Clinical Case Report

Open meningocele associated with a cervical spinal arachnoid cyst and Arnold Chiari malformation type II, a case report

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Introduction: Spina bifida is a congenital malformation involving the vertebral spine with different presentations ranging from occult defects to large cutaneous defects exposing tissue and nerves.
Case report: Here we describe a patient who had open spina bifida with exposure of the nerve roots, which seemed to be functional on physical examination. A few days after surgery for the closure of the lumbosacral defect, the patient presented with severe respiratory distress and quadriparesis. Magnetic resonance imaging showed a cervical arachnoid cyst and Arnold Chiari type II malformation requiring a second surgical procedure consisting of decompression of the bones and ligaments at the craniospinal junction and microsurgical fenestration of the spinal arachnoid cyst.
Discussion: To our knowledge, spinal arachnoid cysts are a rare entity in pediatric
patients. On the other hand, spinal dysraphism, of which myelomeningocele is the most frequent congenital form, is common. In a literature search, no previous reports were found of cases similar to ours with open spinal dysraphism with these specific anatomical characteristics associated with an arachnoid cyst in the cervical spine and Arnold Chiari malformation.
Conclusion: Spinal dysraphism includes different complex disorders. Full spinal magnetic resonance imaging is recommended for the early detection of associated conditions.

Keywords: myelomeningocele, spina bifida, arachnoid cyst, Arnold Chiari.

INTRODUCTION

Spina bifida (SB) is a term used to describe a set of conditions that may occur during neural tube closure leading to incomplete formation of the spine and are associated with bone defects of different degrees of involvement [1,2]. Spinal dysraphism may be closed or open. Among the latter group, the most common variant is myelomeningocele (MMC) characterized by presenting a placode consisting of rudimentary nervous tissue that are the result of disorders in the neurulation process. Unlike meningocele (MC), it is described as herniation of the dura mater and arachnoids through the osseous SB defect. In the majority of cases, the MC is covered by skin [3,4].

The aim of this study was to describe a rare case of a patient with open SB who at birth presented with a lumbosacral defect with features of both MMC and MC without specifically belonging to either of these types of SB.

CASE REPORT

The patient was a male infant. Spinal dysraphism was diagnosed in utero by ultrasonography. The child was born

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by scheduled Caesarean section. Head circumference was 32 cm.

On physical examination a lumbosacral defect was observed measuring 6 x 4 cm with absence of skin, exposure of underlying tissue, and cerebrospinal fluid (CSF) leak. The placode was not identified, but in its place anatomically healthy nerve roots were seen. On physical-motor examination, the lower limbs could be completely lifted (Fig 1).

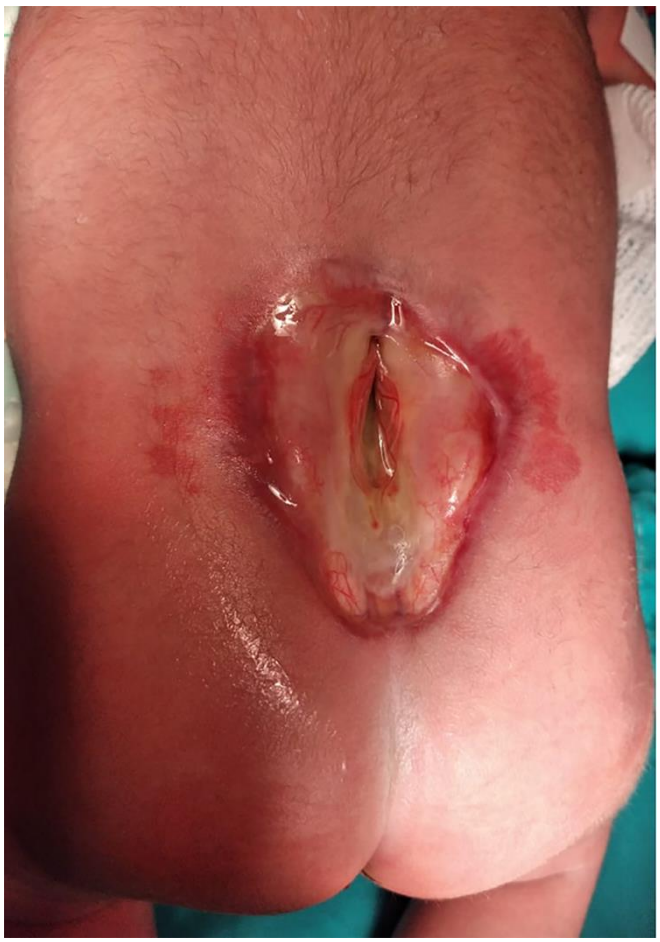

Fig 1- Before closure of the dysraphism. A large open lumbosacral defect without placode is observed. The nerve roots are displaced towards the lateral walls of the defect.

