Unusual Causes of Paraplegia: Literature Review

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Abstract

Paraplegia refers to damage to the neurons in the thoracic, lumbar, or sacral segments of the spinal cord, resulting in an injury that is often associated with the loss of motor or sensory functions. In general, the function of the upper limbs is not affected, and the extent of damage to the lower body is determined by the location and level of the injury. Due to advancements in medical technology, a considerable proportion of paraplegic patients can resume independent functions of daily life and return to work or school, following proper treatment and rehabilitation. However, paraplegia is only a symptom, not a cause; therefore, the early diagnosis of paraplegia is of great importance. The causes associated with paraplegia are numerous, ranging from common trauma to rare infections, and the symptoms can vary. We discussed five uncommon paraplegia causes that have been cited in the literature, including spinal dural arteriovenous fistula, spontaneous spinal epidural hematoma, aortic dissection, decompression sickness, and spinal tuberculosis, and review the pathophysiology and clinical symptoms associated with each cause, as well as their treatment and prognosis. We hope to expand the range of possibilities for the differential diagnosis of paraplegia, to increase the likelihood of proper diagnoses and treatment at early stages.

Key words: paraplegia, arteriovenous fistula, spontaneous hematoma, decompression, aortic dissection, spinal tuberculosis

Introduction

Paraplegia refers to the impairment or loss of motor or sensory function in the thoracic, lumbar, or sacral regions of the spinal cord, secondary to the damage of neural elements within the spinal canal. Arm functioning is generally spared, but the trunk, legs, and pelvic organs may be involved, depending on the level of injury [1]. Symptoms can include the loss of sensation or mobility below the level of injury, the loss of bladder and bowel function, and unexplained pain. Paraplegia can be further categorized into complete and incomplete paraplegia. Complete paraplegia is defined as the complete lack of motor or sensory function at the lowest sacral segment, whereas incomplete paraplegia indicates that some amount of function, either motor or sensory, is preserved below the neurological level. Paraplegia is predominantly caused by spinal cord injuries that result from accident or trauma; however, other causes include iatrogenic injuries, genetic disorders, autoimmune diseases, infections, tumors, and stroke. Treatment varies, depending on the severity, the level of injury, and the etiology. In this article, we briefly discuss some uncommon etiologies, to expand the possibilities available to readers when performing differential diagnosis for paraplegic patients.

Relevant Case Presentation

Case 1: Spinal dural arteriovenous fistula (SDAVF)

A 79-year-old female patient, with a history
of chronic atrial fibrillation under anticoagulation therapy, was admitted to the hospital with persistent coma after a first episode of seizure. Lumbar puncture was performed, and prophylactic enoxaparin was instituted 24 hours later. Abrupt lower limb anesthesia and paraplegia developed 5 days after the lumbar puncture. Magnetic resonance imaging (MRI) revealed a subarachnoid hematoma from T12-L5, and arteriography showed a spinal dural arteriovenous fistula at T12 to L1. Embolization and spinal laminectomy were performed, for subarachnoid hematoma evacuation  

Case 2: Spontaneous Spinal Epidural Hematoma (SSEH)

A 31-year-old female patient, without a recent history of infection, trauma, surgery, or anticoagulant use, presented at the emergency department with weakness in the bilateral lower extremities, after waking up in the morning. The patient then deteriorated to the acute onset of flaccid quadriplegia [American Spinal Injury Association (ASIA) score A]. The initial laboratory tests and blood pressure were within normal limits. Neurological exam disclosed the loss of bilateral lower extremity muscle power (0/5), the loss of skin sensation below the T4 vertebral level, with saddle anesthesia, a positive Babinski sign of the bilateral lower extremities, and reduced bilateral tendon reflex. MRI revealed a posterior epidural hematoma, from the T2 to T3 levels, with spinal cord compression. Spontaneous spinal epidural hematoma was diagnosed. An urgent, posterior-approach, decompression laminectomy with hematoma evacuation was performed, and early rehabilitation was initiated  

