

Cervical Meningocele about 2 Operated Cases

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Abstract

Cervical meningoceles are the least common forms of spina bifida, representing between 4% to 8% of all spinal dysraphisms. They are not accompanied by neurological deficit in childhood but attached cord syndrome can occur with growth. Surgical treatment, in addition to avoiding rupture and correcting the aesthetic damage, will preserve neurological functions. We report 2 cases that we took care of. They are a 5-month-old infant and a 3-year-old girl. The first presented a 7 cm rounded meningocele pedunculated through the C1 lamina defect. The second presented an 8.6 cm polylobed meningocele pedunculated through the C2 lamina defect. They both underwent surgery with good progress.

Keywords

Cervical Meningocele, Spinal Dysraphism, Spina Bifida

1. Introduction

Cervical meningocele is one of the least common forms of spinal dysraphism. They represent only 4% to 8% of all spinal dysraphisms [1]. Meningocele is a lack of closure of the posterior midline which concerns the meninges, the vertebral lamina, and the muscular plane. It is made from an arachnoid herniation through the defect which forms the pocket containing only CSF and covered by skin without neural plate. Cervical meningoceles are not accompanied by neurological deficit in childhood. It has the particularity that the spinal cord is sometimes attached to the dura mater and other soft tissues by a meningo-fibrous element exposing it to an attached cord syndrome with growth. Surgical treat-

ment does not only have an aesthetic objective. Its aim is to preserve neurological functions by removing cord attachment. The aim of our work is to report our experience through two cases that we operated with very good progress.

2. Observations

2.1. Case No. 1

5-month-old female infant with a notion of second-degree consanguinity, born vaginally from an irregularly monitored pregnancy. She was brought to us for consultation for a congenital neck mass. The examination found a rounded cervical mass 7 cm in large diameter, pedunculated at its base, of soft fluctuating consistency covered by thin skin which was translucent and pink at the level of the dome (**Figure 1**). There was no CSF leak. The infant was in good general condition with no neurological deficit, normal head circumference and good psychomotor development. We did not find any other malformations. The CT scan of the cervical spine showed a C1 lamina defect from which arises the very small neck of a cystic mass measuring 61×45.2 mm with subcutaneous expansion suggestive of a cervical meningocele (**Figure 2**). Brain CT showed mild bifrontal cerebral atrophy without ventricular dilatation. During the surgical treatment of the meningocele we performed a circumferential skin incision circumscribing the mass at the level of its pedicled insertion base followed by dissection of the subcutaneous, aponevrotic and muscular planes up to the limit of the spinal bone defect in C1. After collar identification (pedunculated meningeal attachment) between C0-C1, we opened the cyst from the collar in order to cut the meningo-fibrous attachment of the spinal cord. We performed dural closure after sectioning the collar then closing the aponevrotic, subcutaneous and cutaneous planes (**Figure 3**). There were no postoperative complications. The evolution at 6 months postoperatively was good without neurological disorders or hydrocephalus.



Figure 1. Pre-operative image of a large cervical meningocele pedunculated at its base covered with thin translucent and pink skin at the level of the dome.

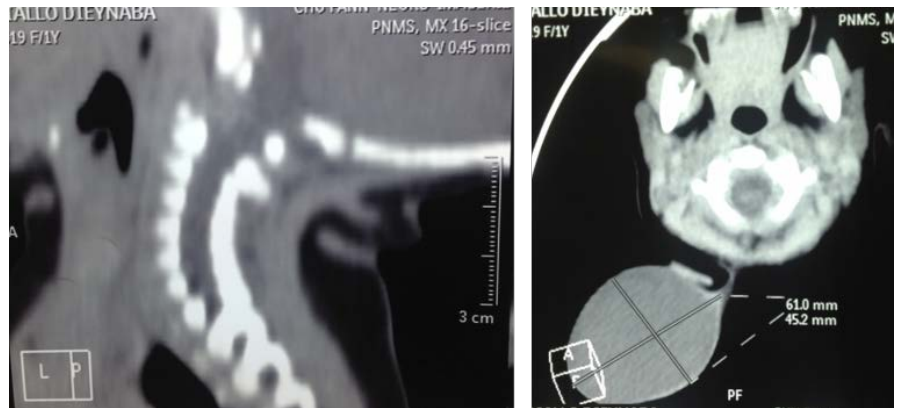


Figure 2. CT scan of the cervical spine showing the cervical meningocele in the form of a cystic mass with subcutaneous expansion arising from the medullary canal from the C1 lamina defect where the very small neck is located.

