Intradural arachnoid cyst associated with cauda equina syndrome following lumbar puncture procedure: A case report

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SUMMARY

Intraspinal lesion is known to cause significant impairment that leads to deterioration in physical function and psychosocial disruption. Delay in detection and definitive intervention of symptomatic spinal neurological deficit often leads to significant morbidity and prolonged functional decline. We present a case of a 13-year-old girl who acquired an acute infection that was complicated with coagulation defect. She underwent a lumbar puncture procedure following an altered mental state. After prolonged mechanical ventilation in the acute phase, she developed critical illness polyneuropathy of all four limbs. Subsequently, she developed cauda equina syndrome secondary to intradural arachnoid cyst at L3/L4 level that was later identified by magnetic resonance imaging of the lumbar spine. She became wheelchair dependent post-acute phase of the disease. With intensive therapy and serial functional evaluation, she achieved significant independence in activities of daily living despite incomplete neurological recovery of the affected lower limb muscles.

INTRODUCTION

Spinal arachnoid cysts (SACs) are rare lesions that can produce concomitant compressive myelopathy through a mass effect on the spinal cord.[1] They commonly do not cause any symptoms and are a common incidental finding in the central nervous system imaging being performed for other non-related reasons.[2] Hence, the occurrence could be underreported in the literature. The aetiology remains unclear; it is ascribed that acquired SAC appears to result from previous trauma or inflammatory condition at the affected site whereas another majority of SAC are idiopathic and congenital.[3]

Lumbar puncture is widely known as a simple procedure that is commonly performed in the ward. It usually does not impose any hazardous complication in most of the patients subjected to it. In cases where lower limb weakness occurred after the procedure, early identification and therapeutic management have shown a reduction in residual impairment and morbidity.[4] We report a unique occurrence of an acquired intradural arachnoid cyst with cauda equina syndrome that resulted after a diagnostic lumbar puncture performed during acute leptospirosis, and how rehabilitation has contributed to the improvement of her functional independence despite the incomplete neurological recovery.

CASE REPORT

A 13-year-old girl who was previously well with normal physical function was admitted for acute confusion state following an episode of intermittent fever for 4 days, myalgia, headache and recurrent vomiting. On arrival, her Glasgow Coma Scale (GCS) was 11/15 (Eye:4, Verbal:1, Motor:6). Blood pressure was 148/79 mmHg, pulse rate was 135 beats per minute, temperature was 38.6°C, and oxygen saturation (SpO2) was 96% on room air. Physical examination revealed dehydration, neck stiffness, and hepatomegaly. Laboratory investigation showed thrombocytopenia, elevated creatine kinase, acute kidney injury, and liver derangement. The diagnosis of leptospirosis was confirmed with serology where Leptospira IgM was detected via ELISA test. Intravenous ceftriaxone and acyclovir was started. She was admitted to intensive care unit (ICU) for further management. Computed tomography scan of the brain revealed no abscess, hydrocephalus, or cerebral oedema. As the clinical scenario was suggestive of encephalopathy, lumbar puncture was performed on day 2 of admission (day 6 of illness) in the presence of thrombocytopenia with platelet count of 34 × 10^9/L but normal coagulation profile. The procedure was carried out in a full sterile manner using lumbar puncture needle size 18 gauge with no difficulty and no repeated puncture. Cerebrospinal fluid (CSF) opening pressure was 22 mmHg. CSF obtained appeared colourless. CSF full examination and microscopic examination showed no red blood cells, glucose of 6.0 mmol/L, and protein of 0.3 mg/ml. Initial phase post-procedure was uneventful as she had no immediate back pain or puncture site swelling.

