

gradients were reduced from 110 to 50 mm Hg in the patient reported here (*Fig. 1*).

The major hurdles to extensive application of balloon dilatation technology in infants in India include the limited availability of hardware and expertise for interventions in this age group. Further, a number of infants in our country with lesions amenable to early intervention may escape timely attention. We believe that this technique is particularly promising in the Indian context because of its relative simplicity and low cost as compared to operative treatment.

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## Enterogenous Intramedullary Cyst

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Spinal enterogenous cysts are rare cystic lesions of the spinal cord, first described by Harriman(1). Many isolated case reports

(2-4) are available in the literature. Majority of them are intradural extramedullary. The intra-medullary cyst has a different clinical presentation and to the best of our knowledge only 6 cases have been reported till now(3,5-8). Based on one of our case and

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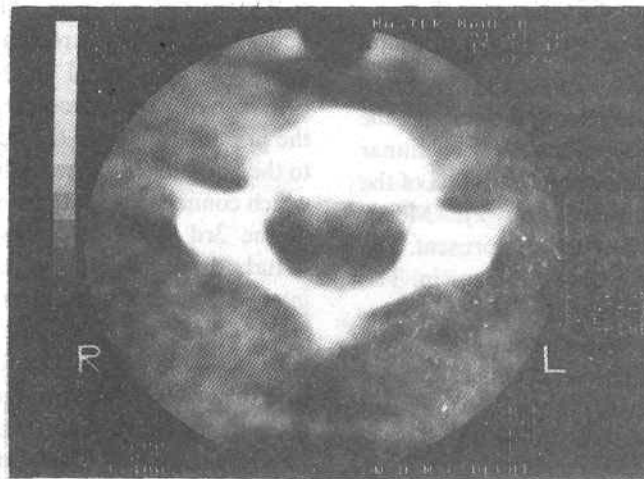
review of literature we have tried to indicate the clinical features of intramedullary enterogenous cyst. Immunohistochemistry, done for the first time in enterogenous cyst is also reported.

### Case Report

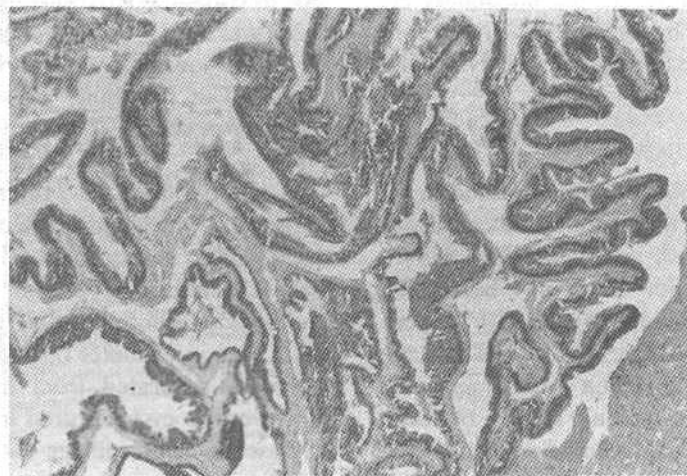
A six-year-old boy was admitted in the pediatric ward with pain in the neck and weakness of both upper and lower limbs following a minor fall a day before. At admission he was able to walk with support. The weakness progressed rapidly and the child became bed-ridden within a period of ten days. There was no history of fever, chronic cough, headache, vomiting, convulsions or weakness in any limb prior to this episode. Neurological examination revealed signs of lower motor neuron lesion in both lower limbs. Motor power was Grade 0 in both lower limbs and there was no appreciable sensory deficit. Clinically, there was no obvious spinal deformity or lymphadenopathy. Plain X-ray spine did not reveal any abnormality. Contrast CT showed complete block dye column at the level of C-7. A low

attenuating lesion at C-6 producing expansion of cord and narrowing of sub-archnoid space was seen (*Fig. 1*). Delayed slices after 24 hours showed no contrast in the cervical region. The possibility of intramedullary lipoma was considered on the basis of CT findings.