Case 3: Acute Aortic Dissection

A 56-year-old female patient, with a medical history of chronic obstructive pulmonary disease, presented to the emergency department with the chief complaints of lumbar back pain and paresthesia and paralysis in both lower extremities. She was in her usual state of health, without walking difficulties, 90 minutes before the presentation of symptoms. She denied any previous surgeries, intravenous drug abuse, fever, or recent trauma. Her vital signs were remarkable for hypertension and tachypnea. The initial laboratory findings disclosed hypokalemia, elevated glucose levels, and elevated white blood cell counts. Computed tomography (CT) scan revealed disc herniation, which resulted in L5/S1 spinal stenosis. After the administration of pain control and antibiotics, she was referred for MRI scanning. Upon arrival, she was noted for a blood pressure of 176/84 mmHg and sustained low back pain. Physical examination revealed flaccid paralysis and both lower extremities were cold and pulseless to palpation. Neurologic examination disclosed anesthesia below the level of T10, with absent patella tendon reflexes. Doppler ultrasound failed to detect bilateral dorsalis pedis and femoral pulses. CT angiography (CTA) showed Stanford type A aortic dissection (AD), extending to the celiac trunk, with an abdominal aortic thrombus proximal to the iliac bifurcation. Anti-hypertensive agents were administered, and urgent surgical repair was performed  

Case 4: Decompression Illness (DCI)

A 46-year-old, experienced, female diver dived to 110 feet for 27 minutes and conservatively decompressed for 13 minutes, at 10 feet. Within 10 minutes after she surfaced, her right foot felt hot and tingly while climbing onto the boat. Then, it became progressively numb, with radiation to the thigh. Her left limb suffered from the same symptoms, and low back pain developed. She could not walk after reaching the shore for 30 minutes. Immediately, pure oxygen was delivered for 60 minutes, and her sensation and strength gradually recovered. She was fully recovered and felt normal one week later  

Case 5: Spinal Tuberculosis

A 33-year-old female presented to the hospital with low back pain that radiated to her lower extremities and the inability to walk. She began experiencing these symptoms approximately four months previously and was incapable of performing her usual activities. One month prior to her presentation, she noticed the progressive inability to move her lower limbs and eventually became bed-bound. Occasional fever, decreased appetite, and weight loss were mentioned, beginning six months prior to presentation. No cough, chest pain, or shortness of breath was noted. On examination, she appeared malnourished, with tachypnea and raised body temperature. Chest auscultation revealed bilateral crackles. Lower limb examination disclosed bilaterally decreased muscle power (3/5), with diminished plantar reflexes and
tenderness at the L1-L2 area. X-ray of the lumbosacral spine region showed L2 compression, with the involvement of the L1-L2 paradiscal area. MRI of the spine revealed spondylodiscitis at the L1-L2 level, with a soft tissue component compromising the spinal canal and a bilateral psoas muscle abscess. Chest X-ray revealed a bilateral heterogeneous opacity, with right upper lobe (RUL) cavitation. Sputum acid-fast staining was positive. She was diagnosed with pulmonary tuberculosis, with vertebral involvement. Spinal decompression was performed, and antitubercular therapy was administered [17].

Discussion

A. Spinal dural arteriovenous fistula

SDAVF is the most common vascular malformation of the spine. SDAVF has a male predilection and many cases become symptomatic during middle age, suggesting that it is likely to be an acquired disease. Most lesions are solitary and are frequently found in the thoracolumbar region (T6-L2). The exact etiology remains unclear [2,3].

 Typically, a radiculomeningeal artery enters a radicular vein and forms an SDAVF on the dorsal surface of the dural root sleeve in the intervertebral foramen. Consequently, the increased pressure within the perimedullary venous plexus results in decreased spinal vein drainage, leading to venous congestion, with intramedullary edema and dilated and tortuous veins, which subsequently cause spinal cord hypoxia and myelopathy [4].

Symptoms progress slowly and are nonspecific, including tingling pain, lower back pain, paresthesia, sensory loss, lower extremity weakness, and unsteady gait. Symptoms become worse during physical activity. Bowel and bladder incontinence and erectile dysfunction often present late during the course of the disease. However, spinal hemorrhage is very rare [3,4].

