Figure 2b:

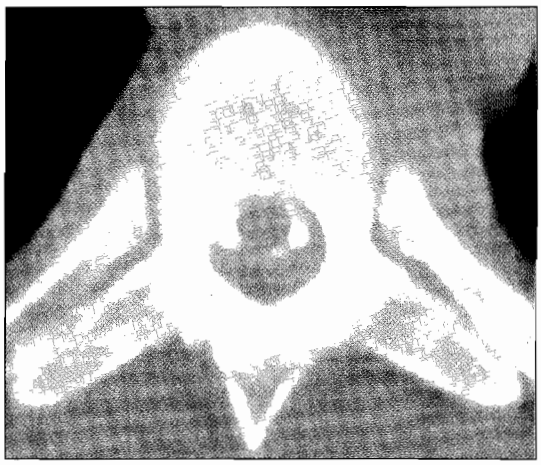

Figure 2c:

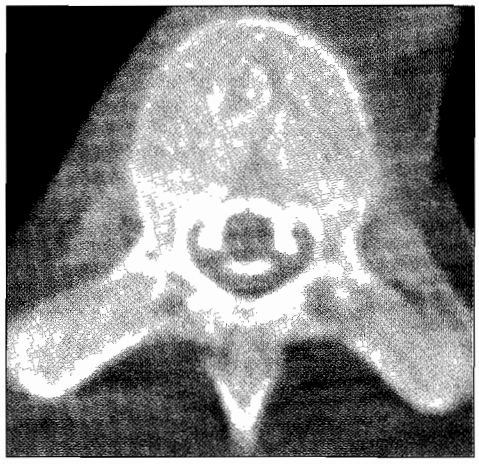

Figure 2d:

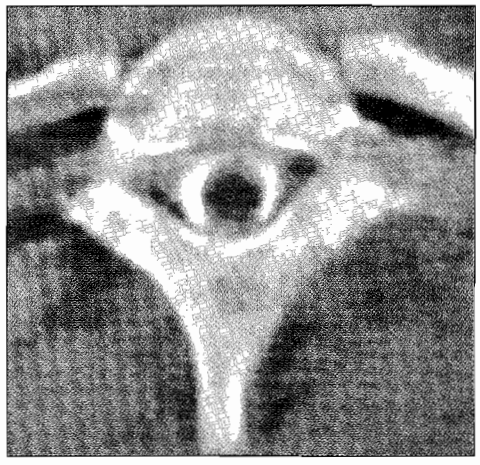

Figure 2b, 2c, 2d:
CT scans at various levels. Fig 2b shows a posterior mass displacing the cord anteriorly. Fig 2c and 2d are scans at levels above and below the mass respectively showing that the spinal cord is centrally located in the spinal canal

An orthopaedic consultation was first made on 7th March 2002. On reviewing the CT Myelogram the opinion was that the diagnosis was epidural lymphoma. He was now noted to also have weakness of the upper limbs and was developing weakness of chest wall muscles. After discussing with the parents it was felt that surgery would help to arrive at a tissue diagnosis but was unlikely to reverse the neurological deficit. The general poor prognosis was explained. They felt we should go ahead with surgery because if lymphoma were confirmed, they would consider buying the chemotherapy.

He received four units of blood prior to surgery and he was taken to theatre on 19th March 2002. Laminectomies were done bilaterally from T3 to T5. A soft, fibrinous, epidural mass was partially removed ant the spinal cord decompressed. Total excision was not possible due to the extent of the lesion. Histology of the tissue confirmed a diagnosis of non-Hodgkin's lymphoma. Worsening pressure sores and breakdown of the surgical wound complicated the postoperative period. The neurological deficit in the lower limbs did not improve, however power in the upper limbs returned to normal. Post operatively he required transfusion of four units of blood. Chemotherapy using a CHOP regime was recommended but the parents could not afford the medications. His general condition deteriorated and he died on 26th April 2002 from a combination of anaemia and septicaemia from infected pressure sores.

Discussion

Clinical Presentation / Features

Primary spinal epidural NHL can present at any age although it is more common in elderly patients with a median age 70 years. A prodrome period of back pain with a median duration of 3 months is common. This is followed by an acute neurological deterioration with a median duration of 6 days[2].

Neurological findings may include paraparesis or paraplegia; a discrete sensory level; hyperreflexia; clonus and loss of sphincter control. In most patients, diagnosis of primary spinal epidural NHL is made when neurologic deficit is already established and histological specimens have been obtained at surgery.

Investigations

Blood tests:

Routine blood investigations include FBC, ESR and peripheral blood film. These may reveal a normocytic, normochronic anaemia; pancytopaenia and atypical cells; elevated ESR. Bone marrow assessment is important to exclude systemic lymphoma. Blood culture is necessary to exclude septicaemia in patient presenting with features of meningitis and fever. VDRL and HIV tests should be done after appropriate counselling.

