CASE REPORT

Primary Extradural Non-Hodgkin's Lymphoma

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Abstract

A 28 years old male presented to our institute with 3 months history of paraparesis with decreased sensation over left foot and loss of bladder and bowel control .The diagnostic work up revealed an extradural mass at spinal level L3 L4.Had laminectomy and the tumour was sub totally resected. Histological examination revealed non hodgkin's lymphoma. The patient was worked up for disease anywhere else and was confirmed to have primary extra-dural non-hodgkin's lympoma(PENHL). Patient was treated with corticosteroides, adjuvant radiotherapy and chemotherapy. The residual tumour completely disappeared and patient is living normal healthy life and is walking without support after 9 years of follow-up.

Key Words

Primary spinal, Non Hodgkin's Lymphoma, Radiotherapy, Chemotherapy

Introduction

No other malignancy is so hetrogeneous and complex in its biological behaviour and hightly variable in prognosis as non-hodgkin's lymphoma (NHL). Every year about 367000 patients are diagnosed to have NHL and extra nodal NHL constitutes about 10-20% of all non-hodgkin's lymphoma cases (1). Spinal cord compression is a rare presentation of non-hodgkin's lymphoma occurring only in 0.1% to 3.3% of patients of primary extradural NHL However, spinal cord compression may occur in 0.1% to 10.2% of NHL patients during the course of disease and is usually aggressive in behaviour (2). Various series have reported favorable prognosis for primary spinal extra dural N.H.L. (3-4). The clinical feature and outcome of treatment in these patients may vary depending upon the duration of symptoms and time of surgical intervention.

Case Report

A 28 years old young male first presented with pain left lower back and and weakness of both lower limbs of 3 months duration progressive with loss of bowel and bladder control since one month.On examination patient was in good general condition, no peripheral lymphadenopathy as well as normal systemic examination. There was a laminectomy scar in lumber region with a bed sore of 8×8 cm in the sacral region. Central nervous systemic examination revealed decreased power in both lower limbs grade 3/5 with decreased sensation over left foot. Patient was with indwelling Foley's catheter.

Investigation revealed Hb.: 13.9, TLC: 6100, DLC:55/39/5/, Plat: Adequate, B.U.:21, S. Crt. : 1.0, S.U.A.:6, RBS:97, S. Bil. -0.6, S.Alk. Po4-6.0, SGOT/SGPT: 21/16, CXR-PA - Normal Study, USG Abd. - Normal. Whereas MRI-(Spine) showed soft tissue mass causing displacement and compression of spinal canal with cortical erosion of L3. Bone marrow examination revealed no Infiltration with NHL, HPR (S-8877/94)-NHL Large cell type.

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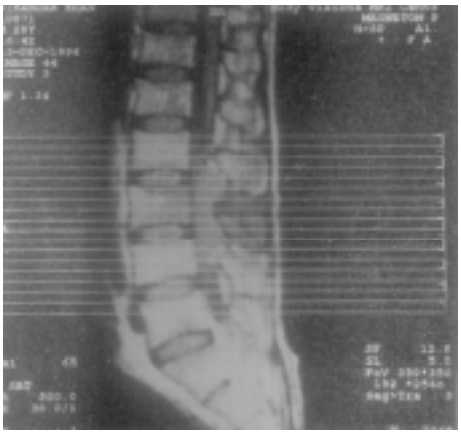

Fig. I Scout MRI film showing extradural mass lesion with erosion of pedicle of L3 vertebra.

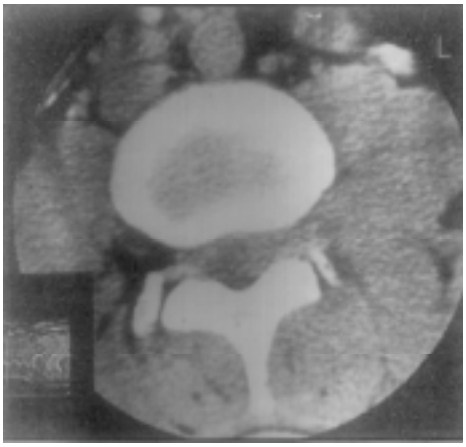

Fig - II Axial cut showing the para spinal mass.

Fig.-III Post treatment axial CT cut showing normal scan with resolution of primary tumour.

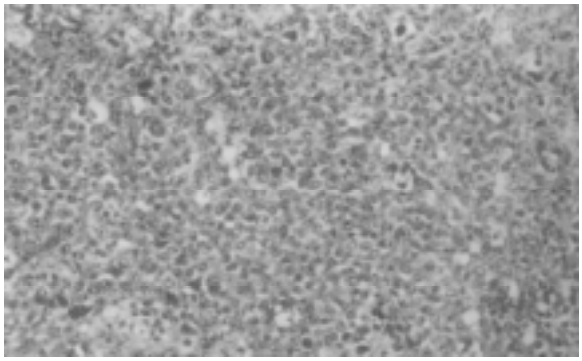

Fig.-IV Histopathology slide showing primary extradural NHL.