Laminectomy at C5-7 revealed and bulge in the dura which was opened and the cord was found to be expanded. The spinal cord was slit open through posterior median sulcus and bluish, lucent glistening mass measuring 1.5 cm in diameter bulged out. It could be easily separated from neural tissues. However the cyst ruptured in the last part of dissection with spillage of clear fluid and a sub-total removal of the cyst wall was done. Postoperatively, patient lost sensation, bowel and bladder control. He also developed bed sores, hyperthermia and CSF leak. Bladder and bowel control were regained in 2 months time and bed sores also healed during this period. He however, continues to have off and on hyperthermia. The septic cause of fever was negative on more than one occasion. The fever used to settle by



*Fig. 1. CT Scan at C<sub>6</sub> showing a low attenuating lesion and expansion of cord.*



*Fig. 2. Photomicrograph of the section from the cyst lined by columnar epithelium, at places by pseudostratified ciliated columnar epithelium and foci of squamous metaplasia. H & E  $\times$  200.*

shifting the child to a cooler atmosphere and hence an autonomic derangement was attributed for hyperthermia in this child. The CSF leak settled at 4th post operative week by elevating the head and use of acetazolamide. At present 12 months after operation, the patient has recovered nearly 50% of sensation. However, there is no return of useful motor power.

Histological examination showed that the lining epithelium varied from low columnar to high columnar and there were foci of the squamous metaplasia (*Fig. 2*). Mucus secreting goblet cells were also present. The lining cells stained positive for mucin. The lining epithelium rested on loose fibrovascular connective tissue without any muscle or cartilage. Immunohistochemical staining revealed positivity for both Desmin and Vimentin.

#### **Discussion**

The dermoid and epidermoid cysts are

the commonest intraspinal dysgenetic cysts with the rare variety being arachnoidal, enterogenous, teratomatous and empendymal cyst(8). The spinal enterogenous cysts \* have an epithelial lining resembling gastrointestinal mucosa. They have been referred to by various synonyms like intestinoma, neurenteric cyst, gastrocytoma and enteric cyst(4,9). The spinal enterogenous cysts originate from the incomplete separation of the primitive gut from the neural groove due to the persistence of the Lieberkiihn channel which connects the entoderm and ectoderm in the 3rd week of embryonal life. This disturbs the development of mesoderm which interposes between the two embryonal layers to form vertebral bodies, leading to vertebral anomalies and enterogenous cysts(1). Immunohistochemical findings suggest the mesodermal interposition in the present case. Bentley and Smith(10) asserted that enterogenous cysts are invariably associated with a vertebral defect but cases without any

vertebral defect have also been reported(4). No vertebral anomaly was encountered in our case. The lining epithelium showed foci of squamous metaplasia as well as ciliated cells as noted by previous authors(1,3,5).

The clinical symptoms associated with enterogenous cysts depend on the site of lesion and are typical for all such cysts. The intramedullary enterogenous cysts present a progressive illness with neurological deficit (Table I). Out of total seven cases, clinical course is not spelled out in two. Four cases including ours have shown progressive neurological deficit. Seventh case had episodic pain in interscapular region but once neural deficit appeared it was progressive. On the contrary in extra medullary enterogenous cysts majority of cases show episodic and waxing and waning neurological

deficit. The neurological recovery after surgical decompression was very little in three patients including the present case. Out of the other four cases, one died in the post operative period and in three cases follow up course is not reported by the authors.

The diagnosis of enterogenous spinal cyst is based on histological findings. The clinical differentiation from other space occupying lesions of the spinal canal is difficult preoperatively. Moreover, CT findings of a low attenuating intramedullary lesion is not characteristic of this lesion.

We feel in such a presentation the possibility of intramedullary enterogenous cyst should be entertained alongwith lipoma, syringomyelia, ependymal cyst and parasitic cysts like cysticercus cellulose and hydatid

TABLE I - Summary of Reported Cases of Intramedullary Enterogenous Cyst

S. No.	Author(s) & year	Age (yrs.) & Sex	Location of cyst	Clinical course	Neurological deficit	Outcome
1.	Longmaid & Jones (1963)(5)	11M	(i) Lumbosacral region (ii) T <sub>8</sub> -T <sub>9</sub>	Not mentioned "	Not mentioned "	Not known "
2.	Rewcastle & Francoeur (1964)(6)	34 F	D <sub>10</sub> Progressive	Paraparesis	Very little recovery	
3.	Silvernail & Brown (1972)(7)	12M	T <sub>2</sub> Episodic	Weakness in upper limbs	Partial recovery	
4.	Deshpande <i>et al.</i> (1972)(3)	11M	above C <sub>3</sub>	Progressive with respiratory & sphincter involvement	Quadriplegia operatively	Died post-
5.	Vivek Sharma <i>et al.</i> (1991)(8)	20M	C <sub>8</sub>	"	Quadriparcsis; upper limb LMN Lower limb UMN	Not mentioned
6.	Present case (1991)	6M	C <sub>6</sub>	"	"	Very little recovery

cyst, although we agree that it is a histological curiosity.

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