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Primary Repair of a Myelomeningocele in a Geriatric Patient: A Case Report

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Abstract

Background: Myelomeningocele is the most common open spinal dysraphism eligible for surgical repair. Traditionally, myelomeningocele patients that received no surgical intervention had a dismal prognosis. Before 1970, 80% of untreated infants were dead at 3 months, while after 1970 more untreated children survived infancy but usually did not live past adolescence. Adult presentation of myelomeningocele is exceedingly rare.

Case presentation: We present a 74 years Caucasian female was transferred to our institution for evaluation of purulent drainage around an unrepaired myelomeningocele site. Per the patients report, she was offered no surgical treatment for her myelomeningocele as a newborn, but was instead treated with percutaneous needle drainage of the lumbar fluid collection on a weekly basis for several months. Examination of her back was notable for a large 6 cm×6 cm fluid filled mass in the lumbosacral region with an abnormal epithelialized covering and purulent drainage. Her brain MRI revealed arrested hydrocephalus and a Chiari II malformation. Due to the infection, she underwent surgical correction of the myelomeningocele. At 6 month follow up she had a well healed wound and no complications from the surgery.

Conclusion: The present case adds to the small body of literature describing adult presentation of spinal dysraphism. To our knowledge, this is the oldest patient to undergo primary repair of a congenital myelomeningocele. Primary repair, even in the elderly, is possible and can be done safely.

Keywords: Myelomeningocele; Spinal dysraphism; Geriatric; Infection; Hydrocephalus

Introduction

Myelomeningocele (MM) is the most common open spinal dysraphism eligible for surgical repair. Traditionally, MM patients that received no surgical intervention have a dismal prognosis. Reasons for not intervening in the past were multifold, but typically referred to the potential burden that MM children could place on themselves, their family, and society [1]. Before 1970, 80% of untreated infants passed away by 3 months [2]. After 1970, more untreated children survived infancy but usually did not live past adolescence [3]. Treated or untreated, those that do survive are often limited by various degrees of bowel and bladder dysfunction, orthopedic disabilities and, to a lesser degree, cognitive abnormalities. Adult presentation of myelomeningocele is exceedingly rare. To our knowledge, this is the first report of a geriatric patient with an unrepaired myelomeningocele and is a remarkable survival story.

Materials and Methods

Our institution's IRB approved this retrospective Case Review.

Results/Case Presentation

The patient is a 74 year female with an unrepaired myelomeningocele who was transferred to our institution for evaluation of purulent drainage around the myelomeningocele site. Several months prior to this presentation she had an episode of cryptogenic bacteremia that was treated elsewhere with broad spectrum intravenous antibiotics. She underwent an MRI of the lumbar spine prior to transfer (Figure 1A). During a discussion with the patient we discovered that she was offered no surgical treatment for her myelomeningocele as a newborn, but was treated with percutaneous needle drainage of the lumbar fluid collection on a weekly basis for several months. Beyond her urostomy, she had no other surgical treatments.

Upon examination, the patient was alert and oriented without any overt cognitive abnormalities. She had full motor strength of her bilateral upper extremities with flaccid paralysis in her bilateral lowers. She had no voluntary bowel or bladder function. Examination of her back was notable for a large 6 cm×6 cm fluid filled protrusion in the lumbosacral region with an abnormal epithelialized covering and foul smelling drainage (Figure 1B). She underwent further imaging at our institution consisting of an MRI of her brain that revealed longstanding radiologic ventriculomegaly and a Chiari II malformation (Figure 2). Due to the active leakage, she was admitted to the hospital to continue her intravenous antibiotics and for surgical correction of the myelomeningocele.

The patient was taken to the operating room were the myelomeningocele was repaired in usual fashion. We were able to dissect the neural placode and nerve roots free from the overlying

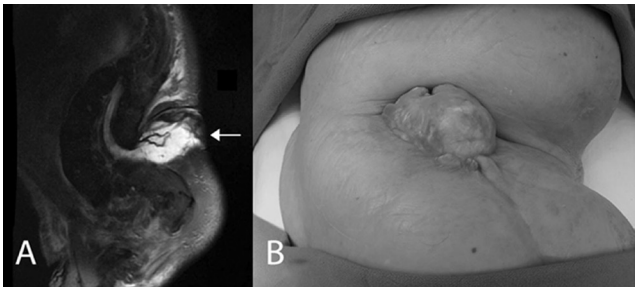

Figure 1: (A) Sagittal MR image T2 of the lumbar spine demonstrating the myelomeningocele with surrounding fluid collection (white arrow). (B) The pre-operative lumbosacral region of the patient with the open myelomeningocele.

