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Quality of life in a 61-year-old paraplegic patient with myelomeningocele: illustrative case

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BACKGROUND Limited data exist on survival of patients with myelomeningocele (MM) into late adulthood. Patients born prior to 1975 received less surgical treatment and demonstrated poorer survival. Patients born after 1975 show lower mortality rates because of advancements in prenatal care, diagnostics, technology, and treatment. The increasing number of adult patients with MM demonstrates these successes but also highlights the need for a better understanding of both medical management and quality-of-life issues in this population.

OBSERVATIONS A 61-year-old woman presented to the hospital with a functional level T12, vertebral level low lumbar MM that had developed a tissue defect with a cerebrospinal fluid leak. The patient had her MM closed at birth and since then had received no additional surgical treatment or repair of her MM defect. After successful treatment, the patient continues to follow up for additional decompression procedures and admits to a good quality of life.

LESSONS This case adds to the limited literature describing management of patients with MM over the age of 60. The patient's self-described good quality of life and longevity align with reports of importance of independence, level of education, bowel management, ability to provide self-care, strong interpersonal relationships, and absence of shunted hydrocephalus.

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KEYWORDS myelomeningocele; spina bifida; congenital; adult; longevity; life satisfaction

Myelomeningocele (MM) is a severe form of spina bifida due to a neural tube defect, and it is commonly associated with other congenital anomalies such as hydrocephalus, Chiari II malformation, bowel and bladder incontinence, kyphoscoliosis, and lower extremity paralysis.

The current literature estimates survival in patients with MM and comorbid hydrocephalus at 30 to 40 years.[1–4] Lesion location is a significant factor in the rate of mortality, with higher disability and mortality rates in patients with MM who have higher level lesions.[5] Other causes of mortality rates are a history of comorbid conditions, specifically hydrocephalus and Chiari II malformation.[6]

In the 1950s, primary repair for patients with MM spina bifida was not typically offered, and only 10% of patients with MM survived past infancy.[7] In addition, patients with MM and associated hydrocephalus born before 1975 did not necessarily receive surgical MM repair and cerebrospinal fluid (CSF) shunting. Comparison of patients treated with MM closure and CSF shunting versus those with no treatment demonstrated a survival benefit with surgical treatment (68% survival to age 16 in treated versus 27% in untreated).[8] Because of improved surgical and medical management, after 1975, 90% of patients with MM treated with closure and CSF shunting survived to age 16, compared with 58% of untreated patients.[8] Although MM closure has resulted in increased survival, timing of shunt placement for MM-associated hydrocephalus remains controversial because of shunt malfunction and rates of infection.[9] When patients with MM who had survived to adulthood (>16 years of age) were independently grouped by hydrocephalic shunt placement, a 20% decrease in survival for 20% of patients with shunted hydrocephalus was observed.[8] This is a continuing area of research and discussion.

Fewer infants are born with MM as a result of aggressive education on the importance of improved maternal nutrition and folate

ABBREVIATIONS CSF = cerebrospinal fluid; MM = myelomeningocele.
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supplementation, and infants who are born with MM have reduced lesion severity.[10] In addition, prognosis for infants born with MM has improved because of prenatal screening and early neurosurgical intervention.[7,8,11] Collectively, these advancements indicate that more patients with MM are living into adulthood.[12] The increasing adult population of patients with MM has resulted in a growth in hospital admissions among this patient population, and complications noted involve hydrocephalus, paraplegia, neurogenic bladder disturbances, and degenerative spinal disease.[13] While there has been an upward trend in patients with MM living past the age of 40, the number of admissions in patients over age 50 is much smaller.[13] We have found only four other documented reports of patients with MM over the age of 60, detailing (1) a primary repair of an MM in a geriatric patient,[14] (2) the benefits of microsurgical decompression therapy in an adult patient,[15] (3) primary repair of an ulcerated lumbosacral MM in adulthood,[16] and (4) a rare case of an elderly patient with a large prevertebral thoracic MM.[17] In accordance with the literature, our patient is a rare example of an individual with an MM and comorbid hydrocephalus who has survived well into adulthood.

Our goal in this report is to document the life of a patient over 60 years of age presenting with MM and nonshunted hydrocephalus to better inform management of future elderly patients with MM. Particular attention is paid to her quality of life.

Illustrative Case

This patient is a 61-year-old woman with a functional level T12, vertebral level low lumbar MM, nonshunted hydrocephalus, associated Chiari II malformation, kyphoscoliosis, bowel and bladder incontinence, and complete paraplegia (Figs. 1 and 2). Her other medical conditions include essential hypertension, coronary artery disease, nephrolithiasis, and restrictive lung disease.