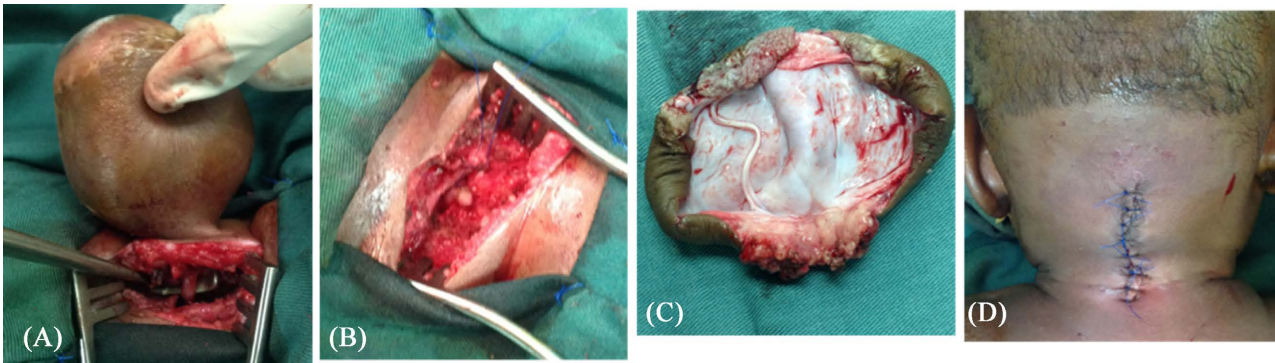


Figure 3. Operative images (A) Skin incision at the base of insertion and dissection of the different planes up to the limit of the C1 bone defect. (B) and (C) Opening of the cyst from the collar in order to cut the meningo-fibrous attachment of the spinal cord (black arrows); (D) End of intervention after closure of the dura, aponeurosis, subcutaneous planes

2.2. Case No. 2

3-year-old girl born vaginally from a poorly monitored pregnancy with no particular history who was brought to us for consultation for a congenital cervical mass gradually increasing in size. The examination found a rounded polylobed posterior median cervical mass 8.6 cm in large diameter, pedunculated at its base with soft consistency and covered by normal skin (**Figure 4**). The child was in good general condition without neurological deficit or macrocrania with good psychomotor development. We did not find any other malformations. The brain and cervical spine scan showed a type 2 of Arnold Chiari malformation associated with a bony defect of the C1 and C2 laminae from which the collar of the hypodense cystic formation (55.1 × 18.7 mm) begins subcutaneous expansion and continuity with the spinal canal suggesting a cervical meningocele (**Figure 5**). We performed a circumferential skin incision circumscribing the mass at the level of its base of insertion followed by dissection of the subcutaneous, aponeurotic and muscular planes up to the limit of the spinal bone defects of C1 and C2. We opened the cyst from the collar which we had clearly defined at the level of the defect of the C2 laminae. We did not find any meningo-fibrous attach-

ment to the spinal cord. We performed dural closure after sectioning the collar and then closing the aponevrotic, subcutaneous and cutaneous planes (**Figure 6**). The outcome at 6 months postoperatively was good without neurological disorders or hydrocephalus.



Figure 4. Pre-operative image of a polylobed cervical meningocele, pedunculated at its base covered by normal skin

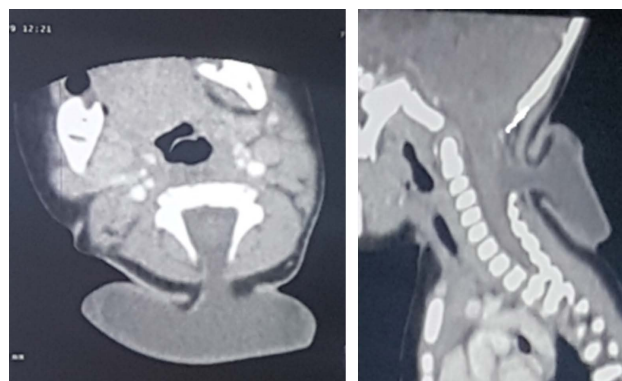


Figure 5. CT scan of the cervical spine showing a bony defect in the laminae of C1 and C2 from which the neck of the hypodense cystic formation expands subcutaneously and is continuous with the spinal canal, suggesting a cervical meningocele associated with a type 2 Arnold Chiari malformation.



Figure 6. Operative images (A) Skin incision at the base of insertion and dissection of the different planes up to the limit of the bone defects of C1 and C2; (B) End of intervention after closure of the dura mater, aponevrosis, subcutaneous and cutaneous planes; (C) Open meningocele cyst without identified meningo-fibrous attachment.

