

Dumbbell-shaped Hodgkin's Disease with Cauda Equina Compression Mimicking a Herniated Inter-vertebral Disc, A Case Report

Jen-Chung Liao, MD; Tsai-Sheng Fu, MD; Wen-Jer Chen, MD; Shih-Ming Jung¹, MD

Hodgkin's disease may involve the spine as a setting of the advanced disease. An initial manifestation of Hodgkin's disease in spine is extremely rare and the major involved sites usually are the thoracic or cervical spine. The mechanisms of pathogenesis for the formation of an epidural mass during Hodgkin's disease are hematogenous dissemination from nodal sites or local infiltration of lymphomatous tissue. We document here a case of a 16 year-old boy who suffered from incomplete voiding due to dumbbell-shaped retroperitoneal Hodgkin's disease with cauda equina compression. He was successfully managed using surgery and adjuvant chemotherapy. Although lymphadenomatous tissue responds well to radiotherapy and chemotherapy, the role of surgery in this case was to achieve immediate nerve tissue decompression and to obtain an adequate specimen for pathological diagnosis. Magnetic resonance imaging (MRI) is a non-invasive and helpful tool when detecting spinal and paraspinal lesions and we emphasize that spinal MRI should be performed without delay if there is persistent back pain or sciatica. (*Chang Gung Med J* 2007;30:458-63)

Key words: dumbbell-shaped tumor, Hodgkin's disease, cauda equina compression

The frequency of Hodgkin's disease (HD) with spinal cord compression is rare (5%) and the spine is usually a setting of the advanced disease.⁽¹⁾ An initial manifestation of HD in the spine is extremely rare (0.2%). The major site of involvement is usually the thoracic spine.⁽²⁾ We describe here a 16-year-old boy who initially presented with impending cauda equina syndrome due to retroperitoneal HD with epidural invasion. The patient was successfully treated with surgery and chemotherapy.

CASE REPORT

A 16 year-old boy suffered from low back pain with radiation to right lower extremity and was diagnosed as a herniated inter-vertebral disc (HIVD) at

local clinic. Pelvic traction and rehabilitation programs were arranged there. This boy was admitted to our emergency department three months later due to persistent low back pain with right sciatica. He could not find a painless position. Perianal numbness had been noted for one week, but anal sphincter function was normal. Although there was no urine retention or incontinence, urine dribbling after voiding happened off and on. A physical examination revealed sensory deficit from right L1 dermatome downwards together with slight motor weakness on right ankle dorsiflexion and right big toe dorsiflexion. A right straight leg raising test was positive at 30 degree elevation.

Emergency plain lumbar spine radiographs showed a soft tissue shadow over right psoas muscle

From the Department of Orthopedics Surgery; ¹Department of Pathology, Chang Gung Memorial Hospital, Taipei, Chang Gung University College of Medicine, Taoyuan, Taiwan.

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Correspondence to: Dr. Jen-Chung Liao, Department of Orthopedic Surgery, Chang Gung Memorial Hospital, 5, Fusing St., Gueishan Township, Taoyuan County 333, Taiwan (R.O.C.) Tel: 886-3-3281200, ext. 2163; Fax: 886-3-3278113; E-mail: jcl1265@cgmh.org.tw

and loss of lumbar lordosis (Fig. 1). A retroperitoneal tumor was first impressed. Due to the neurological symptoms affecting the lower extremities, magnetic resonance imaging (MRI) of lumbar spine was arranged for this patient. Magnetic resonance imaging of spine revealed a huge retroperitoneal mass and lumbar epidural mass that was compressing the cauda equina (Fig. 2). The patient underwent a computed tomography-guided biopsy of the retroperitoneal mass one day later. Posterior surgical decompression was performed on the same day. The patient underwent lumbar laminectomy and a firm yellowish mass encircling the cauda equina was found. The epidural mass also occupied the bilateral L3, L4, L5 roots with extensions to the foramina. Although the tumor mass was not excised completely, the bilateral nerve roots were loose after decompression. The patient was encouraged to sit one day postoperatively; the hemovac was removed two days later and he started ambulation. The patient regained bladder function and the right sciatica regressed.