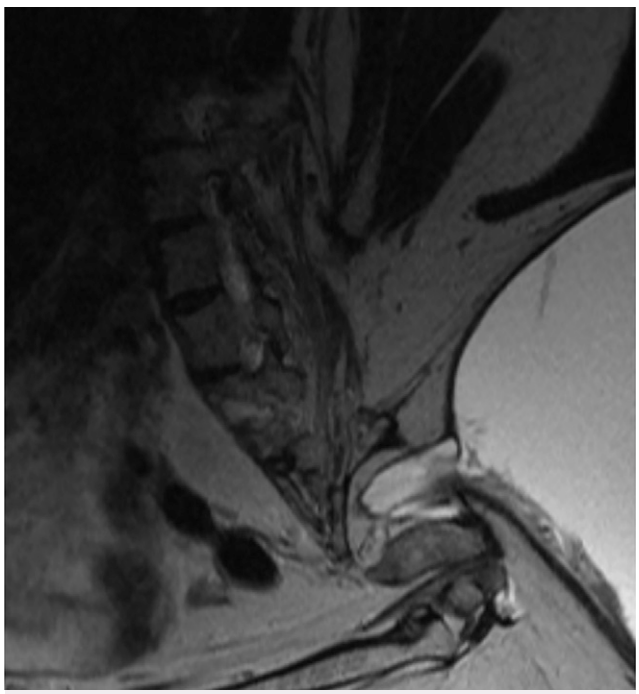

FIG. 1. Sagittal T2 magnetic resonance imaging shows marked scoliosis, spinal deformity, and lumbar MM.

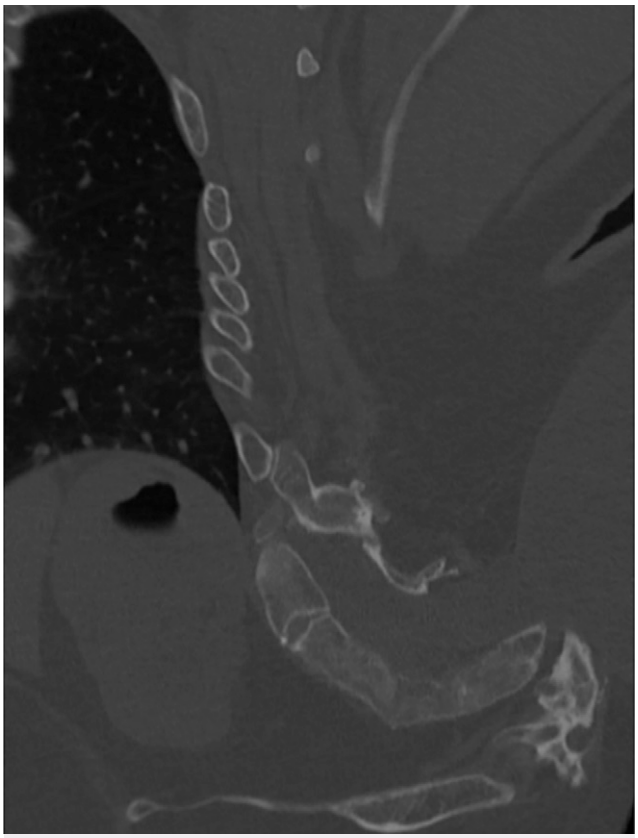

FIG. 2. Coronal spinal computed tomography shows marked scoliosis and lumbar spina bifida.

Patient Oral History

The patient was born in 1959 and grew up in the Chicago, Illinois, area with her older siblings and both parents. She was diagnosed with an MM at birth and underwent closure of the defect on the day of delivery. She was also diagnosed with hydrocephalus that physicians decided not to shunt because they thought she would not survive much longer. When she was born, her parents were told that her chances of survival were low, she would experience lower than "normal intelligence," and she would be "better off being put in a home" for the disabled. Her parents raised her with love and support. Although her father built parallel bars to help her walk with the support of leg braces, the patient has been a wheelchair user since 7 years of age. She began experiencing severe headaches accompanied by nausea and vomiting at age 21 as a result of her Chiari malformation. The patient followed up with a neurologist at University of Illinois and presented to more than 30 different doctors with her parents; however, no additional interventions were performed.

Patient Education

The patient attended school with other physically disabled children, where she was socially active and excelled academically. After high school, she received a bachelor's degree and wrote her thesis on wheelchair accessibility on campus. She then enrolled in a master's degree program, which she was unable to complete because of debilitating headaches caused by her Chiari II malformation.