At day 3 of admission, she became more restless and hypoxic, needing mechanical ventilation. Laboratory investigations at this point showed worsening of renal function, metabolic acidosis, severe liver transaminitis, and coagulopathy. On the following day, she developed right inguinal hematoma from venipuncture because of coagulopathy and thrombocytopenia resembling disseminated intravascular coagulopathy (DIVC). Two cycles of DIVC regime were

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Table I: Nerve conduction study and electromyography

Sensory study				
Nerve/Sites	**Latency Ms (Normal)**	**Amplitude µV (Normal)**	**Velocity m/s (Normal)**	
Right Median	2.75 (≤3.5)	13.6 (≥15)	47.3 (≥50)	
Right Ulnar	2.75 (≤3.1)	54.5 (≥10)	53.6 (≥50)	
Right Radial	2.40 (≤2.9)	38.5 (≥15)	56.3 (≥50)	
Right Sural	3.80 (≤4.4)	27.0 (≥15)	36.2 (≥40)	
Left Sural	4.15 (≤4.4)	5.4 (≥6)	33.7 (≥40)	
Motor study				
Nerve/Sites	**Latency ms (Normal)**	**Amplitude mV (Normal)**	**Velocity m/s (Normal)**	**F-Wave Ms (Normal)**
R Median	3.70 (≤4.4)	10.9 (≥4)	51.9 (≥49)	27.82 (≤31)
R Ulnar	2.60 (≤3.3)	4.7 (≥6)	56.8 (≥49)	28.00 (≤32)
R Radial	3.10 (≤2.9)	1.5 (≥2)	62.2 (≥49)	
Bilateral common peroneal CMAPs absent				
Bilateral tibial CMAPs absent				

EMG findings:
*Left tibialis anterior and left semi membranous showed fibrillation potentials and positive sharp waves. Right lumbar paraspinal muscles showed no spontaneous activity though this is a limited test to suggest root involvement.

*Electrophysiological evidence of bilaterally symmetrical axonal and length-dependent motor and sensory polyneuropathy

Table II: Neurological and functional progress during intensive rehabilitation

Time post neurological detection	Spinal nerve level	Week 2	Week 3	Week 4	4 weeks post-surgery
Lower limb myotomal (MRC)	L2	3/5	4/5	5/5	5/5
	L3	4/5	4/5	5/5	5/5
	L4	0/5	0/5	0/5	0/5
	L5	0/5	0/5	0/5	0/5
	S1	0/5	0/5	0/5	0/5
Anal tone		Lax	Lax	Lax	Lax
Voluntary anal contraction	S2-S4	Nil	Nil	Nil	Nil
Lower limb dermatome (pinprick)	L1–L3	2/2	2/2	2/2	2/2
	L4	1/2	1/2	1/2	2/2
	L5	0/2	0/2	1/2	1/2
	S1	1/2	0/2	0/2	0/2
	S2-S5	0/2	0/2	0/2	0/2
Ambulation		Wheelchair propelled by carer	Wheelchair-self propel	Walk with waking frame, not more than 100 m	Walk with walking frame more than 200 m
Modified Barthel Index (MBI)		31/100	43/100	68/100	80/100

MRC=Medical Research Council motor power grading system
Pin prick test scale: 0=not sharp, 1=less sharp, 2=same sharpness as compared to reference point

delivered. There was no swelling or bleeding at the lumbar puncture site. Her ICU stay was complicated with episodes of hospital-acquired infection resulting in worsening septicemia. She was intubated for 12 days with a total ICU stay of 22 days.

At day 22 of admission, she became more responsive and fully alert. Unfortunately, she was noted to have bilateral upper limb and lower limb weakness and numbness, which limited her physical activities. Limb examination revealed muscle hypotonia, hyporeflexia, and upper limb reduction in motor power of Medical Research Council grading 3/5 bilaterally in both proximal and distal muscle groups. Lower limb examination revealed proximal muscle power of 3/5 at hips and knees bilaterally and distal muscle power of 0/5 over the bilateral ankle. Deep tendon reflexes were markedly reduced at the knees and absent at the ankles. She was treated as critical illness polyneuropathy (CIP) at that point. She was then transferred to another tertiary hospital with dedicated rehabilitation services, 4 weeks after being discharged from the ICU when her medical crisis had resolved. Sensory examination following dermatomal distribution revealed a sensory level of L3 (knee region) where she manifested sensory deficit starting from L4 dermatome downwards. Per-rectal examination revealed lax anal tone with no voluntary anal contraction and loss of deep anal sensation. Apart from that, she also failed trial without catheter where there was no voluntary voiding after removal of catheter, despite water intake of 500 milliliters within 2 hours. Urodynamic study performed 1 week later revealed areflexic low-pressure bladder. Lumbar MRI revealed spinal intradural arachnoid cysts at L3 and L3/L4 levels causing displacement and compression of the descending nerve roots as shown in Figures 1 and 2. Gradient ECHO sequence revealed no evidence of hematoma as shown in Figure 1c.

