Proceeding S.Z.P.G.M.I vol: 9(1-2) 1995, pp. 44-46.

Combined Intradural and Extradural Tumour: A Case of Lymphoma of The Filum Terminale

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SUMMARY

Lymphomas are tumours occurring both in the intradural and the extradural compartment. We report a case of lymphoma of the Filum Terminale with the existence of both an intra and an extradural component in continuity. This has not been described before. The possible reasons for such a configuration are also discussed.

INTRODUCTION

The dura usually presents a thick impenetrable barrier to the spread of spread of malignant tumours from the extradural to the intradural region. We report a recent case with such a contiguous spread.

CASE REPORT

A thirteen year old female presented to our neurosurgical clinic with dull persistent backache for the previous six months. Neurological examination showed a minimal weakness of right big toe dorsiflexion with a loss of ankle jerks bilaterally. A myelogram showed a complete block due to intradural tumour opposite the middle of the body of the L4 vertebra (Fig. 1). A CT Scan performed subsequently confirmed the thecal sac filled with tumour of homogeneous density with asymmetric expansion of the sacral spinal canal but no bony involvement (Fig. 2).

She was started on dexamethasone 4 mg q.i.d. However a few hours prior to scheduled surgery she developed urinary retention preceded by severe backache and left leg pain. There was sensory loss only on the left from S2 to S4. A L4-L5 laminectomy was then done with sacral deroofing to expose a combined intra and extradural tumour with severe distortion of the thecal sac. The tumour arose from the filum terminale intradurally and continued extradurally having a constriction at the point of exit from the dural sac. It was soft and haemorrhagic having necrotic areas. The extradural component was excised completely while the intradural component was excised subtotally (Fig. 3) there

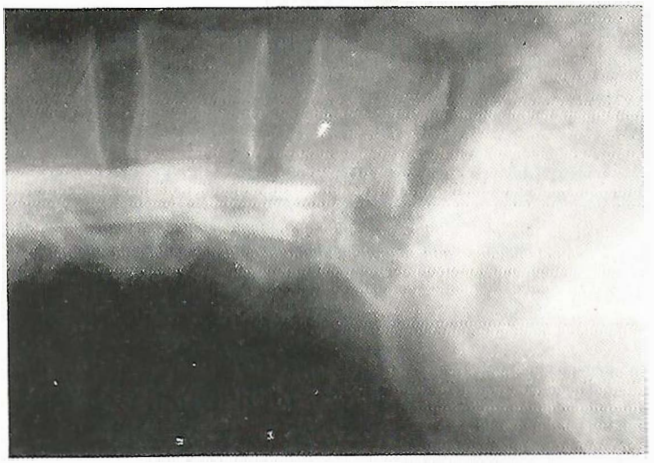

Fig. 1: Myelogram showing block at L4.

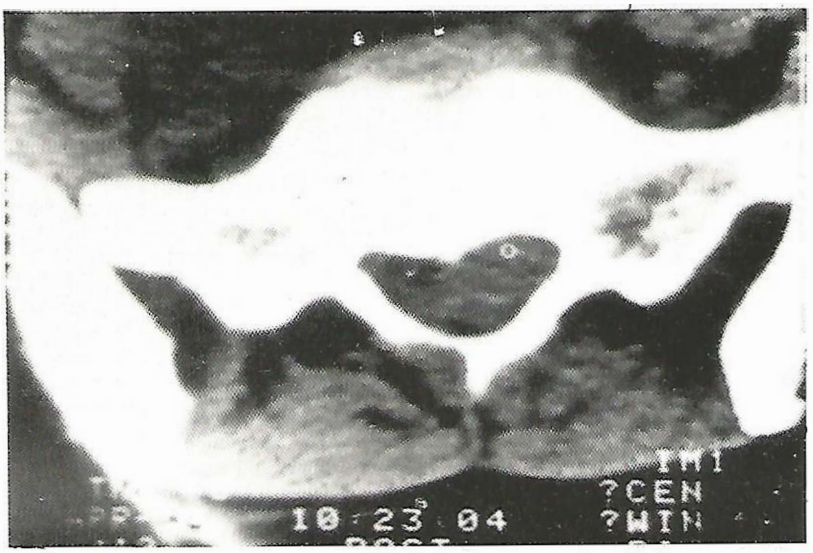

Fig. 2: CT scan at S1 showing expansion of spinal canal.