SDAVF diagnosis is generally confirmed by MRI and spinal angiography. On a T1-weighted scan, after contrast administration, the dilated perimedullary vessels are enhanced. Meanwhile, cord edema appears as hyper-intensity, with peripheral sparing, on a T2-weighted scan. A prominent, serpiginous, and engorged perimedullary venous plexus can be observed on the dorsal aspect of the cord. Digital subtraction angiography (DSA) is the gold standard for diagnosis because it can help identify the location of the fistula. After injection, the delayed return of contrast material in the radiculomedullary arteries, with the early venous filling of the radiculomedullary veins, can be observed [4,5].

Treatment with endovascular embolization is performed using liquid embolic agents, to occlude the distal part of the feeding artery and the proximal part of the draining vein. Other options include surgical therapy to ligate the draining vein, which should be considered if endovascular treatment is contraindicated or fails [2-4].

Prognosis is associated with the duration of the symptoms and disability before treatment. Although motor and sensory impairment benefit the most from treatment, sphincter and erectile dysfunction often respond less effectively [3,4].

B. Spontaneous spinal epidural hematoma

SSEH is defined as the accumulation of blood within the epidural space, without known traumatic or iatrogenic causes, and constitutes less than 1% of all spinal epidural lesions, with an annual incidence of approximately 1 per million individuals. SSEH typically occurs in individuals in their forties and fifties, and the exact etiology remains unknown. Predisposing factors include underlying coagulopathy, anticoagulant use, arteriovenous malformation, increased intra-thoracic or intra-abdominal pressure, and hypertension [6,7].

The origin of hemorrhages remains under debate. However, most evidence supports the hypothesis of venous bleeding. The low pressure, valveless, epidural venous plexus is vulnerable to any changes in pressure from the thoracic and abdominal cavities, such as those that occur during coughing and sneezing, which can result in a venous rupture. According to the arterial bleeding hypothesis, a hematoma forms quickly, resulting in the abrupt onset of SSEH. However, the origin of bleeding is not a major prognostic factor [6,7].

Compared with the ventral epidural side, SSEH is more often observed on the dorsal aspect, due to the larger dorsal epidural plexus and a region called the locus minor resistentiae, which is more susceptible to rupture following minor variations in intravenous pressure. Moreover, the ventral epidural veins receive more support from the posterior longitudinal ligament. SSEH occurs predominantly
at the cervicothoracic and thoracolumbar segments because the increased mobility of these segments applies more tension to the epidural veins [7].

Symptoms vary, depending on the location and severity of spinal cord compression. Typically, patients present with the acute onset of severe neck or back pain, with occasional radiation to the extremities, followed by the rapid progression to nerve root or spinal cord compression symptoms. Hyporeflexia and flaccid paralysis may also present [6-8].

MRI is the diagnostic standard because it can determine both the location of the hematoma and the severity of spinal cord compression. Within the first 24 h of symptom onset, hematoma typically appears hyperintense on T2-weighted images and often becomes hyperintense on both T1- and T2-weighted scans 24 h later. Chronic hematomas may emerge as hypointensities on both T1- and T2-weighted images [7].

Surgical decompression, along with hematoma evacuation, is the standard treatment for SSEH. Based on a small number of case reports, many have suggested urgent surgical decompression within 12–48 h of symptom onset, to achieve optimal neurologic improvements. Only those who are not fit for surgery and those who are asymptomatic, with spontaneous hematoma resorption, can be managed conservatively [7,8].

The time interval to surgery and, more importantly, the degree of preoperative neurological deficits are two major prognostic indicators. Other poor prognostic factors include the involvement of four or more spinal segments, sensory impairment, and the involvement of areas that have narrow spinal canals [6-8].

C. Acute Aortic Dissection

AD is a life-threatening condition that describes a tear in the intimal layer of the aortic wall, leading to blood extravasation into the media of the aorta and the creation of a false lumen. AD may affect other arteries, by blocking blood flow in the true lumen, resulting in decreased blood flow to the vital organs. Dissection weakens the aortic wall, causing aneurysm formation or vessel rupture. The mortality rate increases by approximately 1% per hour during the first 48 hours if the diagnosis is delayed [9,10].