Surgery for the closure of the dysraphism was performed at 16 hours of life. During the procedure the nervous tissue was spared, the dural plane was identified and dissected achieving hermetic closure, after which the musculoaponeurotic plane and skin were closed.

Twenty-two days after surgical closure of the dysraphism, the patient presented with episodes of apnea and marked weakness in the limbs. Quadriparesis of 1/5 on the motor function scale of the Medical Research Council (MRC) was determined [5].

The patient was intubated. Polysomnography (PSG) showed severe obstructive sleep apnea syndrome associated with hypoxemia and hypercapnia.

Magnetic resonance imaging (MRI) of the entire neural axis showed cerebellar hypoplasia and a descent of the cerebellar tonsils through the cervical canal of around 14 mm up to C3-C4 as well as medullary kinking. No ventricular enlargement or transependymal edema were observed. At the lower cervical levels, posterior displacement and a decrease in spinal cord diameter was seen caused a cyst-like mass extending up to C6-C7 (Fig 2,3).

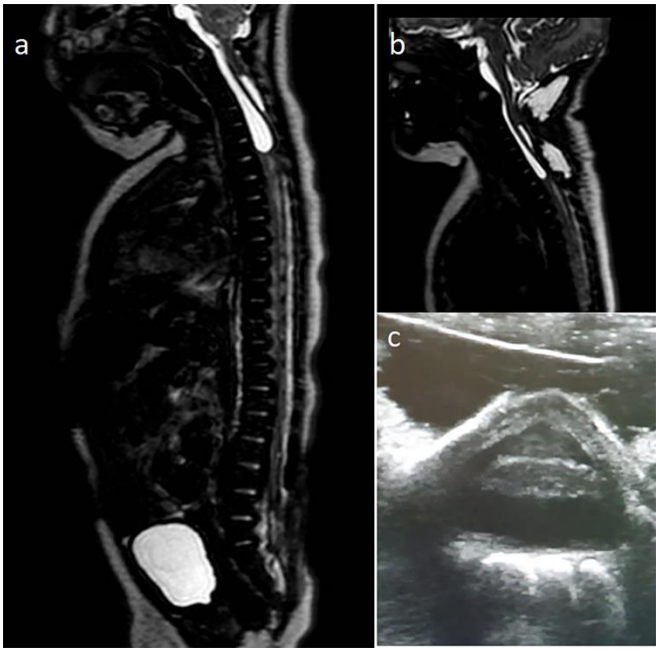

Fig 2- a) Sagittal T2-weighted FIESTA MRI sequence showing a cervical arachnoid cyst ventrally to the spinal cord causing a mass effect and dorsal displacement of the cord; b) On postoperative sagittal T2-weighted FIESTA MRI of the spine a reduced cyst size can be observed; c) Intraoperative ultrasound image showing the bone signal, the spinal cord and the anterior space with the arachnoid cyst.

At 25 days of life, a second surgery was performed consisting of osseoligamentous decompression of the craniocervical junction to resolve the Arnold Chiari II malformation and laminectomy of the 6th vertebra and hemilaminectomy of the 7th cervical vertebrae followed by microsurgical ultrasound-guided fenestration of the arachnoid cyst.

Postoperative MRI showed a decrease in cyst size (Fig 2).

Five days after the second surgery, the patient was extubated requiring non-invasive ventilation for four more days. At one week of life, a postoperative PSG showed no changes compared to the preoperative study.

At 4 weeks of life, the patient did not require supplementary oxygen and was fed orally. He had no

