ABSTRACT

We present a case of metameric arteriovenous malformation (AVM) in a neonate. The neonate appeared normal, except for a dimple and blue discoloration in the lower thoracic spine. Spinal ultrasonography revealed cutaneomeningiospinal angiomatosis in the thoracic spine. Spinal magnetic resonance imaging revealed a definite metameric AVM at the T10-T11 level and a cutaneous hemangioma at T11. The neonate was conclusively diagnosed with spinal metameric AVM at the T10-T11 level. The neonate was asymptomatic with no, abnormal physical or neurological complications, including urinary incontinence, fecal incontinence, or muscle weakness.

Key Words: Arteriovenous malformations, Infant, Newborn, Spinal cord

INTRODUCTION

Pediatric spinal cord vascular malformations are characterized by clinical features that are different from those of adults. The clinical manifestations of spinal cord arteriovenous malformations (AVMs), including pain, acute or progressive myelopathy, and radiculopathy, depend largely on the degree of malformation. Neonates and infants rarely present with spinal arteriovenous shunts. However, a few cases of patients with spinal AVMs who became symptomatic while they were have been reported.

The treatment of spinal cord AVMs during childhood remains controversial. Surgery and embolization, or a combination of the two, are the current and most common treatments used in clinical practice. We describe a very rare case of a neonatal, metameric AVM observed in our hospital.

CASE REPORT

A neonate was born by cesarean section at a local obstetrics and gynecology clinic. At a
gestational age of 38 weeks, a birth weight of 3.36 kg, and 1-min and 5-min Apgar scores of 9 and 10, respectively. Before delivery, screening ultrasonography did not reveal any specific findings, except cephalopelvic disproportion. Therefore, a scheduled cesarean section was performed. The mother's medical history was unremarkable, and the findings of the neonate's physical and neurologic examinations were normal, with the exception of a dimple and blue discoloration along the lower thoracic spine. The dimple was approximately 0.5 × 0.5 cm in size, and the blue discoloration (Figure 1A) was approximately 2 × 2 cm in size (Figure 1B).

Spinal ultrasonography revealed the presence of an AVM and cutaneomeningospinalangiomatosis of the thoracic spine (Figure 2). The skin dimple was identified by the presence of a skin hemangioma; the AVM was more cephalic than the skin hemangioma. Abdominal and pelvic ultrasonography revealed only mild, normal, pelviectasis; results of all other examinations were normal, including the neonatal screening and auditory tests.

The neonate’s neurological examination results were normal, including the light reflex, muscle tone, and neonatal reflexes (e.g., Moro reflex, grasp reflex). The neonate was asymptomatic, with no urinary or fecal incontinence, muscle weakness, or other physical or neurological complications. The neonate showed normal normal feeding and weight gain. Spinal magnetic resonance imaging (MRI) was successfully performed 29 days after birth; the results showed a definite metameric AVM at the T10-T11 level, and a cutaneous hemangioma at T11 (Figure 3). This enabled a conclusive diagnosis of spinal metameric AVM at the T10-11 level.

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**Figure 1.** Gross physical appearance. (A) Physical appearance of the neonate’s 0.5 × 0.5 cm dimple. (B) Physical appearance of the blue discoloration surrounding the dimple.

**Figure 2.** Ultrasonography findings. (A) Color Doppler image of a transverse scan shows focal vascularity within an oval, hypoechoic, nodular lesion (arrow) in the subcutaneous area of the back, at the T11 level. (B) A longitudinal scan shows a hypoechoic, linear lesion (arrow), with markedly increased color flow, and a connection with a hypervascular mass (thick arrow) on the dorsal spinal cord at the T8-9 level.
We consulted the neurosurgery department, and the neonate was kept for continuous observation at the pediatric department and the neurosurgery department.

