

Paraplegia by spinal cord compression as the initial manifestation of Hodgkin's disease: a case report

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Summary. The 90% of Hodgkin's disease (HD) cases are originated by lymphnodes whereas 10% by extranodal regions as epidural space. Neurologic complications of HD can be classified as directly resulting from the disease or indirectly originated from the disease or from its treatment. Patients very rarely present with spinal cord compression (SCC) due to epidural HD. Few cases of HD with such presentation have been reported in the literature. Primary spinal extradural HD with no further organ involvement is extremely rare. We report a case of a child with SCC as initial and unique presentation of HD.

Key words: Hodgkin's disease, spinal cord compression, laminectomy, paraplegia

Background

Hodgkin's disease (HD) originates most commonly in an age range of 16-34 years, with another subgroup presenting later in life. The incidence of HD is estimated at 3.0 per 100.000 in the US. Prior viral infections, especially with EBV and HIV, associated with a severe form, may also play a role in subtypes of HD (1).

The 90% of HD cases are originated by lymph nodes whereas 10% by extranodal regions as epidural space. Patients rarely present with SCC due to epidural HD. Limited cases with such presentation have been reported in the literature (2-12).

Neurologic complications of HD can be classified as directly resulting from the disease or indirectly originated from the disease or from its treatment (6).

Primary spinal extradural HD with no further organ involvement is extremely rare. (2-5).

We report the case of a child with SCC as initial and unique presentation of HD.

Case report

XY, an 8 years-old African male, came to our observation for spastic paraplegia. One year before the child began to accuse back pain and subsequently he presented difficulty in walking, sensory loss and autonomic dysfunction (urinary and faecal incontinence). He was therefore admitted to the local Hospital, where he was submitted to spinal radiographs (negative) and cerebrospinal fluid analysis (which showed only an increase in protein). He received steroid therapy (prednisolone) for 3 months and for suspected tuberculosis (TBC), he started therapy with isoniazid and rifampicin continued for 10 months.

When he was admitted to our Department, at the neurological examination the child appeared oriented in time and space, with excellent understanding, fluid language; he did not report visual disturbances and cranial nerves deficits; arm's strength and tone and neck muscles were within normal limits; OTR present and symmetrical, lower extremity hyperreflexia. The exam

revealed a sensory deficit below the L1 dermatome; signs of spastic diplegia of the legs with the impossibility or extreme difficulty in extension; atrophy of the gastrocnemius; more evident clonus at the left leg; a right heel eschar; difficulty in bilateral dorsiflexion of the feet, with Achilles tendon's retraction. He presented urinary and faecal incontinence and he was unable to maintain a sitting position without support and to walk.

Spinal MRI revealed an infiltrating probable infectious lesion on the epidural space, from D4 to D6 and from D8 to D10 levels (Fig. 1a). Serological tests were negative for acute infections by *Chlamydia pneumoniae*, *Mycoplasma pneumoniae*, *Toxoplasma Gondii*, *Brucella*, *Treponema pallidum*, TBC, CMV, HSV1, EBV, VZV, measles, mumps, Parvovirus B19, Adenovirus, Enterovirus, HIV, HCV and HBV. We decided, in accordance with neurosurgeons, to perform surgical decompressive laminectomy and to remove extradural compressive lesion in the dorsal posterior D4-D10 levels. Pathology examination demonstrated the presence of classic nodular sclerosing HD, with expression of CD20.

The TC scan of the brain, thorax and abdomen, performed for staging, were normal.

After the surgery, following physiotherapy and treatment with muscle relaxants (baclofen), the child presented progressive decrease in legs hypertonicity and recovery of sitting position with neuroradiological improvement (Fig. 1b).

The therapeutic process was continued voluntarily in another Hospital

Discussion

The most common causes of SCC are tumours and less frequently abscesses and granulomas.

Sarcomas (rhabdomyosarcoma, osteosarcoma and other) account more than fifty percent of spinal cord involvement in childhood; the remainder are caused by neuroblastoma, germ cell tumour, lymphoma, leukaemia and drop metastasis of CNS tumour (table 1) (14).

A picture similar to SCC may have other causes in children with cancer including infection, radiation myelopathy, spinal cord infarction and intraspinal hematomas due to a coagulation problem.