A histological analysis of a specimen from the epidural mass showed multiple nodules of lympho-

cytes, eosinophils, plasma cells and large cells with prominent nucleoli and this favored a nodular sclerosing Hodgkin's lymphoma (Fig. 3). Histological analysis of the retroperitoneal lesion showed numerous eosinophils, histiocytes, plasma cells and scattered



Fig. 1 Plain radiographs at emergency room shows a soft tissue shadow (arrow) over right psoas muscle.

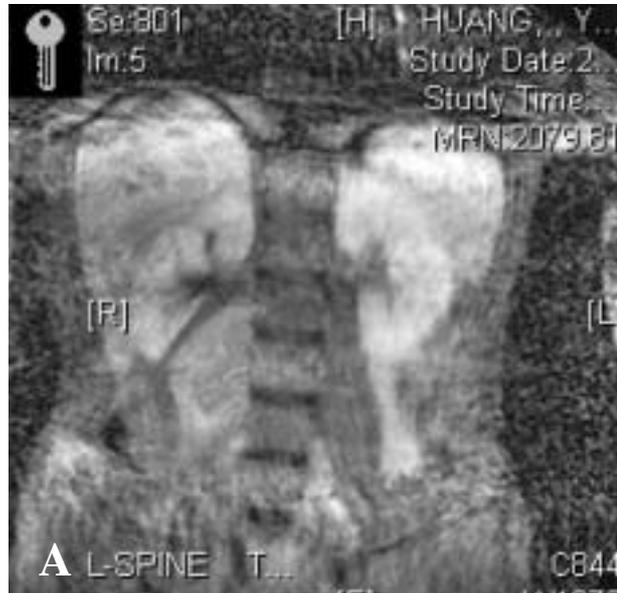


Fig. 2 Coronal view of magnetic resonance image shows a huge mass occupies right lumbar paravertebra area (A). T2-weighted sagittal spin-echo image reveals a soft mass occupies epidural space and compression of cauda equina (B).

large cells that were suggestive of Reed-Sternberg cells (Fig. 4). Immunohistochemical staining was positive for CD 15 antigen and CD 30 antigen, but was negative for CD3, CD20 and LMP1 antigens. Therefore Hodgkin's lymphoma was confirmed as both the morphological and immunological phenotypes are consistent with Hodgkin's disease.

Postoperative clinical evaluation revealed no cervical or inguinal lymphadenopathy. Thoracic and abdominal computerized tomography (CT) revealed no hepatosplenomegaly and no evidence of intra-abdominal or mediastinal lymph node enlargement.

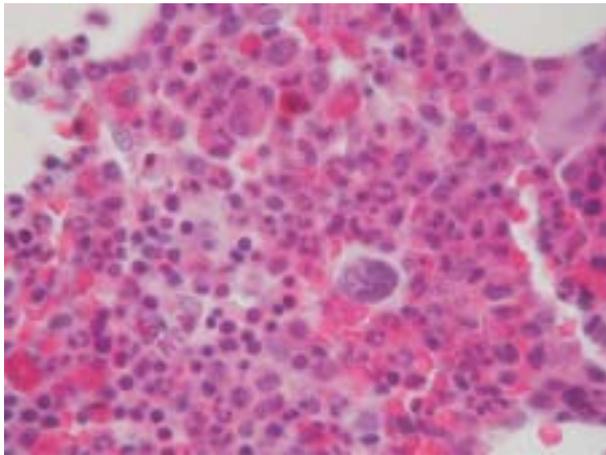


Fig. 3 Histologic resection of specimen from epidural space discloses polymorphous cellular infiltration with lymphocytes, eosinophils and large cells with prominent nucleoli.