**DISCUSSION**

Spinal AVMs are very rare among neonates⁴, with an estimated detection rate of 1.4/100,000 neonates per year in the general population in the United States⁵. Majority of spinal AVMs occur in the thoracolumbar cord. The malformation is a congenital vascular abnormality of the spinal cord, characterized by an arteriovenous shunt, with or without an intervening vascular network between the feeding artery and the draining vein¹². This venous congestion may also be the main pathophysiological feature in neonates and infants. In general, spinal cord malformations may present with a variety of symptoms, including weakness, paraplegia, sphincter dysfunction, and spinal cord hemorrhage⁶. The symptoms can be either acute or progressive⁴, or, as in the present case, the patient may be asymptomatic.

Spinal AVMs have traditionally been divided into three groups according to their anatomical characteristics as follows: type I, which is a dorsal extraspinal AVM; type II or compact, which is an intraspinal AVM; and type III or juvenile AVM². The present case was classified as type I AVM.

In South Korea, a 3-year-old boy with a spinal epidural hematoma presented with lower back pain and fever. He developed paraplegia after two consecutive lumbar punctures performed over the course of 2 days. The spinal epidural hematoma was not detected on lumbar spinal MRI; however, it was detected on cervicothoracic MRI, because the lesion was at the C7-T6 level. Laminectomy was performed to evacuate the hematoma and a specimen was obtained. The pathologic findings indicated the presence of an AVM. The boy required continuous physical therapy
because of, the unsatisfactory results of the hematoma aspiration procedure and laminectomy treatment. He was the youngest individual diagnosed with spinal AVM in South Korea before the present case\(^{10}\).

In addition to spinal AVMs, congenital AVMs of the scalp have also been reported. An 8-day-old male was referred to our institution because of a pulsatile scalp mass in the right parietal area in conjunction with congestive heart failure, which was later determined to be caused by an AVM of the scalp. Embolization of the two feeding arteries, arising from the right superficial temporal artery 11 days after birth resulted in complete cure without recurrence\(^{11}\).

MRI diagnosis of an AVM is based on the presence of serpiginous or tubular structures, corresponding to large draining veins\(^{2}\). However, accurate identification of the involved shunt is difficult by MRI\(^{2}\). Angiography remains the gold standard for the analysis of the anatomical, morphological, and architectural features considered for therapeutic decisions\(^{2}\). When a neonate presents with a dimple, imaging of the spine should be considered when a congenital anomaly of the spinal canal or spinal cord is suspected. The preferred imaging modalities for infants, up to the age of 1 year, are spinal ultrasonography and spinal MRI\(^{7}\). Most studies recommend spinal ultrasonography for neonates presenting with a dimple in the lumbosacral region, excess hair, a pigmented area, multiple cutaneous stigmata, or a hemangioma measuring >5 mm that is located high on the back (>2.5 cm from the anus)\(^{8,9}\).

Once diagnosed, the treatment of a spinal cord AVM typically involves surgery and/or embolization. The treatment of choice depends on the size and shape of the lesion. In some studies, the natural history of the disease indicates progressive deterioration, without spontaneous improvement\(^{1}\). Therefore, early diagnosis and treatment are important\(^{2}\). In the present case, the selected approach was observation without surgery.

**REFERENCES**

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신생아에서 척수동정맥기형은 매우 드문 질환이다. 신생아 척수동정맥기형은 변형의 정도에 따라 통증, 급성부터 진행성, 척수병증, 신경근병증에 이르기까지 증상이 매우 다양하다. 척수 자기공명영상촬영을 통해서 어느 정도 진단할 수 있지만, 혈관조영술을 통해 진단하는 것이 가장 정확하다. 척수동정맥기형의 치료는 색전술 또는 수술이 제시되고 있지만, 아직까지 논란이 많다. 본원에서 매우 드문 질환인 신생아 분절성 동정맥기형이 보고되었다. 신생아는 출생 당시 아래쪽 흉추부위에 소와(dimple)와 주위 피부색의 변화가 동반되었다. 척수 초음파를 시행한 결과 흉추의 피부수막척수혈관종이 발견되었고, 척수 자기공명영상촬영(MRI)에서 T10-T11 부위의 분절성 동정맥 기형이 명확히 보였고, T11부위의 피부혈관종이 밝혀졌다. 신생아는 어떤 신경학적 합병증 없이 무증상이었으며, 신경외과와 상의한 결과 추적 관찰하기로 하였다.