Nerve conduction study (NCS) and electromyography (EMG) were first performed 8 weeks after she was discharged from the ICU as shown in Table I, for which the test revealed electrophysiological evidence of bilaterally symmetrical axonal and length dependent motor and sensory polyneuropathy. Lumbar paraspinals test did not show spontaneous activity to suggest lumbar nerve root involvement. Follow-up NCS performed 3 months later showed mild improvement of the neuropathy.

Rehabilitation

Intensive rehabilitation aiming to achieve neurological recovery and promote functional independence was carried out to enable her to return to school and optimal social function. She had good improvement of the upper limb neurology and function, where the power improved to 5/5 bilaterally. She was able to carry out her personal activities of daily living in bed with minimal assistance. Later, upon agreement for surgical intervention, she underwent laminectomy and marsupialisation at L3/L4 level at 5 weeks post-detection of the neurological manifestation. After the surgery, she continued the intensive rehabilitation which enabled her to achieve independent walking with a walking frame. Neurological, mobility, and functional (assessed using Modified Barthel Index) improvement of the lower limbs with intensive therapy was observed, as shown in Table II. She showed some sensory improvement after the surgery.

Because of the flaccid ankle with the absence of bilateral motor power, we provided her with a pair of solid ankle-foot orthosis (AFO) to maintain the ankle in plantigrade position to assist locomotion. As for bladder and bowel management, she was trained to perform clean intermittent self-catheterisation (CISC) four-hourly and stool manual evacuation once daily. Bladder and bowel continence was achieved. After 6 months of rehabilitation, she was able to walk independently, whilst wearing the bilateral AFO.

DISCUSSION

In patients who undergo complicated acute illness with extended bed rest, hypoactivity leading to multisystemic deconditioning especially involving the neurological and musculoskeletal system is not uncommon.[5] This patient was noted to have all four-limb weakness of lower motor neuron features after being bedridden for a prolonged period that gave rise to CIP. CIP classically develops in the setting of critical illness and immobilisation. The actual onset of the weakness was unknown. Nonetheless, there was no electrophysiological study of the neurological condition being performed early upon detection. Looking at the timeline of this patient's illness, Guillain-Barre syndrome or muscle necrosis from rhabdomyolysis were the possible differential diagnoses. Therefore, it would be ideal if NCS and EMG could be done at the onset of weakness identification

Concerning the available result from NCS and EMG investigation, the denervation changes seen in the lower limbs and not in the paraspinal muscles could be due to the CIP that was still present. The subsequent post-surgery improvement may also be due to the spontaneous ongoing recovery from CIP and only co-incidentally seen as a response following surgery, especially given that the L2 and L3 myotome had been showing progress from the beginning of intensive rehabilitation. Nevertheless, the findings and interpretation from NCS and EMG studies have some elements of operator-dependent and patient-dependent, where the invasive part of the EMG could cause discomfort and difficulty for the patient to relax the targeted muscle properly.

Despite the identification of sphincter involvement along the way, suspicion of intraspinal pathology was not raised early in this case, thus there was no imaging done initially. In the absence of spinal mechanical signs such as back pain, back swelling, or tenderness, the presentation of bilateral paraparesis, sensory loss, neurogenic bladder, and neurogenic bowel is always suggestive of intraspinal pathology. Neurological manifestations commonly cited in CIP are typically limited to peripheral nerves.[6] Adding on to the insights, this patient was asymptomatic of any spinal syndrome pre-morbidly. Coagulopathies are the most common causes of subarachnoid hematoma occurring at the spinal region where the aetiology may either be pharmacologically induced or a complication from systemic diseases constituting 40.5%.[7] The patient discussed in this report manifested spontaneous bleeding episodes at the right inguinal region from a femoral venous puncture site as well as left antecubital fossa. She also had elevated liver enzymes and thrombocytopenia. Hence, she was at high risk to develop spinal hematoma following a lumbar puncture even when the procedure was smooth. In this case, however, lumbar MRI investigation following the development of typical cauda equina syndrome revealed arachnoid cysts and no evidence of hemorrhage in gradient ECHO sequence.

