

# Spinal Neurenteric Cyst and Hydrosyringomyelia with Vertebral Anomalies in an infant

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**Abstract:** Spinal neurenteric cysts are rare intradural development lesions, usually composed of a thin walled cyst whose lining mimicks gastro-intestinal or respiratory epithelium: A3 months old baby presented with backward deviation of neck since birth along with scoliosis of spine. MRI revealed multiple vertebral anomalies along with hydrosyringomyelia and spinal neurenteric cyst. The case reported being submitted on the account of rarity of anomalies of these cysts with hydrosyringomyelia.

## Introduction

Spinal neurenteric cysts are rare intradural developmental lesions, usually composed of a thin walled cyst whose lining mimicks gastro-intestinal or respiratory epithelium. Older age and cervical location are found to be statistically characteristic of solitary cysts in contrast to younger age and lumbo-sacral location for dysraphic cases (1). Association of these cysts with hydrosyringomyelia has not been reported. We report a case of spinal neurenteric cyst and hydrosyringomyelia with multiple vertebral anomalies.

## Case Report

A 3 month old male baby born to non consanguineous parents was brought with history of fever for 15 days, cough and nasal discharge for 12 days. There was history of backward deviation of neck since birth. There was history of drug intake antenatally for a male baby. The exact nature of the drug was not known. Baby was 6th in birth order. Two elder siblings were females. There was history of death of two elder female siblings at the age of 1 and 2 yrs due to pneumonia. Developmental history was normal.

On examination weight was 3.3 kg, length was 53cms and head circumference was 39cms. Vitals were stable. Anterior fontanella was 1x1cms and was at level. There was a bulge on right side of chest. Scoliosis of spine towards right side was present. Examination of chest revealed bilateral vesicular breathing with crepitations on back on right side. Cardiac and central nervous system were

normal. Liver was palpable by 3.5cm below right costal margin and span was 5cms. Rest of abdominal examination was normal. On investigations haemoglobin was 10gm%, TLC 16000/mm<sup>3</sup> with polymorphs 51%, lymphocytes 37%, monocytes 3%, eosinophils 3% and band cells 6%. Blood culture showed no growth of pyogenic organisms. CSF examination was normal. X-ray chest showed kyphotic deformity towards right along with vertebral anomalies in upper thoracic region. MRI of spine showed scoliosis of cervico-dorsal spine along with multiple vertebral anomalies with fused block vertebrae, hemivertebrae and butterfly vertebrae. Spinal cord showed hydrosyringomyelia at two levels cervico-dorsal and lower-dorsal. Dural ectasia was also present (Fig.2). A posterior mediastinal mass was also seen. It was well define and hyperintense on T2 weighted images and hypointense on T1 weighted images with internal separations. The mass extended caudally till D10 (Fig.2). It was spinal neurenteric cyst in the thoracic spine. Patient was diagnosed as a case of pneumonia with spino-vertebral anomalies. Treatment in the form of I/V fluid, oxygen inhalation, inj. Ceftriaxone 100mg/kg per day in divided doses I/V and inj. Vit. K 2mg I/M OD was given. Patient responded to the treatment.

## Discussion

Neurenteric cysts are rare congenital lesions of spine and are lined with endodermal epithelium. They result from anomalous endodermal-neuroectodermal adhesion in the 3rd week of embryonic life with persistence of canal of Kovalevsky. The most common location is cervico-dorsal region and usually lie ventral to spinal cord<sup>2</sup>. A detailed review of literature revealed 80 cases of solitary spinal neurenteric cysts which were analysed and compared regarding clinical and pathological aspects with 56 such cases of those with concomitant evidence of dysraphism. Older age and cervical location are found to be statistically characteristic of solitary cysts in contrast to younger age and lumbo-sacral location for dysraphic cases, magnetic resonance imaging is the diagnostic modality of choice<sup>1</sup>. A definite diagnosis can only be made by biopsy and histological examination<sup>3</sup>. Solitary cyst are mainly composed by endodermal derivatives while dysraphic cases also have mesenchymal and ectodermal elements indicating an earlier area in development<sup>1</sup>. Therapy of choice is complete resection<sup>4</sup>.

Associated vertebral anomalies, scoliosis, hemivertebrae and anterior spina bifida<sup>5</sup> gut cysts, bowel duplication, the presence of keratin markers and mucin secreting cuboidal or columnar intestinal

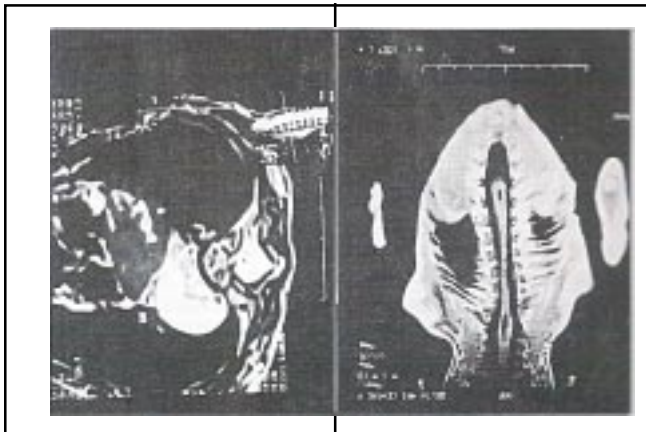


Fig.1: MRI showing hydrosyriago myelia at cervieu dorsal and lower dorsal region. Fig.2: MRI showing spinal neuro enteric cyst.

epithelium in their walls confirm their endodermal region<sup>2</sup>.

In our case neurenteric cyst was present in thoracic region. It was associated with multiple vertebral anomalies and hydrosyringomyelia.

Syringomyelia is a cystic cavity within the spinal cord that may communicate with the CSF pathways or remain localised and non-communicating<sup>6</sup>. It is associated with congenital malformations as well as trauma and tumours<sup>7</sup>. Incidence of scoliosis with syringomyelia has been found to vary from 4% to 20%<sup>8,9</sup>. In one series, it was 18.4% in boys and 2.6% in girls<sup>8</sup>. Scoliosis develops in children as a result of damage done to the anterior horn which innervates the muscles of trunk by an asymmetrically expanded syrinx<sup>10</sup>. Absent superficial abdominal reflexes in patients with scoliosis is an indication for investigations for underlying syringomyelia. Patients with thoracic curves are generally asymptomatic. Their neurological signs are subtle. Patients with thoracolumbar curves have neurological signs<sup>11</sup>. Spontaneous shrinkage of syringomyelia in children is not unusual and is associated with improvement in tonsillar herniation, scoliosis and the neurological deficit<sup>12</sup>.

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