Treatment Given

Patient was treated with Ext. Radiation to dorsolumbar spine by two posterior oblique fields by isocentric technique by field size of 14x6 cm centring L 3 with a dose of 35Gy\15#\3weeks and was started with CCT with CHOP regime :

Inj. Adriamycin	...	50 mg.
Inj. Endoxan	...	800 mg (D1 & D8) x 4 wkly xD1
Inj. VCR	...	1.5 mg
Tab.PDN	...	60 mg.OD - (D1-D14)

Received 4 cycles when readmitted on 17-4-95 with history of fever and swelling left testis. Investigation revealed (Table - I)

Table I : Cyclic CHOP regime

Dates:	18-4-95	22-4-95	25-4-95	2-5-95	9-5-95
Hb.	11.2	9.9	12.6	10	10.5
TLC	500	500	8800	10900	4500
DLC	--	30/60	76/17/3/4	81/15/2/2	65/23/8/4
Plat	1.49×10^5	45000	32000	2.14×10^5	2.98×10^5
BU	30	33	75	22	20
S.Crt	1.0	1.3	2.7	1.2	1
S.UA	6.6	8.1	--	6	--
S.Bil.	--	0.6	0.8	0.5	--

CXR - Normal Study Urine C/S - Sterile
Throat Swab - Staph aureus & Kl. Pneumoniae
USG (Testis) - S/O Orchitis USG Abd. Normal Study
FNAC (Testis) A-1874/95 Acute inflam. Cells S/O Orchitis

Patient was managed with I/V antibiotics and supportive therapy and improved, received further chemotherapy cycles and completed treatment in June 1995.

Patient is on regular follow-up without catheter, walks of his own with slight left leg limp. His last CECT Abd. And Pelvis and x-ray chest dated Sept. 2004 are normal. There is no clinical evidence of any disease.

Discussion

Many authors have questioned the origin of primary epidural non-hodgkin's lymphoma (PENHL). Rubinstein (5) hypothesis believes them to be arising from normal lymphoid tissue present in epidural space. They propose the theory of antigenic stimulation with transformation cascade under appropriate conditions can produced primary spinal lymphomas from mesodermal cells lining the epidural space. Some authors support that these tumours originate from the para vertebral lymph nodes or from the vertebral body and later extend and cause extradural cord compression (6-7). Immuno-cytochemical and electron microscopic studies have shown that these tumours most frequently originates from the B-cells (8).

PENHL are seen more commonly in males (66-70%) with a male: female ratio of 1.6: 1. It usually presents in fourth to fifth decade of life. The most common presenting symptoms are backache and radicular pain of few months with a sudden neurological deterioration. Most common site of involvement is thoracic spine (69%) followed by lumber (27%) and cervical region has only (4%) of cases (9). The reasons for higher percentage of involvement of thoracic spine, is most likely the greater length of the thoracic portion as well as more concentration of lymphatic drainage in this region also the outcome is less favorable in patients where there is involvement of D5 and D8 vertebra (10). This is possible due to fewer radicualr arteries to the spinal cord and thus this region is more susceptible to ischemic injury secondary to epidural compression (11). The triad of most common presentation in PENHL is weakness of lower limbs (70-92%), localized back pain (50-60%) and bladder dysfunction (28-35%) (12). All patients need a thorough work up like nodal lymphomas to arise at exact staging and ruling out any other site of primary nodal disease. Plain radiographic abnormalities viz. - Pedicle arosion, Destruction of Vertical Body and Compression fracture and para-spinal mass may be seen in 15-42% of cases (13). The role of CT-Myleography is decreasing because of MRI and CT scan together provide greater details and more information about soft tissue involvement and extent of disease (14). Pathologically most patients have high/intermediate grade histology (4,15) some series have reported low grade histology also.

Patients who are seen with spinal cord compression and no known primary lesion elsewhere, require surgical intervention for diagnosis and relief of neurological deficit (14-17). As there is mostly absence of bony erosion by these tumorus consequently, laminectomy did not jeopardize spinal stability. Aabo and Walkbom have reported no difference in outcome between patients undergoing decompressive laminectomy and radiotherapy vs. those receiving spinal radiation only. The role of decomperessive laminectomy in PENHL presenting with peraplagia of recent origin has shown significant neurological improvement (15).

Role of Radiotherapy and Chemotherapy

Irradiation in PENHL is an important and effective treatment (15). Survival rates appear to correlate with the total dose of radiation delivered. Also the fractionation schedules are important as the late complications of radiation are dependent on dose per fraction and lymphomas are highly radiosensitive and potentially curable tumorus. Non-hodgkin's lymphoma requires a dose of 3500 cGy. to 4000 cGy. Delivered in 20-25 fractions over a period of 3-4 weeks to achieve radical cure (18). Successful chemotherapeutic treatments of epidural spinal cord compression in metastatic non-hodgkin's lymphomas have been reported (19) showing favorable response. PENHL is a chemo-sensitive tumour like other non-hodgkin's lymphomas. Hence chemotherapy is also used with a premise to prevent the distant failures. Various chemotherapy drugs have been used with variable response rates but CHOP regime (Cyclophophamide, Adriamycin, Vincristine and

Prednisolone) remains the gold standard of treatment.Primary spinal extradural NHL has poor prognosis for age > 50 years and patients with more aggressive histological types including diffuse large cell variety. Patients having longer duration of paraplegia and bladder and bowel involvement have poor outcome. Many studies highlight that type of surgery whether complete or sub-total lemectomy does not effect the survival pattern. As chemo-radiotherapy remains the main stay of treatment of PENHL.Primary spinal extradural NHL is an uncommon lesion usually presence with a syndrome of back pain, followed by an acute neurological deterioration in a person with no prior history of any malignancy. Surgery followed by spinal irradiation can result in significant neurological improvement. Addition of chemotherapy leads to favorable long term survivals especially in younger patients.

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