Technical Note & Surgical Technique

Pseudomeningocele masquerading as monomelic amyotrophy

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A R T I C L E   I N F O

Article history:
Received 3 June 2016
Accepted 12 June 2016

Keywords:
Pseudomeningocele
Monomelic amyotrophy
MRI
CT myelogram
Neurosurgery
Myelopathy

A B S T R A C T

Pseudomeningocele is a known cause of myelopathy, however this is classically associated with bilateral symptoms. We discuss a patient presenting with single upper extremity weakness, initially diagnosed as monomelic amyotrophy, which further investigation revealed to be a giant pseudomeningocele. This case illustrates a unique presentation of pseudomeningocele, including prominent changes in MRI, CT myelogram, and EMG. A review of the literature for pseudomeningocele including pathology, diagnosis, imaging, and treatment as well as the initial presentation of monomelic amyotrophy is provided.

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1. Introduction

This case illustrates an atypical presentation of pseudomeningocele (PM) that appeared to mimic monomelic amyotrophy (MMA). Pseudomeningoceles are rare extradural collections of cerebral spinal fluid, with a giant PM defined as being greater than 8 cm in length. It is formed by a breach in the dural-arachnoid layers, typically either traumatic or iatrogenic. This case is the first reported instance of unilateral myelopathy. We describe the clinical course, electromyogram, and radiologic findings of a 38-year-old Indian male diagnosed with giant PM.

2. Case description

A 38-year-old right-handed Indian male presented to the clinic for evaluation of right arm weakness and atrophy. The patient stated that in 2005 he was in a car accident and broke his right humerus. He recalls mild residual atrophy and weakness that mostly resolved. In the last two years, his friends and family noted the right upper extremity (RUE) atrophy and weakness had progressively worsened. He was seen by multiple specialties and referred to neurology for evaluation of possible motor neuron disease with previous RUE brachial MRI showing muscle wasting. The patient denied any symptoms in his left upper or bilateral lower extremities but did complain of some fasciculation intermittently in the RUE with no cramping. He reported no dark urine, fever/chills, or weight loss. On examination, his RUE strength was graded as follows: deltoid 3, spinati 2, triceps 3, biceps 4-, wrist extensors 4, interossei 4. He had normal sensation and slightly reduced reflexes. The remainder of his examination was unremarkable.

Given his history of gradually progressive weakness and atrophy in the setting of an ethnic Indian background, monomelic amyotrophy (MMA) was the initial diagnosis. Other possible etiologies were ruled out with testing for paraneoplastic and vasculitic causes. EMG showed normal motor and sensory conduction with needle exam showing extensive chronic focal disorder of motor neurons, their axons, or both in the RUE from C5–C8 with some ongoing denervation. MMA was still the leading diagnosis but the span of the neuropathy caused some doubt and imaging was obtained to rule out spine pathology and possibly confirm MMA. A cervical spine MRI was obtained and showed a giant PM; this was confirmed by CT myelogram showing the collection was predominantly on the right (Figs. 1 and 2). This appears to be the first report of single limb involvement of giant PM. The patient was offered two options for a surgical correction of the leak. First option would be open surgery involving finding a source of leak and correcting it. Given location, the procedure has high morbidity associated with it and there was no clear identifiable source of leak. Second option was placing a cystopleural shunt which would help to decompress the cyst and has lesser complications associated with it. After discussing options and risks, the patient opted for the shunt which was done by C3–C4 cervical laminectomy followed by decompression of the cyst and placing catheter in this cervical region and pleural space. After 2 months he returned to the clinic with repeat MT myelogram showing slight reduction in size and patient reporting improvement in RUE strength.
The initial concern was for MMA given the clinical exam, history, and EMG findings. MMA is defined by slow onset of muscle wasting and weakness of one arm with no sensory loss. The etiology is debated, with two main hypotheses: the Hirayama “dynamics” hypothesis postulates that neck flexion causes a circulation deficit, while the “imbalanced growth” hypothesis of Toma involves inelastic dura resulting in microtrauma [1,2]. Regardless of the mechanism, the result is progressive loss of anterior motor neurons of the cervical spine. Males ages 15–25 of Asian descent, most commonly Japanese and Indian, are primarily affected [3]. The hypothenar and thenar muscles are affected more severely in patients with MMA (compared to cervical spondylotic amyotrophy or amyotrophic lateral sclerosis), with an ulnar/median nerve CMAP ratio of 0.55 ± 0.41 [4]. Imaging in MMA results in a wide variety of findings including abnormal cervical curvature, asymmetric cord flattening, localized lower cervical cord atrophy, and noncompressed intramedullary high signal intensity, with loss of attachment between the posterior dural sac and subjacent lamina being the most statistically important finding with sensitivity of 93% and specificity of 98% [5]. The disease plateaus over several years, but attempts to prevent progression with neck collars or fusion surgeries have been promising.