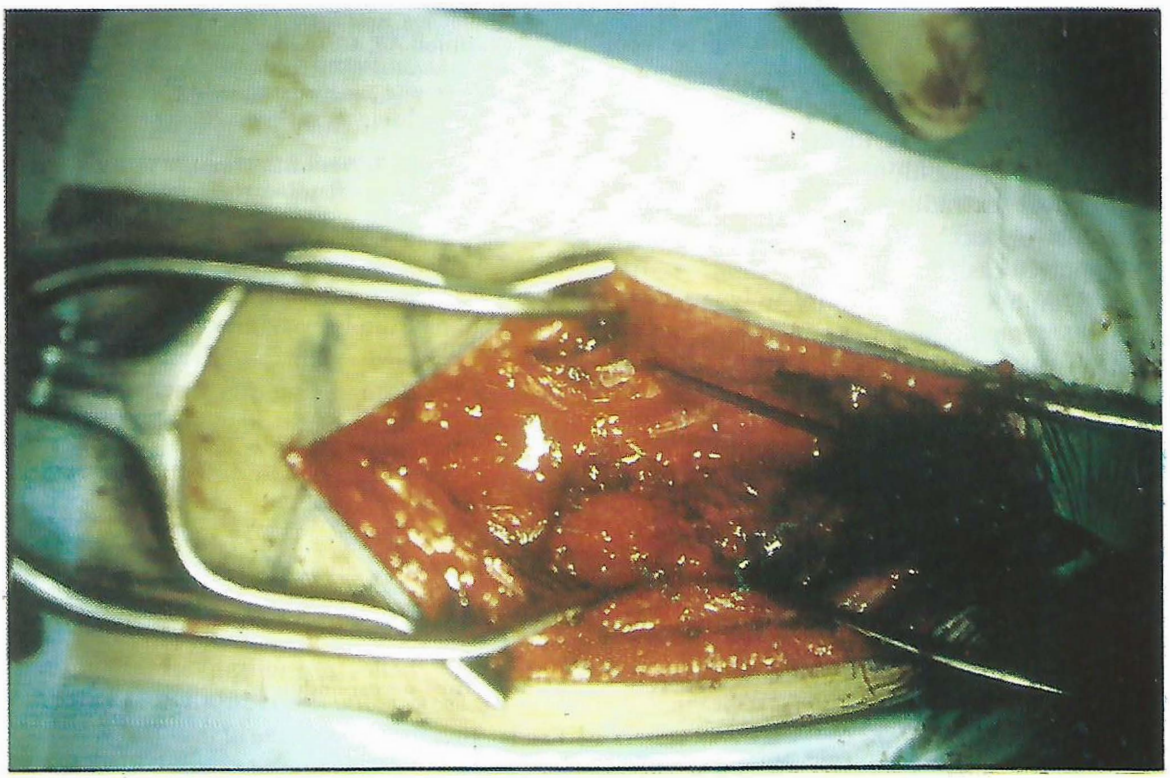

Fig. 3: Operative photograph showing intradural and extradural tumour.

being small fragments left adherent to the intradural nerve roots. Histopathologic examination revealed a tumour with small cells, pseudorosette formation and areas of haemorrhagic necrosis, probably a lymphoma.

Postoperative recovery was rapid and she recovered her urinary control. However her parents opted initially for her not to have radiotherapy and combination chemotherapy these being started only after she again developed urinary retention four

months after surgery. She received a modified CHOP (cyclophosphamide, adriamycin, vincristine and prednisolone) chemotherapy regime and remained well until dying of systemic spread and local recurrence about eighteen months after diagnosis.

DISCUSSION

Spread of tumours from the intradural to the extradural compartment and vice versa is resisted by the tough fibrous dura which provides a barrier to the infiltration of tumours. However transgression of the spinal dura by adjacent primary tumour such as Ewing's sarcoma may be seen rarely[1]. Transdural spread of spinal epidural lymphomas is even more rare with Russell and Rubinstein having seen only one example in a fifteen year old boy[2]. Benign tumours such as neurofibroma arising from dorsal nerve roots may infiltrate along the nerve root sheath and achieve an hourglass configuration with one part within the thecal sac and one outside it[3]. Such a mechanism is likely in our patient where tumour arising from the filum terminale infiltrated along it to appear extradurally. To our knowledge after an extensive search of the literature, it is the only such reported case.

Spinal lymphomas commonly present with symptoms of cauda equina or spinal cord compression[4]. Diagnosis is usually established by positive contrast myelography to localise the lesion which may not cause any bony changes. MR imaging is a recent welcome addition to the diagnostic armamentarium[5]. Treatment is by laminectomy with subtotal excision and nervous tissue decompression followed by radiation locally and combined modality chemotherapy[6]. This was the option chosen for our patient. Worthwhile improvement and survival has been reported[7].

An interesting fact is the response of these lesions to steroid therapy with control in some tumours being achieved by steroid therapy alone[8]. The acute deterioration on the initiation of steroid therapy in our patient along with the operative and histological findings of acute necrosis is evidence of response of lymphomas to these drugs. Our report serves to highlight a possible mechanism of spread of intradural lymphoma to the extradural compartment. We await with interest further reports of such tumours.

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