Intradural SAC is rare and commonly does not cause any significant symptom.[2] There have been similar cases reported after clinical procedures or spinal trauma which are syringo-subarachnoid shunt operation, lumbar myelography, operation of an epidural spinal hematoma, and traumatic vertebral fracture.[1,8,9] Acquired SAC occurring after procedures such as lumbar puncture or spine surgery could be the result of inflammation and subsequent meningeal adhesions, where trauma to the inner dura layer causing arachnoiditis was the identified cause of cyst development.[1,9] Histologically, choroid plexus producing CSF were found in the cyst wall.[3] The clinical history of our patient and the level of the SAC indicated the possibility of it to be secondary to the lumbar puncture procedure she went through during the acute presentation of leptospirosis, similar to the cases reported by Kriss et al in 1997.[9]

In the case of reversible pathology, early intervention may contribute to neurological improvement leading to significant functional improvement. Regarding definitive management aiming for neurological recovery, as with any spinal cord compression syndrome, the time of onset to surgery is an important factor in achieving recovery after treatment. Delayed decompression intervention of more than 48 hours has been proven to have significant association with poor outcome, as also seen in our patient.[10] The increased functional independence score in Modified Barthel Index (MBI) shown in Table I after the surgery was

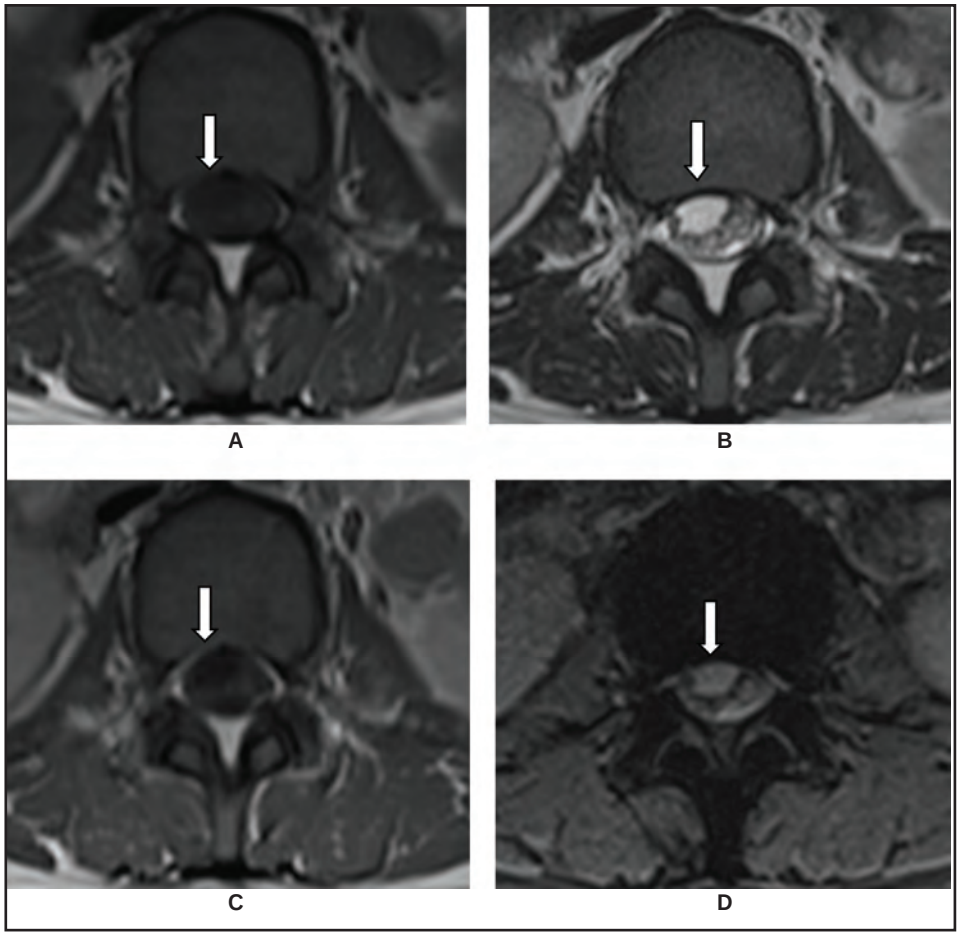

Fig. 1: Lumbar MRI – axial section. Axial lumbar MRI at L3 level revealed intradural cystic lesion at anterior aspect of the thecal sac which, (A) demonstrates low signal on T1-weighted image, (B) bright on T2-weighted image, and (C) not enhanced on post gadolinium administration. (D) Gradient echo sequence (GRE) revealed no evidence of hematoma.

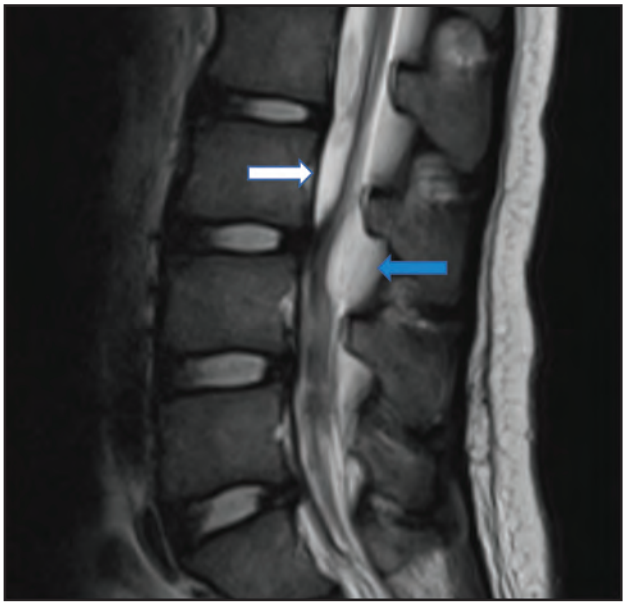

Fig. 2: Lumbar MRI – sagittal section. T2-weighted sagittal image showing another cyst seen posteriorly and slightly inferiorly at L3/L4 level (blue arrow). These cysts caused mass effect to adjacent nerve roots.

contributed by CISC training done post-surgery. She was scored 0/10 for the bladder control domain in the MBI assessment while she was on indwelling urinary catheter, and later achieved the score of 10/10 after she has become independent in managing her bladder with intermittent catheterisation.

