

A rare cause of Cauda equina syndrome: Epidural high grade primary non-Hodgkin lymphoma

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ABSTRACT

Cauda equina syndrome (CES) may be caused by herniated disc, tumor, trauma, and spinal infections. However, CES due to epidural high-grade non-Hodgkin lymphoma (NHL) is very rare. Up to our knowledge, few cases have been reported in the literature. We report a case of epidural high-grade NHL presenting as CES. A 55-year-old man presented with CES caused by extradural compression by primary NHL. The patient underwent an L4-L5 laminectomy. The operative findings were suggestive of well-demarcated epidural tumor. The final histopathological diagnosis revealed epidural high-grade NHL. NHL causing CES is rare. This report highlights the importance of keeping afresh the various causes of CES for prompt diagnosis and management.

Key words: Cauda equina syndrome, cauda equina tumours, non-Hodgkin lymphoma, timing of surgery

INTRODUCTION

Cauda equina syndrome (CES), is a combination of signs and symptoms resulting from affection of the nerve roots in the Cauda equina. Typical manifestations associated with the disorder can be low back pain, unilateral or bilateral sciatica, bilateral weakness of the lower extremities, saddle or perianal hypoesthesia or anesthesia, sexual impotence, together with rectal and bladder sphincter dysfunction.^[1] The most common cause of CES is a intervertebral disc prolapse. But rare causes of CES do exist which include fractures, tumors, infection, spontaneous spinal hematoma, etc. Knowledge about these rare causes of Cauda equina will help the orthopedic surgeon in making proper decision and plan the surgery.

Non-Hodgkin lymphoma (NHL) rarely involves the central nervous system. When it does it most commonly affects the brain (2% of primary and 7-9% of metastatic brain

tumors) and only rarely directly infiltrates the spinal cord or Cauda equina. In series of Cauda equina tumors, the overall incidence of lymphoma is less than 2%. However, there have been several case reports of direct lymphomatous involvement of the Cauda equina, both by primary localized, disseminated, and recurrent NHL.^[2]

We report here a case of epidural high-grade NHL presenting as CES causing extra-dural compression with a clinical feature specific for Cauda equina tumors. We would like to highlight the importance of keeping various causes afresh.

CASE REPORT

A 55-year-old male presented with progressively increasing low back pain since past 40 days, which worsened on recumbency and sneezing, with recent complaints of urinary incontinence. The patient presented to us 2 weeks after the onset of urinary complaints. His medical history was unremarkable. The patient complained of low back pain with bilateral lower limb radiculopathy with urinary incontinence since 15 days. There was no history of bowel disturbance or weakness of lower limbs. On neurological examination there was bilateral extensor hallucis longus weakness (grade 4/5), absent ankle jerk, decreased anal sphincter tone on digital rectal examination, absent

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bulbocavernous reflex, and decreased sensation in the perianal area. Bilateral straight leg raising test was free and bilateral knee reflex was normal. Upper limb neurological examination was normal. His general and systemic examination was normal routine biochemical investigations, chest and lumbosacral X-rays were normal. HIV serology was negative. Magnetic resonance imaging (MRI) of the lumbosacral region showed a mild L4-L5 disc prolapse with a hypo intense mass in both T1W and T2W images arising from posterior aspect compressing the thecal sac, most probably a hypertrophied ligamentum flavum, as reported by a senior radiologist. The most striking feature was the clinical presentation, of aggravation of symptoms on recumbency and partial relief on assuming a sitting posture; hence, he would be sitting continuously. The pain was so severe that the patient could not lie down even for few minutes; we had to sedate the patient for the MRI. This clinical feature is considered specific for Cauda equina tumors. Not satisfied with the MRI report a contrast enhanced MRI scan was done, which showed a L4-L5 contrast enhancing posterior epidural mass compressing on the dural sac measuring 2.5 cm craniocaudally and 1 cm anteroposteriorly at the L4/5 level [Figure 1]. The patient was operated 3 weeks after the onset of his bladder symptoms. The patient underwent L4-L5 laminectomy with en-masse excision of the tumor. Intra-operatively there was a friable dark tissue in the epidural region. On histopathological examination a high-grade NHL was noted, [Figure 2]. Immediately after surgery, he had complete relief of his backache and was able to lie down comfortably. His extensor hallucis longus weakness persisted, which was not symptomatic. His urinary incontinence did not improve for which he was on condom catheter. After suture removal the patient was referred to an oncologist for further management of NHL. Further investigations did not reveal any other foci. The patient was started on chemotherapy

for NHL. Patient received CHOP regimen and completed 6 cycles. After 1 year of follow up patient is asymptomatic, urinary incontinence has not improved for which he is on condom catheter. PET scan at 1 year follow up shows no evidence of active disease in the body.