In this case, the breadth of the neuropathy and the patient’s age were two factors that caused suspicion for an alternative diagnosis and led to focused cervical spine imaging. It was this imaging that led to the final diagnosis of PM. The question of the etiology for our patient remains. PMs result from a breach in the dural-arachnoid layer, causing an extra-dural cerebrospinal fluid (CSF) collection with eventual formation of a fibrous capsule. Most commonly it results from iatrogenic (intentional vs. accidental) durotomy, and rarely from trauma or congenital abnormality [6]. Epstein et al. did a thorough review of inadvertent traumatic durotomies and found that 3–11% of surgeries not involving the dura had a post-op dural tear [7]. Infrequent nonsurgical causes of PM include needle puncture, trauma, direct trauma, excessive dural or nerve root traction, dural laceration from sharp bone fragments [8]. Similar events of large PMs from traumatic brachial injury have previously been reported [9–11].
Kotani et al. presented a similar case that started with upper extremity monoparesis but proceeded to affect all the limbs soon after onset [10]. The current case is unique in that the patient had only one limb involvement that had slowly progressed for 2 years without any effect on other limbs. The patient’s prior auto accident with lingering residual weakness may have been a brachial injury that caused this PM.

PM may be diagnosed via multiple imaging modalities. CSF collections were distinguished on MRI by absence of mass effect, low T2 signal complexity, and low T1 signal intensity [12]. There may also be communication with the thecal sac combined with spinal imaging of dilatation of the epidural venous plexus and diffuse dural thickening and enhancement [12]. However, MRI of some congenital defects, cysts, and tumors may show similar signals and need to be considered if suggested by the clinical history. CT Myelography is better for determining the source of the leak, especially when combined with retrograde radionuclide or delayed imaging techniques [13]. Absence of the nerve sheath and pulsatile leakage of contrast into the extra arachnoidal sacs are typically visualized on CT [14,15].

The treatment of PM is controversial, with management dependent upon many factors including location, sac size, and symptoms. Given that PMs may stop and resolve spontaneously, it is recommended to observe initially with bed rest in the Trendelenburg position for 7–14 days [16]. Further conservative treatment with watertight skin closure and repeated subcutaneous punctures of the PM is also recommended [17]. Epidural blood patches and aspiration have been applied successfully to treat patients with incidental durotomies with PMs [18]. Temporary catheters may result in resolution but long term catheters have also been used, typically in the setting of poor surgical access [19,20]. Indications for surgery include progressive radiculopathy, signs and symptoms of myelopathy, or failure of conservative measures. The indications for surgery include progressive radiculopathy, signs and symptoms of myelopathy, or failure of conservative measures. I n d i c a - tions for surgery include progressive radiculopathy, signs and symptoms of myelopathy, or failure of conservative measures.

The definitive treatment for PMs is surgical dural repair, stitching the leak closed with fibrin sealant, often supplemented with a dural, muscle, or fat graft placed over the area of the persistent leak [21,22]. In this case, given that we were unable to locate a source of the leak and that the PM was anterior to the cord preventing extensive intraoperative exploration, a T4-5 laminectomy was performed with placement of an arachno-pleural shunt.

4. Conclusion

Pseudomeningocele is a rare cause of myelopathy and should be included in the differential even if there is no evidence of trauma.

References