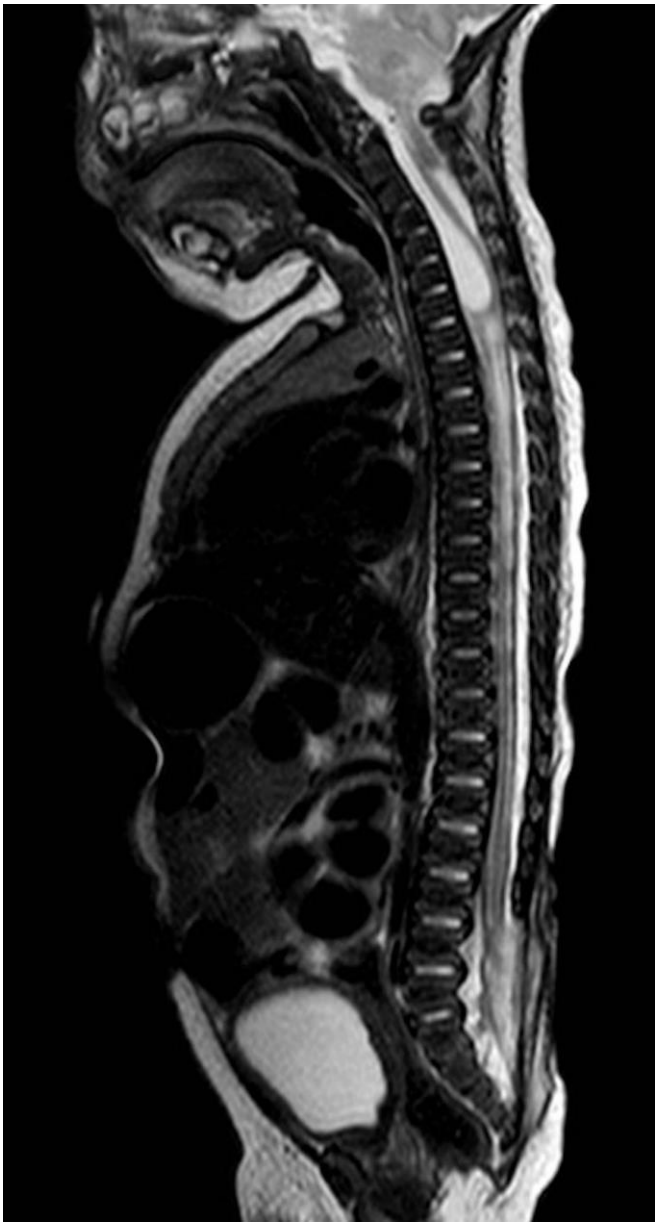

Fig 3- Sagittal T2-weighted MRI sequence showing the medullary cone at the level of the third lumbar vertebra.

hydrocephalus and moved his limbs with a paresis of 4/5 on the MRC motor function scale [5].

DISCUSSION

Among the different types of spinal dysraphism, MMC is the most common congenital defect, characterized by exposure of nervous tissue leading to failure of primary neurulation [6]. MMC is associated with a tethered cord, Arnold Chiari type II malformation, hydrocephalus and has been related to maternal folate deficiency [7-10]. MC, on the other hand, is believed to occur during secondary neurulation and may be associated with Arnold Chiari type I malformation, hydrocephalus, and various genetic syndromes [11-12]. MC is most often a closed, i.e. skin covered, defect although it may be open in rare cases [3-12].

In our case, we could speculate that the defect occurred at some time between primary and secondary neurulation as the spinal cord and nerve roots had developed completely, but closure of the dura and overlying tissues was not achieved. Our patient had a lumbosacral malformation that did not strictly meet the criteria for MMC or MC described in the literature.

Multiple and complex mechanisms are involved in the development of the central nervous system. Although the trend is to classify conditions to facilitate research and understanding, there are rare cases that do not fit into the existing classifications.

Patients with MMC associated with a lumbar cyst posterior to the closure of the spinal defect have been reported; however, in none of the cases they were diagnosed at the time of birth [13]. Spinal arachnoid cysts are an uncommon cause of neural compression in children, with rare reports of the association of these sacral cysts with occult SB [14]. Chiari Type II malformation consists of significant herniation of the cerebellar tonsils through the foramen magnum and is characterized by elongation and caudal displacement of the cerebellar vermis and brainstem, also the presence of a MMC in virtually all cases.

Our patient presented with a cervical spinal arachnoid cyst in addition to tonsillar descent causing severe respiratory symptoms and quadriparesis in the first days of life. Surgical treatment consisted of microsurgical fenestration of the arachnoid cyst and osseoligamentous decompression of the Chiari type II malformation which showed to be successful up to the moment of this report resulting in improvement in postoperative images and motor symptoms. However, the respiratory symptoms remained unchanged after surgery. The pathophysiological mechanism explaining the above-described picture may be the extrinsic compression of the medulla by the arachnoid cyst causing the motor deficit in an anatomically preserved motor pathway. On the other hand, alterations on the PSG that remained unchanged after decompression may have been secondary to intrinsic malformation of the cranial nerve nuclei occurring during gestation in some patients with this type of disease. According to this hypothesis, dysfunction is due to failure of cranial nerve formation rather than secondary to extrinsic compression and does therefore not respond to surgery [15].

The great variability of patients with spina bifida highlights the need for multidisciplinary treatment and follow-up. This favors early detection of associated pathology.

We have presented an extremely rare case in which an interdisciplinary treatment and follow-up was carried out, which allowed an early diagnosis of its comorbidities. This

favored an adequate surgical approach and a favorable evolution.

CONCLUSION

Spinal dysraphism involves a spectrum of different, complex conditions. Complete MRI of the neuraxis is recommendable for early detection of associated disorders. Here were describe a rare case of spinal dysraphism with an atypical presentation due to the anatomical shape of the lumbosacral defect and its association with Arnold Chiari type II malformation and a spinal arachnoid cyst.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Ethics approval was not required for this study.

Consent to participate

The patient gave consent to use his information and images for research proposes.

Consent for publication

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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CONTRIBUTIONS

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