terms "diastematomyelia" and "diplomyelia" have been used interchangeably[5].

Pang et al.[16] have suggested that these confusing and misleading terms should be abandoned and have proposed an alternative classification to deal with all double spinal cord malformations. They also have proposed a unified theory that explains the embryogenetic mechanisms of all variants of SSCMs. All SSCMs originate from one basic error that occurs at the time at which the primitive neurenteric canal closes. This basic error is the formation of an accessory neurenteric canal between the yolk sac and amnion, which is subsequently invested with mesenchyme to form an endomesenchymal tract that splits the notochord and neural plate. They have defined two types of SSCMs.

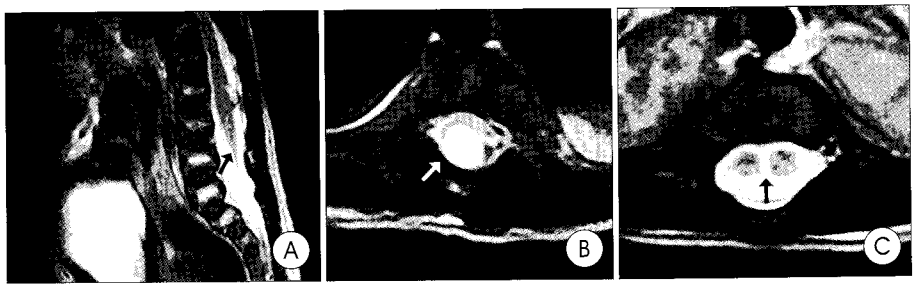

Fig. 3. Spinal magnetic resonance images show multiple spinal anomalies including a tetherd cord (A, arrow), hydromyelia in thoracic and lumbar spinal cord (B, arrow) and diastematomyelia from T11/12 to L4 level (C, arrow).

Fig. 4. Photomicrographs of the tumor specimen. A : Intestinal epithelium is showed (H & E, X40). B : Squamous epithelium and gland are showed (H & E, X40). C : Immature cartilage (open arrow) and bone (black arrow) are shown (H & E, X100).

Table 1. Anantomical grading system for newborn sacrococcygeal teratoma(SCT)

AAPSS type	SCT description
I	Tumor primarily external with minimal, if any, presacral component
II	Tumor primarily external, some presacral component
III	Tumor primarily presacral, small external component
IV	Tumor primarily presacral, may extend into pelvis or abdomen

AAPSS : the American Academy of Pediatrics Surgical Section

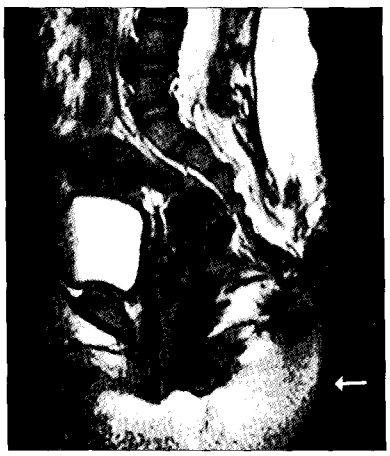

Fig. 5. Postoperative follow up magnetic resonance image after 7 months showing no residual mass (arrow).

A type I SSCM consists of two hemicords, each contained within its own dural tube and separated by a durasheathed rigid median septum. A type II SSCM consists of two hemicords housed in a single dural tube separated by a nonrigid, fibrous median septum.

The association of teratoma with diastematomyelia had been reported 17 cases from 1951 to 1999[3-6,8-11,13,17-19,20,22]. The incidence is extremely rare. In 15 cases, the teratoma was at the same level of the split cord malformation and the teratomas of the other 2 cases were remote from the split cord malformation. Ten cases presented as overt spinal malformations in which a true teratoma was associated with a split cord malformation and various other spinal and extraspinal abnormalities. In 10 cases, the teratomas were all extramedullary (intraor extradural) and 2 only extraspinal. Eight cases of 17 cases were presented less commonly in an occult fashion, so the diagnoses were delayed; 4 were discovered in childhood and 4 in early adulthood. Seven of these teratomas were intradural and the other was extradural.

Ersahin et al.[5] were reviewed the cases of 74 patients with SSCMs who were surgically treated between January 1, 1980 and December 31, 1996. Mean age was 33.08 months. There were 46 girls (62%) and 28 boys (38%). The majority of SSCMs were located in the lumbar and lower thoracic region. The conus medullaris was located below L-2 in all but one patient. Sixty-two patients (83.8%) had at least one associated spinal lesion that could lead to spinal cord tethering. The most common associated spinal lesion was a thick filum terminale that was seen in 30 patients (40.5%). Myelomeningocele and meningocele followed the thick filum terminale in frequency. The association of the teratoma was seen only in one patient (1.35%). The presenting symptoms, in decreasing order of incidence, were skin stigmata, orthopedic deformities of foot, spina bifida, lower paraparesis, scoliosis, bladder and bowel disturbances, etc. They treated patients surgically. Any associated spinal lesions that might lead to spinal tethering were also surgically treated. The clinical importance of SSCMs is that abnormalities associated with split cord malformations are common, and the lesions are capable of tethering the spinal cord. Ersahin et al.[5] reported that 85% of patients had more than one spinal lesion which capable of tethering the spinal cord. Pang[15] found at least one unrelated tethering lesion in all lumbosacral and lower thoracic SSCMs and in a much smaller number of cervical SSCMs. He has suggested that the entire neuraxis should be studied rediographically and , if a second tethering lesion is found, it should be treated surgically. In our case, the operations for the diastematomyelia and cord tethering were not performed due to patient's disagreement and no neurologic abnormalities.

Conclusion

We experienced a extremely rare case of SCT with SSCM. It should be considered that multiple spinal congenital anormalies may occur at multiple levels of the spinal canal in SSCM.

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