AD can be classified into two types, DeBakey or Stanford, depending on the site of the dissection. DeBakey ADs can be further subdivided into type I, which involves both the ascending and descending aorta, type II, which involves only the ascending aorta, and type III, which involves the descending aorta, distal to the left subclavian artery. Stanford ADs can be subdivided into type A, which involves the ascending aorta, and type B, which involves the descending aorta. Dissection is more commonly observed at proximal sites [9,10].

Risk factors for AD development include hypertension, age, being male, smoking, connective tissue disease, and congenital anomalies [9,10].

Typically, the patient presents with abrupt, severe, tearing, sharp chest pains that may radiate to the back. Other symptoms include syncope, heart failure, cerebral vascular accidents, shock, pulse deficits, limb ischemia, and a murmur of aortic insufficiency [10].

However, paraplegia as an initial presentation observed in fewer than 2%–5% of all AD cases, most likely caused by anterior spinal artery syndrome, due to direct compression from the dissection or dissection of branch vessels. For instance, the artery of Adamkiewicz, between T10-L2, can cause spinal cord ischemia. AD often starts with the loss of pain and temperature sensation while maintaining intact vibration and proprioception sensations. Depending on the duration of ischemia, paraplegia may then develop [9,11-12].

Diagnosis includes history taking, physical examination, and imaging evaluations, such as chest X-rays, trans-esophageal echocardiogram, CT, or CTA [10].

However, if paraplegia is the initial manifestation, doctors may be misled by the lack of spinal cord compression on MRI. Therefore, the prompt suspicion of acute AD should be considered when symptoms and signs of distal vascular abnormalities, asymmetric radial pulses, uncontrolled blood pressure, or persistent chest pain occur [9,11-12].

Treatment plans depend on the type of acute AD. In general, intravenous beta-blockers are administered first, followed by intravenous vasodilators, to reduce the heart rate below 60 bpm and blood pressure to the lowest level that preserves perfusion. If the ascending aorta is involved, urgent intervention, with surgical management, is usually required. If only the descending aorta is involved, conservative treatment with medical therapy is generally sufficient [9,11-12].
Moreover, if spinal cord ischemia develops, a lumbar drain can be placed, to lower intraspinal pressure, optimize cord perfusion, and improve neurologic outcomes [9,12].

The prognosis of paraplegia due to acute AD is often considered poor [11].

D. Decompression illness

Decompression illness (DCI) is a dysbaric disease that includes decompression sickness (DCS) and arterial gas embolism (AGE). Factors, such as depth-time exposure, breathing gas, diver exertion, and water temperature can affect the probability of DCI. The estimated incidence of DCI is approximately 0.03%, with increased risks among patients with persistent patent foramen ovale, and the mean age of DCI patients is 39 years [13,14].

Decompression sickness occurs due to Henry’s Law, when nitrogen gas comes out of solution and supersaturation occurs during ascent, forming bubbles in the tissue and venous blood. The level of bubble formation depends on the depth and duration of the dive and the rate of ascent. Once bubbles develop, they result in vascular injuries, such as intravascular coagulation and plasma leakage, and central nervous system (CNS) injuries, caused by secondary oxidative stress and inflammation. DCS presentations differ in severity, with pain in the joints or muscles and paresthesia being the most common symptoms. The classification depends on the manifestations of DCS. Type I includes joint pain, skin rash or marbling, swelling or pain in the lymph nodes, and focal edema. Type II includes cardiorespiratory failure, inner ear presentations, such as vertigo, and neurologic symptoms involving the brain or spinal cord, including weakness or paralysis, visual disturbances, bowel or bladder dysfunction, lethargy, and confusion. Most patients become symptomatic within the first 24 hours [13-15].

An arterial gas embolism occurs when a diver breathes compressed gas at depth and rapidly ascends, without exhaling the air from the lungs. According to Boyle’s Law, this series of events may result in alveolar rupture and subsequent air leakage into the surrounding spaces, leading to barotrauma, such as pneumothorax, and pneumomediastinum. Moreover, air can enter the pulmonary arterial circulation, causing embolisms that occlude the terminal arteries, such as those in the brain, resulting in the development of stroke-like events. Manifestations vary depending on the sites that are involved. Pain, respiratory distress, headache, unconsciousness, seizure, blindness, or paresis may be presented [13-15].