Lumbar puncture

Analysis of CSF is important. Young et al [4] report that CSF abnormalities are found in 97% of patients of NHL with CNS involvement. Routine microscopy, culture and sensitivity testing of CSF must be done. This may reveal raised protein and cell count with a predominance of lymphocytes. Cytology of the CSF may be done but has been shown to be positive in only 67% of cases of lymphoma with CNS involvement4. India ink staining should be done to exclude cryptococcal meningitis.

Urine and stool

Urine and stool analysis are important to investigate schistosomiasis as a cause of paraparesis / paraplegia.

Radiological

Plain x – rays of the spine are invariably normal with no evidence of bone destruction. Myelography and CAT scan will reveal an extradural mass compressing the spinal cord, usually extending over more than one vertebral segment. MRI scan should be done where available to further delineate the extent of

the lesion. Abdominal ultrasound, chest x-ray, CAT scan of the abdomen and chest are necessary to exclude systemic disease and for staging the disease.

Histology

Histological specimens from primary epidural NHL are obtained at the time of surgical decompression of the spinal cord. CAT guided biopsy of lesion may be attempted prior to surgery if the lesion is accessible.

In patients with NHL and CNS involvement, between 80% and 98% have a diffuse histological subtype CNS involvement in nodular lymphoma is uncommon and is reported in only about 3% of patients with NHL[4].

For proper sub typing immunohistochemistry and immunocytochemistry are important tests.

Treatment

Most patients with primary epidural NHL present with rapidly progressive or advanced neurological deficit. Diagnosis is usually confirmed by histological specimens taken at time of emergency surgical decompression of the spinal cord. Surgery is therefore important for decompression and tissue diagnosis. Surgery involves laminectomy and resection of the tumour. Spinal irradiation and systemic chemotherapy are important adjuvant treatment, which have been shown to increase disease free survival. Intrathecal chemotherapy can be considered in relapse cases. The ten-year experience at the Mayo clinic report shows that 8 patients had primary spinal epidural NHL and all underwent decompressive laminectomies, subtotal tumour resection and local spinal radiotherapy. Four patients died and four remain alive and well and free from disease. The twenty-year experience at the Memorial Sloan – Kettering Cancer Centre showed that 5 patients had primary spinal epidural NHL. Two patients died and three remained alive and diseases free after a median follow up period of ten years. Response in AIDS / HIV patients is similar to that seen in non-AIDS patients [5].

Prognosis

Some poor prognostic factors especially with respect to resolution of paraplegia and disease relapse include:

- Rapidly progressive paraplegia with lack of motor function at presentation (Fraenkel A, B) and long duration of complete paraplegia before presentation have a poor chance of neurological recovery.
- Poorly differentiated, diffuse histological grade is associated with increased relapse rate.

These two cases raise several discussion points. Primary spinal epidural NHL can present at any age, although the median age in most series is 70 years. MC is a young man who is HIV negative whereas LT is middle aged man and is HIV positive. There is a belief that with the HIV / AIDS pan epidemic the number of cases of primary spinal epidural NHL will increase. The literature suggests that the response to chemotherapy and radiation in AIDS patients is similar to non-AIDS patients.

Both patients reveal the difficulty in diagnosing this condition. During the prodrome period of back pain both patients did seek medical attention. However by the time they were referred to the central hospital, neurologic deficit had been present for some time. A high index of clinical suspicion is needed to consider this rare condition as a differential diagnosis in patients presenting with paraplegia. The other major constraint in arriving at a diagnosis is the lack of radiological investigations especially Myelography. In both these patients CT Myelography was done after some delay because the patients had to pay for it themselves at a private institution because QECH did not have Myelography contrast media and the CT scan is out of commission.

MC had a more rapid progression and on presentation had no motor function and already had developed some pressure sores. Although surgery was done, it was known that this was mainly to aid in tissue diagnosis and would not reverse the neurological deficit. His neurological deficit has remained. LT had a slower progression and had residual motor function at the time of presentation. Surgical decompression has resulted in improvement of the neurological deficit.

Chemotherapy in both cases has to be purchased by the patients, as these drugs are not on the hospitals pharmacy list. They are expensive and both patients were not able to purchase the medications. This undoubtedly will affect outcome in patient LT. It is debatable whether chemotherapy would have prevented the death of patient MC. Radiotherapy has been shown to be a useful adjuvant therapy but unfortunately is not available in Malawi. Primary spinal epidural NHL should be considered in patients who give a history of back pain; followed by rapid development of features of spinal cord compression; have normal plain x-rays but whose CAT / myelogram reveal an extradural mass. Urgent surgical decompression followed by chemotherapy and radiotherapy improves the outcome of such patients.

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