3. Discussion

Cervical spinal dysraphisms are less common than those of the lumbosacral region where they are mainly observed [2]. Their proportion in all spinal dysraphy varies between 1 to 8% according to some studies [1] [3] [4] [5]. This rarity means that most authors only report one or two cases [1] [5] [6] [7] [8] [9] with the exception of Habibi *et al.* [3] who reported 16 cases of cervical myelomeningocele and Salamao *et al.* [4] who reported 10 cases of cervical siege in a series of 18 cervico-dorsal spinal dysraphism. The nomenclature and classification of cervical spina bifida according to the type of lesion and the contents of the malformed cyst remain a subject of semantic divergence. We adhere to the classification established by Salamao *et al.* [4] into three types based on the structures found in the cyst. It distinguishes type I (Stalks) in which neuroglial or fibrovascular tissue originating from the posterior surface of the spinal cord attaches to the wall of the cyst. Type II (myelocystocele) contains a secondary ependymal cyst herniating inside a meningocele that is to say the external cyst which is in continuity with the subarachnoid space while the ependymal cyst is in relation to a central canal hydromyelic. Type III (true meningocele) is a herniation of the meninges through the defect constituting a cyst filled with CSF. No neuronal elements are found inside the cyst and only arachnoid adhesions can attach the spinal cord. According to this classification, our first case is a type I and our second case a type III.

This is a malformation that is discovered antenatally using ultrasound or early postnatally. In our two cases, the diagnosis was postnatal at 5 months and 3 years without neurological disorder. The female predominance in our series as in that of Salamao [4] is confirmed by Copp *et al.* [10] in his study of 140,000 annual cases of neural tube anomalies.

The usual clinical presentation is that of a soft posterior midline cervical mass [1] which may be pedunculated or sessile. The dome of the lesion may be covered either by thin translucent pink skin (squamous epithelium), by fibrous tissue or by normal skin. In all cases, the base of insertion and most of the wall are covered by normal skin [4]. Leakage of CSF through the lesion is exceptional; Salamao [4] reported one case in 10 cervical spina bifida observed. One of our cases had the dome covered with fine pinkish translucent skin without CSF leak while the other had normal skin. The large diameters of the cervical masses of our two cases (6.1 and 8.6 cm) were greater than those of Chandra [6], Pessoa [7] and Wong [9] which were respectively 6 cm, 4 cm and 3.5 cm. In larger series, Andronikou [11] and Salamao [4] respectively measured diameters varying from 2.5 to 11.5 cm and 2 to 16 cm.

There is no neurological disorder in the forms discovered in newborns and children without other associated malformations like our cases as well as those of other authors [1] [3] [5] [6] [7] [9]. However, Adronikou [11] reported in his series a case of tetraparesis and mental retardation in a 14-year-old child with an anomaly of the corpus callosum. The association with congenital malformations

such as hydrocephalus and Arnold Chiari malformation was reported by Salamao [4] in his series of cervico-dorsal spinal dysraphima.

The exploration and diagnosis of spinal dysraphisms in general and cervical meningoceles in particular is ideally done by MRI (magnetic imaging), which remains the examination of choice [11]. MRI better shows small abnormalities of the spinal cord and nerves and offers good soft tissue contrast allowing determination of the meningocele or myelomeningocele type with better resolution as well as the associated anomalies and malformations [11] [12]. In the absence of MRI, spinal Computed tomography (CT) scan is a perfect complement. CT with multiplanar reconstructions is a good modality for diagnosing vertebral abnormalities but offers less contrast in soft tissues making it a deblurring examination. Our two patients were only explored by CT of the cervical spine and brain in search of associated anomalies. In our two cases the bone defects concerned the laminae of the C1 and C2 vertebrae and a type 2 Arnold Chiari malformation was associated in the second case. Our patients didn't have an MRI because it's costly for them. In Salamao study [4], preoperative imaging was only found in 10 out of 18 patients and included ultrasound, CT and MRI in only 8 patients. He noted that malformations can occur at all cervical levels and the associated malformations were: hydromyelia, hydrocephalus and Chiari type 2.

Surgical treatment is unanimous in all reported cases. It does not only have an aesthetic aim because in addition to avoiding rupture and therefore leakage of LCS; it helps preserve neurological functions by removing the attachment of the cord. Surgery must be performed early and consists of resection of the cystic pocket after intradural exploration in order to free the cord from the fibromembranous attachment which is excised and then sealed dural closure [1] [4] [6] [7] [9]. The search for and removal of the spinal cord attachment is essential to avoid the occurrence of neurological disorders that can occur with growth in the context of a tethered cord syndrome. The choice of our surgical technique is motivated by the watertight correction of the cosmetic problem and the avoidance of the occurrence of neurological disorders.

The postoperative outcome is often favorable without CSF leak or neurological disorders when surgery is performed quickly in the absence of other associated malformations as in our cases as well as in Chandra, Feltes and Wong [1] [6] [9]. However, in Salamao's study [4], the evolution was marked by neurological disorders such as spasticity, tetraparesis, mental retardation and death in cases associated with other malformations.

4. Conclusion

Cervical meningoceles are very rarely reported. They are not accompanied by neurological deficit in childhood but lead to the occurrence of attached cord syndrome with growth because the spinal cord is sometimes attached to the dura mater and other soft tissues by a meningo-fibrous element. Surgical treatment must be early and not only for aesthetic purposes. Its aim is to preserve neuro-

logical functions by removing cord attachment. The postoperative evolution is good without neurological disorder or CSF leak when there are no other associated malformations.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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