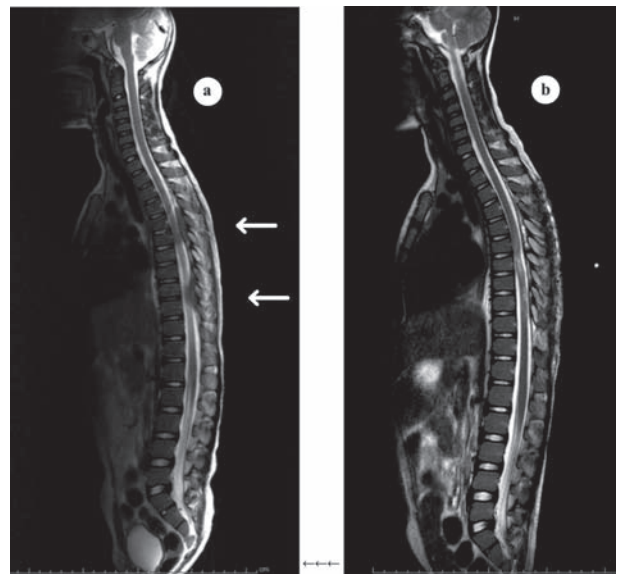


Figure 1. Spinal MRI of lesions on the epidural space, from D4 to D6 and from D8 to D10 levels before (a) and after (b) surgical decompressive laminectomy. Arrows indicate the infiltrating lesions.

Radicular syndromes as first signs of lymphoma are exceptional. The thoracic segment is the most common site of spinal epidural infiltration in patients with HD (15).

Symptoms of SCC include back pain (progressive, worse when lying flat, and improved with walking), weakness, sensory loss, autonomic dysfunction (painless urinary retention, faecal incontinence, and impotence), and ataxia. Signs of SCC result in sensory level, paraparesis, hyperreflexia and presence of the Babinski response.

A simple four-point scale (9) was used to grade functional ability retrospectively:

- Grade 1: normal or minor sensory symptoms only, with no motor or sphincter impairment;
- Grade 2: assistance needed to ambulate;
- Grade 3: inability to bear weight;
- Grade 4: paraplegia.

None of the available data clearly define the optimal treatment for HD presenting with SCC. Some authors advocate a multidisciplinary approach, including local radiation therapy with or without surgical decompression; others prefer chemotherapy as the primary treatment (15-17).

Table 1. Neoplastic causes of SCC in children (modified by Nicolin 14)

Pathology	Causes of SCC (% of all cases)
Neuroblastoma	27
Ewing's sarcoma	23
Rhabdomyosarcoma	15
Osteosarcoma	12
Other soft-tissue sarcoma	7
Hodgkin's Disease	5
Germ cell tumour	4
Non Hodgkin's Lymphoma or Leukaemia	3
Wilms Tumour	2
Other	1

Standard emergency treatment is to administer dexamethasone. If a cord-compressing space occupying lesion is confirmed, a decision between immediate surgical decompression vs. radiotherapy or chemotherapy has to be made. This decision will be influenced by a number of factors, principally:

- a) the availability of a histological diagnosis;
- b) the anticipated response of the specific tumour type to radiotherapy or chemotherapy;
- c) the extent of neurological deficit and
- d) the speed of its progression.

The immediate goal is to restore neurological function (14).

The surgical approach is decompressive laminectomy, which consists in the removal of the posterior arch of the medullary canal to a level above and one below respect to the seat of SCC. Following this approach, the tumour is not removed, but the laminectomy allows the relaxation of the spinal cord from the neoplastic mass front. In Klein and Raffel's retrospective studies the patients undergoing to surgery treatment had reported a best neurological recovery compared to exclusive chemotherapy or radiotherapy (16-17).

Surgery is reserved for neurologic compromise, radiation failure, spinal instability or uncertain diagnosis.

Correale et al. stated that patients having laminectomy combined with chemotherapy and radiotherapy (favourable results in five of nine patients), and

patients having only chemotherapy and radiotherapy (favourable results in 11 of 12 patients) did not differ in their prognosis (7). In Hodgkin patients with spinal findings, the general attitude is to perform decompression with laminectomy (6).

At once, in our case, we decided to perform a decompressed laminectomy for the uncertain diagnosis and to obtain early functional recovery. We have not performed steroid therapy because, at the beginning of the symptoms, it had already been administered by the African colleagues without improvement. As the child had been voluntarily removed by his mother, we could not complete the diagnostic process and begin the appropriate chemotherapy.

Conclusions

The ultimate outcome for these children depends on the extent of the cancer at diagnosis and the response to treatment, but their ultimate "quality of life" depends on the long-term neurological sequelae. Effective team work and carefully planned and decisive action is the key for the best outcome for these children.

Early diagnosis and treatment of these complications are of utmost importance in order to avoid further physical disabilities of these patients.

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