The diagnosis of both DCS and AGE is based on clinical evaluations, including diving history, depth-time profile, onset of the symptoms, and physical examination. Laboratory and imaging studies generally provide little benefit to the diagnosis. Although chest X-ray and CT can help to detect complications associated with pulmonary barotrauma, they are often only used to exclude other etiologies. MRI may aid the diagnosis of neurologic DCS; however, MRI results are often normal [13,14].

The severity of the injury and the time to effective treatment greatly affect neurologic outcomes. DCS and AGE are associated with similar presentations, often occur together, and management strategies are essentially the same for both syndromes. The immediate administration of 100% oxygen can promote the oxygenation of hypoxic tissues and accelerate the rate of nitrogen removal. The timely transport the patient to a recompression chamber for hyperbaric therapy with oxygen as the primary treatment, as this treatment can reduce bubble volumes and inflammation and improve tissue ischemia and edema. Additional management strategies, such as the use of non-steroidal anti-inflammatory drugs (NSAIDs) can decrease the number of recompression therapy but do not alter clinical outcomes. Fluid resuscitation is necessary for potential hypovolemia and hemoconcentration. Anticoagulants are administered prophylactically in the event of paraplegia. The avoidance of hyperglycemia and hyperthermia is also recommended [13,15,16].

The risks of DCI can be reduced by adopting decompression procedures, which allow divers to ascend at a rate that is compatible with the slowest tissue that accepts and releases gas for the depth and duration of the dive. The avoidance of vigorous exercise, the use of alcohol, and cold conditions during decompression after diving is also recommended [13,15,16].

In general, the long-term neurologic prognosis is good, and patients achieve complete recovery, likely because most injured divers are young and have better health conditions [13,15,16].
E. Spinal tuberculosis

Spinal tuberculosis (TB), often referred to as Pott’s disease, accounts for 1%–2% of total tuberculosis cases and is the most common manifestation of musculoskeletal tuberculosis. Spinal TB is most often observed in poorly developed countries and is more common among HIV-infected patients. Spinal TB generally occurs secondary to an extra-spinal source of infection, such as the lungs. Mycobacterium tuberculosis spreads through hematogenous routes to the vertebral bodies, where thoracic and lumbar segments are most commonly involved. The anterior aspect of the vertebral body, adjacent to the subchondral plate, is usually affected. As bone destruction progresses, collapsing vertebral bodies and kyphotic deformities may present, forming an internal gibbus. Associated abscesses, granulation tissues, and tubercular debris may result in spinal cord compression and neurologic deficits. Inflammation edema, cord atrophy, or even myelomalacia may develop [18,19].

The presentation is insidious, and constitutional symptoms, such as fever, night sweats, and weight loss are often reported. Non-specific, chronic back pain may also be noted. If the cervical spine is affected, the disease may manifest as neck pain and stiffness, dysphagia, stridor, and hoarseness. Cutaneous sinuses, paraspinal abscesses, muscle rigidity, and spasm may be detected. Neurologic abnormalities present as early spasticity, with exaggerated deep tendon reflexes. The loss of motor power, including weakness and paraplegia, develops prior to the reductions in pain and temperature sensations. As compression increases, flaccidity and flexor spasm, complete sensory loss, and the disturbance of bowel and bladder sphincters may occur [18,19].

To describe the severity of paraplegia, some experts have proposed a 4-grade classification, based on motor weakness, walking ability, and neurological deficits, as detected on neurological exam. This classification system can help plan and manage spinal TB and paraplegia symptoms [18,20].

Grade 1: Negligible weakness, ability to walk without help, clinician detects plantar extensor, muscle power grade 4-5.

Grade 2: Mild weakness, ability to walk with help, brisk tendon jerks, muscle power grade 3, paresthesia.

Grade 3: Moderate weakness, confined to a bed but able to move the limbs, brisk tendon jerks, sustained muscle clonus, muscle power grade 1-2, hyposthesia, or anesthesia.

