EXTRADURAL TUMOURS IN CHILDREN PRESENTING AS PARAPLEGIA

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Paraplegia as a complication of malignant disease is rare in a paediatric practice. It is usually misdiagnosed as Guillian Barre Syndrome, poliomyelitis or traumatic paraplegia. Spinal cord compression due to epidural metastasis occurs in 1-4% of paediatric cancers[1]. This study summarizes the clinical characteristics and treatment responses of children with spinal cord compression.

PATIENTS, METHODS AND RESULTS

Of 214 children with malignant tumours admitted in the Children's Hospital between 1987 to 1991, six (2.8%) had spinal cord compression. The diagnosis and levels of spinal cord compression were substantiated by Omnipaque myelography in all patients. Location of the lesion was confirmed by laminectomy. Biopsy and histopathology were done to confirm the tumour types. Spine roentgenography as well as examination of CSF and bone marrow were performed in all patients. The tumours presenting with spinal cord compression are shown in Table I.

TABLE I. Tumours presenting as spinal cord compression.

Primary tumours	Total No. of patients	No. and % of patients with cord compression
Ewing's sarcoma	6	1(16%)
Rhabdomyosarcoma	16	1(6.2%)
Lymphoma	43	1(2.3%)
Neuroblastoma	7	1(14.2%)
Acute myeloid leukaemia (AML)	27	2(7.8%)

It was seen more frequently in solid tumours than in leukaemia. The clinical signs and symptoms are shown in Table II.

TABLE II. Signs and symptoms in spinal cord compression.

		No.	%
Symptoms	Weakness in limbs	6	100
	Sphincter dysfunction	6	100
	Pallor	4	66.6
	Back pain	4	66.6
Signs	Hypotonia	6	100
	Sensory loss	5	83.3
	Tenderness over spine	5	83.3
	Hyporeflexia	4	66.6
	Hyperreflexia	2	33.3
	Abdominal mass	1	23.3

The initial symptom was back pain with gradual progression to profound weakness of the extremities in all patients. This was followed by sensory impairment in 83.3% and loss of sphincter control in 100% of cases. All patients had paraplegia at diagnosis. One patient was diagnosed as Guillian Barre Syndrome, one as poliomyelitis and one as traumatic paraplegia. Spine x-rays demonstrated bony abnormalities in 4 patients including osteolytic destructive lesions and wide disc spaces. CSF showed increased proteins in all patients. Histopathological examination of biopsy specimens confirmed solid tumours in 4 patients and leukaemia in two, which was also confirmed on bone marrow biopsy. The level of spinal cord compression in different tumours is seen in Table III.

TABLE III. Level of lesion and response in different malignancies.

Type	Age and sex	Mean duration of symptoms	Level of lesion	Status before treatment				Treatment		Clinical improvement	Tumour remission	Current status
				Motor deficit	Sensory loss	Bladder and bowel dysfunction	Reflexes	Laminectomy	Chemotherapy radiotherapy			
AML	11 yr/F	1 yr.	T5	+	+	+	Absent	+	Cytosar, cosmogen	No	No	Death in 5 days due to intracranial bleeding
AML	11 yr/M	2 months	T11-T12	+	-	+	Decreased	-	Thioguanine cytosar, VP-16	Yes	PR	Survived 1 yr.
Ewing's Sarcoma	10 yr/M	15 days	L4	+	-	-	Brisk	+	VCR, bleomycin dactinomycin, cyclophosphamide	Yes	CR	Survived 1 yr.
Rhabdo myosarcoma	13 yr/F	2 months	T5-T6	+	+	+	Decreased	+	VCR, actinomycin cyclophosphamide	No	PR	Survived 6 months
Metastatic neuroblastoma	7 yr/M	1 month	T9-T10	+	+	+	Decreased	+	VCR, Prednisone endoxan, MTX	Yes	CR	Survived 3 yrs.
Lymphoblastic lymphoma	3 yr/M	1 month	T5-T9	+	+	+	Decreased	+	VCR, prednisone MTX L-asparginase	No	PR	Survived 5 months

The results of treatment are also summarized in Table III. The duration of survival after spinal cord compression differed according to tumour types. All patients hadsymptoms for more than 1 week. One patient with the shortest duration of symptoms (15 days) had complete recovery of all functions and one showed improvement in muscle tone, power and bladder function. The response to treatment was negligible in patients with long durations of symptoms. Four patients survived, 2 are in complete remission one year after diagnosis. Two patients still have evidence of disease.

COMMENTS

The spinal cord and cauda equina may be compressed by a tumour in the epidural or subarachnoid space or by metastatic spread to the cord parenchyma by infiltrating through the inter vertebral foramina[2]. It is more frequent in solid tumours than in leukaemia[1]. Sarcomas account for 43-65% of metastatic spinal cord disease in children. Ewings tumour, neuroblastoma, lymphoma and leukaemia account for the rest[3]. Other causes are rare enterogenous intraspinal cysts and treatment related myelitis[4]. Congenital intraspinal neuroblastoma has been reported to cause compression in 11 patients[2]. Back pain which is seen in 80% of cases may be local or radicular[3]. Reliable localizing sign is tenderness to percussion in 80-90% of patient. There is loss of motor strength in the extremities. The progression of the clinical signs and symptoms is related to the growth rate and location of the tumour and therefore varies from one patient to another[6]. They may develop suddenly or progressively. Radiological abnormalities are seen in less than half of the children[7]. Diagnosis is confirmed by myelography. MRI has recently proved to be a useful diagnostic test[7]. Spinal cord compression is a medical emergency. Any plan of therapy for newly diagnosed patients should take into account the restoration or preservation of neurologic functions, the reduction of skeletal deformities and the potential for radiation induced second malignancies[1]. If severe or progressive, dexamethasone 1-2 mg/kg i/v is given to reduce cord edema followed by immediate myelography[8]. If an epidural mass is demonstrated then the spinal cord must be decompressed immediately. Both local radiotherapy and surgical decompression can be used[9]. Radiotherapy is given if the tumour is radiosensitive. A total of 2000 to 3000 CGY is given depending on the tumour histology[6]. Chemotherapy is given to patients with newly diagnosed leukaemia or lymphoma[10] and started as soon as possible after diagnosis. In these patients laminectomy may not be necessary except for biopsy[10]. These patients have to be observed closely as an occasional child may require neuro-surgical intervention. Hayes et al have demonstrated the effect of chemotherapy alone in patients with epidural metastasis from neuroblastoma and Ewings sarcoma[10].The prognosis for recovery in patients with spinal cord compression depends on the neurologic findings at the time of diagnosis[1]. Full recovery is possible if appropriate treatment is initiated early[11].

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