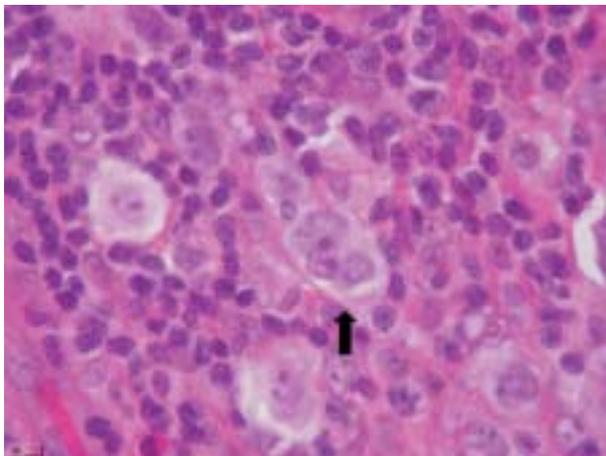


Fig. 4 Histologic analysis of retroperitoneal mass biopsy reveals numerous eosinophils, histiocytes, plasma cells and scattered large cells suggestive of Reed-Sternberg cell (arrow).

However, a pelvic CT scan showed a mass at right iliac crest. The patient was finally staged as suffering from stage 4 nodular sclerosis Hodgkin's diseases.

The patient was transferred to the pediatric hematology department and underwent chemotherapy. He received eight courses of Epirubicin, Bleomycin, Vinblastin and Dacarbazine (EBVD) from November 2004 to August 2005. A follow up MRI after the eight courses of chemotherapy showed an absence of any epidural or retroperitoneal mass (Fig. 5). At the last follow up, 12 months after surgery, the patient remains fully ambulatory without lower extremities weakness and has normal bladder function. His physical examination shows a negative straight leg test.

DISCUSSION

Spinal cord compression caused by a malignant lymphoma is generally a late manifestation of the illness.⁽³⁾ There are about three times more patients with non-Hodgkin's lymphoma than with Hodgkin's disease.⁽⁴⁾ Spinal cord compression as an initial pre-



Fig. 5 T2-weighted sagittal spin-echo images after 8 courses of Epirubicin, Bleomycin, Vinblastin, Dacarbazine (EBVD) shows no evidence of mass in lumbar spinal canal.

sensation of lymphoma is unusual. A handful of case reports have described patients with Hodgkin's disease who have an initial presentation with spinal cord compression.^(2,5,6) The major involved site is the thoracic or cervical spine.

Several mechanisms of pathogenesis for the formation of an epidural mass in Hodgkin's disease have been suggested. Hematogenous dissemination from nodal sites or infiltration of lymphomatous tissue from the vertebral body have been proposed.⁽⁷⁾ In the current case, histological examination of both the retroperitoneal mass and the cauda equina epidural mass all shows classic Reed-Sternberg cells. The retroperitoneal space may be the original site of this Hodgkin's disease. Contiguous spread through intervertebral foramen into cauda equina epidural space followed by dissemination to right iliac area hematogenously seems the most likely mechanism of pathogenesis.

Lymphadenomatous tissue responds well to radiotherapy and chemotherapy. A good clinical response and improved neurological function after conservative treatment for Hodgkin's disease have been described in previous reports.⁽⁷⁻¹²⁾ The roles of surgery are decompression, stabilization and obtaining a specimen. Surgery is usually reserved for those patients where there is a failure of conservative treatment or for those with an initial presentation that involves a severe neurological deficit due to epidural compression or bony destruction. Initial surgical management in cases of severe neurological deficit is able to provide a good functional return.^(2,5,6,13)

Cauda equina syndrome is a complex of low back pain, bilateral sciatica, saddle anesthesia and motor weakness in the lower extremities with rectal and urine incontinence. A herniated disc, a tumor, hematoma formation or a traumatic vertebra with cauda equina compression may cause cauda equina syndrome. A significant improvement in motor and sensory deficit as well as rectal and urinary function occurs in patients with cauda equina syndrome who undergo early decompression surgery.⁽¹⁴⁾ The initial presentations of this patient was low back pain with sciatica and incomplete voiding due to cauda equina compression by an epidural mass. Laminectomy can provide nerve tissue decompression immediately and in the process allow an adequate specimen to be obtained for pathological diagnosis. However, surgery is not able to eradicate the tumor mass com-

pletely. Local control of tumor can be achieved by adjuvant local irradiation or the metastatic mass can be controlled by chemotherapy. After eight cycles of chemotherapy, the right iliac and retroperitoneal mass was no longer present on the MRI at the last follow-up.