Grade 4: Severe weakness, inability to move the limbs, paraplegia to extension or flexion, muscle power grade 0, total sensory loss, bowel and bladder incontinence.

History taking and examination are important for diagnosis. With a high index of suspicion, percutaneous CT-guided biopsy can be performed, to obtain bone tissue and abscess samples for acid-fast bacilli staining and culture. A biopsy can also allow the therapeutic drainage of paraspinal abscesses. Nucleic acid amplification tests, which have higher specificity and sensitivity and faster results, can also be performed. Histological findings showing features of caseating granuloma and giant cells can also assist the diagnosis. Hematological lab data can present with elevated erythrocyte sedimentation rates (ESRs) and usually display normal white blood cell counts [18-21].

The evaluation of spinal images can also provide an adequate diagnosis. Plain radiographs may show lytic lesions, the wedging of the anterior part of the vertebral body, the destruction of the intervertebral disc, and paravertebral shadow, with or without calcification, suggesting abscess formation. CT scans provide better bony details and allow the earlier detection of disease. The maintenance of disc height until late in the disease and abscess with calcification favor tuberculous lesion over pyogenic diseases. MRI is the gold standard for evaluating disk-space infections and osteomyelitis of the spine, allowing the visualization of cord compression [18-21].

Conservative anti-tubercular therapy, consisting of a 4-drug regimen (isoniazid, rifampicin, ethambutol, and pyrazinamide), is the usual course of spinal TB management. Many experts have suggested treatment for 9 to 12 months, using the directly observed treatment short-course (DOTS) method. Most patients can achieve pain relief and neurological improvements within 3 months of treatment onset. The rate of lesion recrudescence is approximately 2%–5%. Drug resistance has been noted to occur in 5%–10% of cases, requiring the use of second-line drugs, such as quinolone and aminoglycoside [18-21].

Although surgical intervention remains controversial, it is generally advised in patients with neurological deficits, including paraplegia with
acute deterioration, established or predicted spinal deformity with instability or pain, large paraspinal abscesses, non-diagnostic percutaneous biopsies, and poor response to medical therapy. Surgery can decompress the spinal cord, correct kyphosis, and result in more rapid pain relief. Some surgical interventions include anterior decompression, with debridement and posterior instrumentation for stabilization. However, laminectomy is contraindicated during typical anterior spinal tuberculosis because it makes the spine unstable [18-21].

Other management strategies include bed rest and the use of braces. However, the efficacy of these strategies remains questionable [18-21].

Overall, the prognosis is good, with almost all patients achieving improvements in neurological deficits and pain [18-21].

Conclusions

Paraplegia is a variable condition, and the extent of functional loss is correlated with the degree of spinal cord injury and the area that is affected. Immediate and proper treatment may result in the best chances of recovery. Therefore, the early diagnosis of the underlying cause is important. Though not commonly observed, these etiologies should be considered whenever unusual paraplegic situations are encountered.

References

非尋常病因之截癱：文獻回顧

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摘要

截癱是指脊髓的胸、腰或薑段中神經元的傷害，導致運動或感覺功能的損傷或喪失。一般而言，上肢的功能不受到影響，而其下半身受損的程度則依從傷的位置和範圍決定。以現今的醫療水平，截癱的病人在經過適當的治療與復健之後，仍有相當比例的患者能恢復生活獨立自主的功能，甚至重返工作崗位或學校進修。然而，截癱只是純表象而非病因，因此，早期的診斷顯得更重要。其造成的原因眾繁不及備載，症狀也是各式各樣，從常見的創傷到少見的感染都有可能。本篇文章列舉了五個不常見的原因，包括脊髓硬脊膜動靜脈瘻管、自發性脊椎硬腦膜外血腫、主動脈剝離、潛水減壓症與脊椎結核病，並且從病例的呈現到簡單討論個別病因的病生理機轉、症狀和治療等，希望藉此使各位讀者在遇到截癱的病人時可以有更多的鑑別診斷，從而提早進行治療。

關鍵詞：截癱、動脈剝離、硬脊膜外血腫、主動脈剝離、潛水減壓症、脊椎結核病

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