DISCUSSION

With this case report we would like to highlight the importance of knowledge on rare causes of CES, specific clinical features of Cauda equina tumors, and the significance of timing of surgery in CES.

CES is itself rare. Most of the cases are due to large central lower lumbar prolapsed intervertebral disc. More than 50 causes of CES have been listed in the literature which include fractures, tumors, infection, spontaneous spinal hematoma, etc.^[1,3,4] Though Cauda equina tumors have been reported, NHL causing CES is rare.

Several studies support the critical importance of timing of surgery, and suggest CES should be operated within 6 h of onset of symptoms. Literature review showed that patients operated within 6 h or operated within 48 h showed no difference in outcome.^[5-7] However, recent studies suggest that timing of surgery is not the prognostic factor in the recovery of bladder functions. It is the severity of bladder involvement at the time of surgery, the most important prognostic factor in the recovery of bladder functions.^[8] Though some recent reports suggests that CES is not an emergency as thought earlier,^[8] it is important to come at a definitive diagnosis quickly. Thus having a thorough understanding of the clinical presentation, which in this case pointed toward Cauda equina tumor, and getting the necessary investigations done in time for early decompression would affect the final outcome. The patient presented to us 2 weeks after the onset of urinary

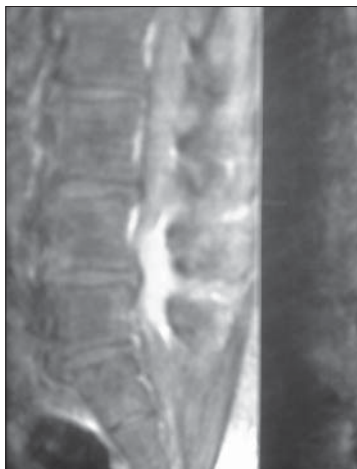


Figure 1: Contrast magnetic resonance imaging of the lumbosacral spine showing the contrast enhanced mass

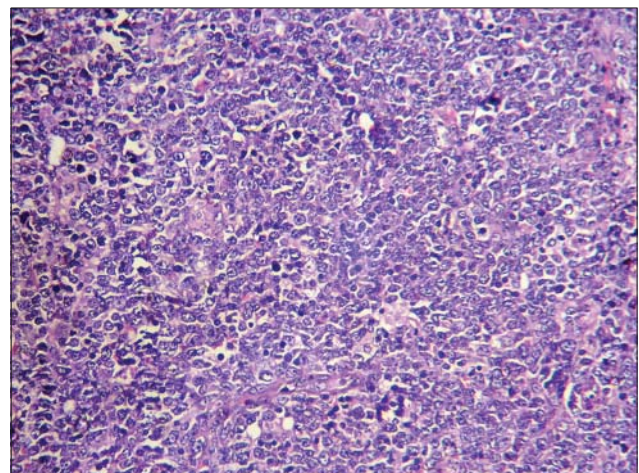


Figure 2: Histopathological picture showing high grade non-Hodgkin lymphoma

incontinence. Had he presented immediately after the onset may be the outcome would have been more favorable. Perceived delays in diagnosis and treatment may result in litigation due to the devastating impact of disordered bowel, bladder, and sexual function on quality of life. Surgery under optimum conditions, by experienced surgeon and with proper planning may give the best results.^[8] This case report shows the importance of keeping the causes of CES afresh to arrive at an accurate diagnosis in time.

CONCLUSION

In conclusion, we report a rare epidural high-grade NHL causing extradural compression and leading to CES. The knowledge of the clinical presentation, and the necessary conventional radiological studies and MRI with contrast enhancement allows for timely diagnosis of the cause and that post-operative recovery can be expected if unnecessary delay is avoided.

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