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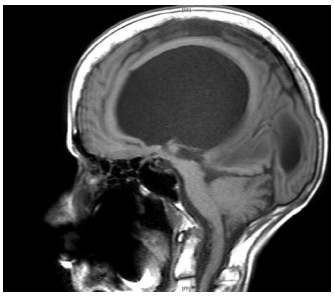

Figure 2: Sagittal T1 MR image of the Brain showing ventriculomegaly and a Chiari II malformation.

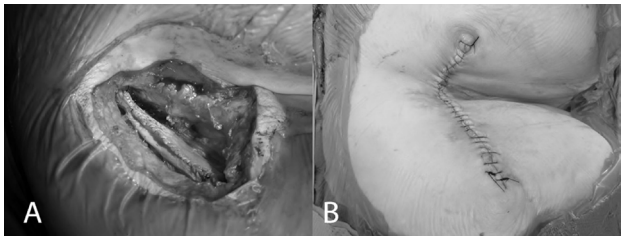

Figure 3: (A) An intra-operative photo showing the thecal sac fashioned with remnant dura closed with running nurolon and (B) the skin closure of the myelomeningocele with a running nylon suture.

tissue and fashion a thecal sac from the dural remnant (Figure 3A). The closure of the neural tube was reinforced with a small piece of dural substitute and dural sealant. The fascia and was able to be primarily closed over the thecal sac and the overlying subcutaneous tissue was mobilized and approximated using care to eliminate dead space. The skin was also mobilized and closed with a running suture (Figure 3B).

Post-operatively she progressed well without evidence of symptomatic hydrocephalus or leakage from her surgical site. She was discharged without complication on a full course of oral antibiotic regimen. The patient was seen in the outpatient clinic for suture removal that was uneventful and was noted to have continued good wound healing at her 6 month follow up.

Discussion

We described a case of a 74-year-old woman who presented with a large, infected, and unrepaired myelomeningocele with an associated Chiari II malformation. This case is remarkable for many reasons, but even more so when taken in historical context.

Before 1975, repair of a myelomeningocele with CSF shunting was noted to have a significant impact on survival to the 16th birthday [3]. However, there was an accepted practice of relegating some children to supportive therapies only. The criteria that delineated which children were chosen for conservative treatment varied, but included children with 'unshuntable' hydrocephalus (central nervous system infection or bleeding preventing CSF shunting), severe lower extremity paralysis, or additional birth defects complicating medical management (cardiopulmonary or renal complications [1,3,4]. Not surprisingly, 80% of the untreated infants before 1970 died by 3 months of age. Our patient was relegated to the supportive care population shortly after her birth. She did not have surgical closure of her defect and did not receive a CSF shunt. Astoundingly she never proceeded to get a life threatening infection and, besides her urostomy, she had no other surgical treatments.

The treatment of myelomeningocele has evolved dramatically over the last half century. In particular, the 1970s saw a revolution in the care of myelomeningocele patients. Many factors, including prenatal referral to tertiary care centers, improvements in fetal ultrasound, computed tomography, advances in CSF shunting, and broad spectrum antibiotics have led some experts to propose MM as a "new disease" since the mid-seventies [3,5]. Childhood survival data at our institution reflects this revolution, as childhood survival improved from 60% in 1960 to 90% in 1985 [3]. Currently, in developed countries, there are very few infants who do not have immediate repair of their myelomeningocele with the first 1-2 days of life, often with rotational flaps [6]. The literature has a few case reports of adult cervical meningoceles and 1 case of a cervical myelomeningocele [7,8]. Interestingly the cervical myelomeningocele was in a 52 years old man who presented asymptomatically. Adult presentations of other spinal dysraphisms have been reported as well. These include split cord malformations and various congenital lesions that cause adult onset tethered cord [9-13].

The present case adds to the small body of literature describing adult presentation of spinal dysraphisms. To our knowledge, this is the oldest patient to undergo primary repair of a congenital myelomeningocele. Primary repair, even in the elderly, is possible and can be done safely.

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