To author's knowledge, the case described here is only the third case report of a patient with Hodgkin's disease whose initial presentation is cauda equina compression by an epidural mass. Toprak et al. presented a 20-year-old male with initial presentations of back pain and radiation to the legs. An MRI showed an epidural mass compressed cauda equina, hepatosplenomegaly and multiple lymphadenopathy. Hodgkin's disease was confirmed by bone marrow and lymph node biopsy. The patient was successfully treated with chemotherapy.⁽⁷⁾ Riffaud et al. reported a 14-year-old patient who experienced low back pain and urine incontinence; imaging studies showed multiple lymphadenopathy and an epidural mass compressed cauda equina. This patient underwent emergency surgical decompression. Hodgkin's disease was confirmed by histopathological examination of the surgical specimen. Adjuvant radiotherapy and chemotherapy were given and successful treatment was achieved finally.⁽¹³⁾ Our case seems similar to these cases; however, there are still some differences between them. In our case, only a huge retroperitoneal mass with cauda equina invasion and right iliac metastasis was seen and no other organs had tumor invasion. Furthermore, there is also no cervical, mediastinal or intra-abdominal lymphadenopathy in our case.

MRI is a noninvasive method and helpful tool for the detection of spinal and paraspinal lesions. We emphasize that spinal MRI should be performed without delay if there is persistent back pain or sciatica. Early detection of spinal epidural lymphomas is able to prevent complications due to spinal cord compression such as paraplegia or sphincter dysfunction and such lymphomas can be successfully managed with conservative treatment.

We conclude that Hodgkin disease involving epidural space as the initial presentation is very rare. The optimal treatment in this situation is multidisciplinary. We chose to treat this patient with early surgical intervention because of his impending cauda equina syndrome. Adjuvant chemotherapy was essential because of his iliac crest metastasis. This

patient represents an unusual case of retroperitoneal dumbbell-shape Hodgkin's disease with initial presentation involving cauda equina symptoms.

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啞鈴型哈杰金氏淋巴疾病壓迫脊髓馬尾，仿似椎間盤突出 — 病例報告

廖振中 傅再生 陳文哲 容世明¹

哈杰金氏疾病可能會侵犯脊椎，不過通常是在末期。哈杰金氏疾病一開始的表現以脊椎侵犯壓迫的病例非常少見，主要好發在胸椎及腰椎。哈杰金氏疾病形成硬膜外腫塊的病理機轉可能是從遠處淋巴節經由血行，或是淋巴組織局部浸潤。我們報告一個病例，這是一個十六歲少年，由於後腹腔哈杰金氏淋巴腫瘤穿越神經孔，壓迫腰椎脊髓導致馬尾壓迫症候群。病人在接受手術和化學治療之後，疾病獲得成功的治療。雖然淋巴組織對於放射線治療以及化學治療的反應很好，手術對於此病人的功能主要在於使神經組織獲得立即的減壓，同時取得足夠的標本做病理診斷。核磁共振是一種非侵襲性且有效的去檢測脊椎或脊椎旁病變的工具。我們也強調如果有持續性的背痛或坐骨神經痛，應該使用核磁共振去偵測脊椎病變。(長庚醫誌 2007;30:458-63)

關鍵詞：啞鈴型腫瘤，哈杰金氏淋巴疾病，脊髓馬尾壓迫

長庚紀念醫院 台北院區 骨科部，¹病理科；長庚大學 醫學院

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通訊作者：廖振中醫師，長庚紀念醫院 骨科部。桃園縣333龜山鄉復興街5號。Tel: (03)3281200轉2163; Fax: (03)3278113;

E-mail: jcl1265@cgmh.org.tw