Patient Residency and Work

The patient lived with her parents until their passing in 2004, and as of this writing, she remains at the residence. Throughout adulthood, the patient held several part-time jobs based from home and worked as an intern at a local nursing home. She currently works as a reading and writing tutor. The patient lives alone with assistance from a nurse two to three times per week as well as nearby family members.

Patient Mental Outlook

The patient describes her quality of life as "good" and has maintained a positive outlook, relying on strength, motivation, and faith. Her largest support system is her family. Her older brother lives in Michigan, and her older sister and her family live close by and visit several times per week. The patient describes her overall life as "happy"; however, she admits bouts of depression due to the limitations of her condition. She states that her regrets are her inability to complete her education or have children of her own, although she has found joy from her nieces and nephews. Tutoring her niece gives her a sense of purpose and worth. Her everyday life consists of rehabilitation two to three times a week, managing her finances, watching TV, maintaining her social contacts, and advocating for disabled persons. She has found her purpose and chooses to live by her motto "survive, succeed, and surpass." From our patient's perspective, factors that are important to quality of life when living with MM include (1) having supportive family and friends, (2) believing in a higher power, (3) receiving an education, (4) feeling academically stimulated, (5) finding purpose in everyday life, and (6) accepting the condition itself.

The patient's neurological function has remained fairly stable over time. She has no cognitive impairments. Her MM and comorbid conditions have led to her lower extremity paralysis, urinary and bowel incontinence, and debilitating headaches. She manages bowel and urine incontinence with colostomy and urostomy bags. Over time, the patient developed a basketball-sized mass at her MM site (Fig. 3) and has received five operations consisting of decompression and drainage over the past 2 years.

The patient had received no prior surgical treatment since the original repair of her MM and presented in August 2018 with CSF drainage from her MM site. This was managed initially by simple suturing of the small defect; however, the CSF leak recurred after 2 weeks, at which time the patient received an additional procedure. This procedure consisted of aspiration of the MM and closure of the soft tissue defect using plastic surgical techniques. Approximately 2 L of fluid was aspirated directly, which allowed for decompression. A CSF fluid sample showed no evidence of infection. Further tissue mobilization and plastic surgery repair of the defect were then performed. The patient remained stable after the procedure and is currently doing well. Magnetic resonance imaging and computed tomography of the brain revealed stable ventriculomegaly, Chiari II malformation, marked thinning of the cortical mantle of the parietal lobes bilaterally, and superficial siderosis within posterior fossa.

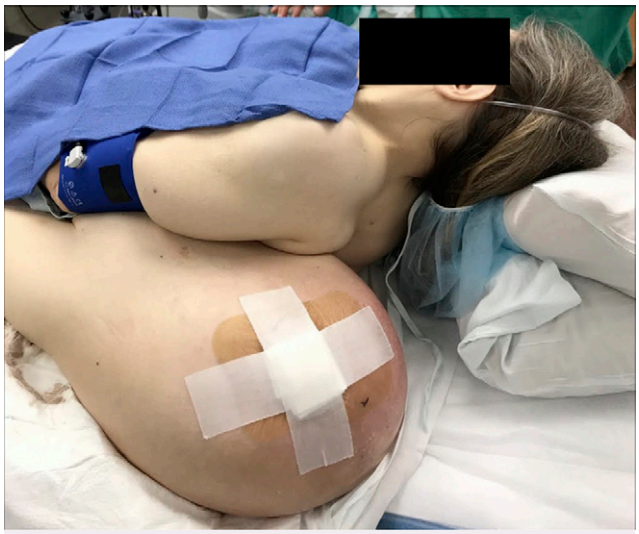

FIG. 3. Distended MM sac.

Discussion

Patients living with MM over the age of 60 are rare, and the need for more research on the management and care of this patient population is warranted. We have been able to identify four reports of individuals with MM over the age of 60. Overall, these case reports depicted patients with different corresponding lesion levels as well as unshunted hydrocephalus, all without cognitive abnormalities. The first study detailed a 65-year-old woman with congenital thoracolumbar kyphoscoliosis, spina bifida aperta, and MM with symptoms of radicular pain and weakness of the left lower extremity. The patient was found to have spinal canal stenosis and received microsurgical decompression surgery. The report details the effectiveness of the surgery in relieving the patient's weakness and pain while improving her overall quality of life.[15] A second study described the primary repair of an MM in a 62-year-old woman who presented with ulceration and CSF leakage at the site of her MM, lower extremity weakness due to a tethered spinal cord, and hydrocephalus. The patient received successful primary repair of her MM, detethering, and placement of a lumboperitoneal shunt, improving her symptoms and decreasing her risk for further complications.[16] The third case report involved a 72-year-old man who presented with a sudden-onset weakness and impaired sensation in his left lower extremity. The patient was found to have a thoracic prevertebral myelomonocyte and tethering of the spinal cord. The patient received a partial T4 corpectomy with microsurgical detethering, allowing him to recover ambulation after the procedure.[17] The final study followed a 74-year-old woman with purulent drainage from her unrepaired MM site. This patient received surgical correction and is considered to be the oldest patient in the current literature to undergo primary repair of an MM,[14] demonstrating that safe primary repair in elderly persons is possible. The limited number of case reports detailing the management of patients over age 60 with MM and associated conditions demonstrates the importance of sharing our patient's journey and neurosurgical care.

