Hodgkin’s Lymphoma Presenting As Extradural Spinal Cord Compression

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Abstract

Objectives: Hodgkin’s lymphoma presenting with spinal cord compression is rare. Reports estimate that only 5% of patients with Hodgkin’s lymphoma have spinal cord compression. The objectives of this study were: (1) To review the histology of all cases of Hodgkin’s lymphoma causing spinal cord compression. (2) To correlate the findings with the clinical details - lymphadenopathy, hepatosplenomegaly, and marrow involvement.

Method: This was a retrospective study, the period of study being between 1987-2002. All cases were taken from the histopathology record files, pathology department, NIMS, Hyderabad. The clinical profile of each case was noted and the histology reviewed. When necessary, immunohistochemistry with the necessary markers was performed.

Result: There were 12 cases, of which 11 were males, and one was a female. The patients ranged from 8 to 62 years of age, with a median age of 32 years. The cord involvement was at different levels - thoracic (8), lumbar (2), and cervical (2).

Conclusions: The histologic diagnosis of Hodgkin’s lymphoma presenting as cord compression requires awareness of the condition. The close differential diagnoses include non-specific inflammation, non-Hodgkin’s lymphoma, tuberculosis and eosinophilic granuloma. Detailed clinical evaluation is essential for interpreting the histology.

INTRODUCTION

Hematological malignancies like non-Hodgkin’s lymphoma and multiple myeloma sometimes present with spinal cord compression. However, Hodgkin’s lymphoma presenting like this is rare. Only 5% of all Hodgkin’s lymphoma cases develop spinal cord compression, and only 0.2% cases, spinal cord compression occurs as the initial presentation. The objectives of this study were (1) To review the histology of all cases of Hodgkin’s lymphoma causing spinal cord compression; (2) To correlate the histology with the clinical details with emphasis on features like lymphadenopathy, hepatosplenomegaly, and marrow involvement.

METHODS

This was a retrospective study, carried out in the pathology department, Nizam’s Institute of Medical Sciences, Hyderabad. The study period was between 1987-2002. All cases of Hodgkin’s lymphoma causing spinal cord compression, were retrieved from the histopathology files. The complete clinical and imageological details were noted. The histology of each case was reviewed. If the patient had a lymph node biopsy done, at any time, it was also reviewed. As part of staging procedure, bone marrow examinations were done in few cases, and these were also examined. In one case, immunohistochemistry with LCA (Leucocyte common antigen), CD15 and CD30 (kits supplied by DAKO), were also performed.

RESULTS

During the study period, there were 12 cases of Hodgkin’s lymphoma presenting with spinal cord compression, of which 11 were males and one female. The patients ranged from 8 to 62 years of age with a median age of 32 years. The symptoms varied depending on the level of spinal cord involvement. The neurologic symptoms included paraplegia, weakness of lower limbs, pain at the involved sites, bladder/bowel involvement, sensory loss. The sites of spinal cord involvement were thoracic spine (8 cases), lumbar spine (2 cases) and cervical (2 cases).

Imageology revealed that the bone was involved in six cases and not involved in six cases. There was vertebral...
body destruction in two cases, lytic lesion in one case and altered signal intensities in three cases.

The temporal association between diagnosis of spinal cord involvement and lymph node biopsy is given in Table 1. There were three cases previously diagnosed as Hodgkin’s lymphoma on nodal biopsy, who later developed spinal cord compression. There were five cases presenting with cord compression, with nodal lymphomas being detected at a later date. There were four cases diagnosed with cord compression, who did not have detectable lymph nodes, even on follow-up.

The histologic features noted in 12 cases is shown in Table 2. All the cases had a polymorphous population of cells, comprising of lymphocytes, plasma cells, histiocytes and eosinophils. All the cases also showed mononuclear Reed-Sternberg cells. Ten cases showed classic binucleate Reed-Sternberg cells. For one case, immunohistochemistry was performed. This was a case which showed a polymorphous cell infiltrate, few mononuclear large cells but there were no binucleate Reed-Sternberg cells. Further, this patient did not have a past history of Hodgkin’s lymphoma and an accurate tissue diagnosis was essential. The lesion showed strong LCA positivity, focal CD15 positivity in the large cells and CD30 was negative.

As part of staging procedure for Hodgkin’s lymphoma, bone marrow examination was done in seven of the 12 cases. Of the seven cases, four showed marrow involvement by Hodgkin’s lymphoma, indicating stage IV disease.

**DISCUSSION**

Only 5% of cases of Hodgkin’s lymphoma may develop spinal cord compression. In only 0.2% cases, cord compression is the initial presentation. Hodgkin’s lymphoma within the spinal cord most often occurs as a complication of widespread dissemination or in concert with disease relapse. It is thought that the lymphomas usually enter the epidural space by contiguous spread from a paravertebral mass through the vertebral foramen and expanding into the epidural space. These paravertebral masses could be mediastinal or retroperitoneal lymph nodes. The vertebral bodies are often spared, hence plain films are often normal, in epidural spinal cord compression, secondary to lymphomas.