This clinical case has raised some pertinent learning points for clinicians from its weakness and strength. Suspicion of an intraspinal pathology upon the manifestation of bilateral lower limb weakness and sphincter involvement was not raised early in the primary hospital despite the presence of classical signs resembling red flags of the spinal cord. In addition, the upper limb weakness has improved remarkably as compared to the lower limb severe weakness, prolonged lax anal tone, and neurogenic bowel and bladder. The failure to recognize the syndrome during the post-acute phase could be due to the concurrent presence of other neurological conditions such as delirium, reduced conscious level, and CIP. Besides that, reduced vigilance of the treating team after an exhaustive management of her severe acute illness is an important lesson to consider. Consequently, early electrophysiological investigation to enable follow-up comparison was also missed. We strongly suggest that all cases with bilateral limb neurological manifestation associated with sphincteric involvement must raise the suspicion of a spinal cord pathology and immediately be subjected to spinal imaging. Furthermore, CIP is rarely associated with bowel or urinary sphincter involvement.[6] Nevertheless, this case has brought the insight that collective interdisciplinary effort in rehabilitation management is one of the key factors in aiming to increase the patient's functional independence level.

CONCLUSION

Localisation of spinal cord pathology can be delayed due to deterioration of an acute medical illness and when other neurological condition such as CIP is also present. Delay in the detection and intervention of symptomatic spinal pathology leads to significant morbidity and reduced functional outcome. All clinicians must be able to recognize the red flags of spinal pathology and the subsequent action required. It is crucial that such patient's neurological progress to be closely monitored right from the onset of symptoms and evaluated vigilantly using appropriate modalities. Additionally, intensive rehabilitation is essential to provide significant functional improvement and promote independence in activities of daily living.

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