Observations

Whereas these case reports all discuss elderly patients with MM, different factors play a role in the appropriate type of neurosurgical management for individual patients. Many patients with MM have different levels of disability and different associated conditions that affect their treatment plan. The patient presented here has a vertebral level low lumbar MM associated with an increased risk of disability and mortality compared to a sacral lesion.[5] In addition, the

patient suffers from comorbid conditions such as compensated hydrocephalus and Chiari II malformation, which increase her risk of mortality.[6] Interestingly, similar to the four case reports in the literature, our patient has not had her hydrocephalus managed with shunt placement. Some evidence shows an increased likelihood of survival past the age of 35 in patients born before 1975 who did not receive shunts for their associated hydrocephalus.[8]

Quality of life for an elderly patient with MM was a focus of this report. Several studies have examined factors that account for quality of life among patients with MM and other spinal dysraphisms. An examination of the predicting factors that lead to permanent disability among patients with spinal dysraphism, including patients with MM, used patient self-assessment to determine their level of disability. Although it is important to acknowledge that patients define disability in different ways, the findings of the study concluded that the strongest predictors of self-reported permanent disability are level of education and degree of stool continence.[18] In addition, the patient's level of self-reported disability correlated with participation in society as well as overall life satisfaction. An additional study explored quality-of-life satisfaction of patients with MM in more detail by examining the patients' level of education, employment status, reproductive status, and relationships in addition to their medical conditions. Similar to our patient, most patients in the study had completed a high school education, with more than 50% completing a college degree. Most patients received their income from disability benefits, made less money than the national average, had higher levels of unemployment, and required a higher level of assistance. In general, participants with hydrocephalus as well as a lack of independent mobility were found to have more limitations in terms of obtaining education, having meaningful employment, living independently, forming partner relationships, and having children. Patients who had greater difficulty in meeting these adult milestones reported lower life satisfaction. Overall, life satisfaction was most associated with family relationships and ability to perform self-care.[19]

These studies highlight the effects that MM and comorbidities have on adult patients and their quality of life and highlight the different factors that play a role in patient life satisfaction. Physicians treating adult and elderly patients with MM must consider all the factors that contribute to the patient's physical condition and overall quality of life. Although it is easy to assume a patient's quality of life by his or her medical impairments, disability for patients with MM is not always defined by physical limitations, and individual fulfillment is achievable in this patient population. Our patient has stool incontinence managed by a colostomy, and based on the literature, stool management significantly impacts quality of life. Our patient has been able to live semi-independently and reports a great amount of familial support, which are likely contributing factors to her high self-reported quality of life. There is benefit for neurosurgical intervention in elderly patients with MM, with the caveat that more research needs to be done on MM. Quality of life can be good from the patient's perspective.

Lessons

MM is the most severe form of spina bifida and is often associated with nerve malfunction as well as hydrocephalus, Chiari II malformation, paralysis, and bowel and bladder incontinence. While current literature estimates a long-term survival rate between 30 and 40 years for patients with MM and comorbid hydrocephalus, more patients are living into adulthood. There is an ongoing dialogue regarding timing of hydrocephalus shunting in patients with MM, and the case reported herein highlights a fifth elderly patient who did not receive a shunt for her hydrocephalus. Our case demonstrates that patients with MM who are over age 60 can experience a full and meaningful life. Furthermore, current advances managing adults with MM will lead to improved life expectancy. It is important to identify factors that improve quality of life for seniors with this condition to improve patient longevity and overall patient care.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: both authors. Acquisition of data: Borowsky. Analysis and interpretation of data: Borowsky. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Administrative/technical/material support: Ruge. Study supervision: Ruge.

Supplemental Information

Previous Presentations

Portions of this work were presented as a clinical poster at the M3 Clinical Poster Competition at Chicago Medical School, Rosalind Franklin University of Medicine & Science, North Chicago, IL, June 9, 2021.

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