In Hodgkin’s lymphoma, epidural involvement is quite extensive with more than three spinal levels involved. In this study, there was vertebral body destruction in six of the 12 cases. Apart from this mode of entry, it is suggested that the tumor may arise de novo, from extradurally located lymphoid rests. In the present study, three cases were diagnosed as Hodgkin’s lymphoma on lymph node biopsies, prior to the cord compression. In five cases, the spinal cord compression occurred first, followed by tissue diagnosis of Hodgkin’s lymphoma. These were later detected to have lymphadenopathy, and the diagnosis confirmed. However, in the remaining four cases, there was no detectable lymphadenopathy on follow-up, after the primary diagnosis was made. These cases may either be considered to have Hodgkin’s lymphoma involving the spine, of de novo origin; or the search for the nodal lymphoma was not adequate.

It is reported that in the spinal cord, the thoracic segment

![Fig. 1: Section shows extradural soft tissue infiltrated by polymorphous population of cells (H & E x 100).](image1)

![Fig. 2: Section shows mononuclear and binucleate Reed-Sternberg cells amidst a polymorphous infiltrate (H&E x 400)](image2)
is most commonly affected, followed by lumbar region, and rarely the cervical region. It is suggested by Epelbaum et al. that this occurs because there is more tolerance for bulky disease in the thoracic cavity. In the neck, the lymphoma would present as a mass before causing spinal cord compression. In the present study, eight of the cases (67%) had thoracic spine involvement, with two cases each having lumbar and cervical spine involvement. In addition to the effects of local compression on the spinal cord, neurologic symptoms of ‘transverse myelitis’ may occur. This is due to the interruption of blood supply in the region of the intervertebral foramens. Very rarely, the infiltrate breaks through the dura to involve the pia and spinal cord itself.

Surgery is essential to obtain sufficient material for a pathologic diagnosis. Hodgkin’s lymphoma involving the epidural space is very responsive to radiation therapy and chemotherapy, with a good prognosis for both functional recovery (86%), complete response (61%) and long-term survival. Further operative procedures may be left in reserve for those patients (14%), who do not exhibit a good clinical response.

To make a diagnosis of Hodgkin’s lymphoma, one needs to find the characteristic milieu of cells- lymphocytes, plasma cells, histiocytes and eosinophils; as well as mononuclear and binucleate Reed-Sternberg cells. When there is previous history of Hodgkin’s lymphoma diagnosed on nodal biopsy, histologic interpretation becomes easy. In the absence of a previous biopsy, a diligent search has to be made for Reed-Sternberg cells. When there is previous history of previous biopsy, status of lymph nodes, liver, spleen is required. The diagnosis is easy, when previous history of Hodgkin’s lymphoma is available. However, when cord compression is the first manifestation, the diagnosis is difficult and the tissue has to be carefully examined.

Non-Hodgkin’s lymphoma would have a more monotonous population of lymphoid cells. These cells would be leucocyte common antigen (LCA) positive, as well as positivity for T-cell or B-cell markers. To differentiate Hodgkin’s lymphoma from non-specific inflammation, careful search for either mononuclear or binucleate Reed-Sternberg cells is essential. In tuberculosis causing cord compression, either ill-or well-defined granulomas, areas of necrosis or positivity for acid fast bacilli may be noted. However it must be noted that granulomas may be seen in Hodgkin’s lymphoma as well. Occasionally, a Hodgkin’s bone lesion may have to be differentiated from eosinophilic granuloma of bone. In eosinophilic granuloma, the histiocytes in some places, tend to show a meshy arrangement. Any areas of necrosis present are not ringed by connective tissue, as they are likely to be in Hodgkin’s lymphoma. Apart from these differences, Reed-Sternberg cells would not be seen.

Bone marrow examination done as part of staging procedure, showed involvement in 4/7 cases (57%). Reports indicate that usually, marrow involvement in Hodgkin’s lymphoma occurs in 5-15% cases. This unusually high incidence of marrow involvement indicates widespread disease at the time of presentation.

CONCLUSIONS

In the present study, the thoracic spine is the most commonly involved site. Histologic diagnosis of Hodgkin’s lymphoma presenting as cord compression, requires awareness of the condition. Detailed clinical evaluation, with history of previous biopsy, status of lymph nodes, liver, spleen is required. The diagnosis is easy, when previous history of Hodgkin’s lymphoma is available. However, when cord compression is the first manifestation, the diagnosis is difficult and the tissue has to be carefully examined.

REFERENCES