

Mediastinal neurenteric cysts in children: A case series

Kamal Nain Rattan¹
Ananta Rattan³

Hemant Yadav² Divya Srivastava²

¹ Senior Professor, Department of Paediatric Surgery PGIMS, Rohtak

² Senior Resident, Department of Pathology PGIMS, Rohtak

³ Junior Resident, Department of Paediatrics RNT Medical College, Udaipur

[Address for correspondent](#) Dr. Hemant Yadav, Department of Pathology PGIMS, Rohtak.
(e-mail: hemantyadav7022@gmail.com)

How to cite this article:

Rattan K.N, Yadav H, Srivastava D, Rattan A. Mediastinal neurenteric cysts in children: A case series. *Iranian Journal of Pediatric Surgery* 2016; 2(1): 36-39.

Abstract

Introduction: Foregut duplication cysts along with vertebral anomalies are called neurenteric cyst. About 30 cases have been reported in the literature so far. Posterior mediastinal neurenteric cysts are very rare and few studies could be found in the literature even after extensive search. Due to this rarity we are prompted to present 7 cases of posterior mediastinal neurenteric cysts to further add on to the literature.

Material and Methods: All details regarding patients with neurenteric cysts who underwent surgery, in the department of paediatric surgery from 2010 to 2015 were gathered from patient case files.

Results: There were 7 patients with neurenteric cyst including 3 males and 4 females with an age range of 4 months to 8 years. The most common presentation was recurrent respiratory infection. In two cases, patients also suffered from dysphasia. In 5 cases there were right sided cysts while two had cyst on left side in posterior mediastinum. In 2 cases the cysts extended to the abdomen while one patient had separate ileal duplication cyst. Another case had mediastinal cyst which had a communication with ileum. All the mediastinal cysts were excised through posterior thoracotomy and subsequently confirmed on histopathology.

Keywords

- mediastinal
- neurenteric cyst
- paediatric

Conclusion: When considering the differential diagnosis of mediastinal cysts in children, neurenteric cysts should be in mind.

received: 20 October 2015

accepted: 17 May 2016

ISSN 2423-5067.

Introduction

Neurenteric cysts are among the most rare congenital anomalies with only 30 cases being reported in literature so far to the best of our knowledge.¹ Developmentally these are enterogenous type duplication cysts, which are associated with vertebral anomalies because of persistence of foregut connection with notocord.² These cysts mostly present in the first year of life but a few cases have also been diagnosed antenatally.³ The location varies from intracranial to abdominal.⁴

Posterior mediastinum as a location is very rare.⁵ Herein we report 7 cases of posterior mediastinal neurenteric cysts, all of which were operated on in the department of pediatric surgery.

Materials and methods

In this retrospective study we included patients with mediastinal neurenteric cysts all of which underwent surgery in the department of paediatric surgery at a

tertiary care hospital during 2010 to 2015. All the details were archived from the patient case files. A detailed study about age of presentation, sex, symptoms, preoperative investigations, diagnostic modalities, intraoperative findings and any associated anomalies was carried out. All patients were followed with chest x-ray and abdominal ultrasonographic examination.

Results

There were 7 patients with mediastinal neurenteric cyst. Their age ranged from 4 months to 8 years. Out of a total 7 patients there were 3 males and 4 females (Table 1).

Preoperative hematological parameters were within normal limits in all of the cases. Radiological investigations included ultrasonography, x-ray and computed tomography scan (Figure 1).

Table 1: Complete details of all cases of posterior mediastinal neurenteric cysts

Case no	Sex	Age	Chief complaints	Side and other anomaly	Surgical detail	Histopath detail
1	Male	7 months	Chest infection	Left, thoraco-abd	Excised	Type I with columnar lining
2	Male	4 months	Failure to thrive	Right	Excised	Type II with columnar lining
3	Female	8 years	Chest infection and dysphasia	Right, thoraco-abd	Excised	Type II with pseudo-stratified lining
4	Female	2 years	Chest infection	Right, scoliosis	Excised	Type I with columnar lining
5	Female	8 years	Chest infection and dysphasia	Right, scoliosis	Excised	Type II with columnar and gastric lining
6	Female	5 and half months	Chest infection and failure to thrive	Right with a separate ileal duplication cyst	First ileal cyst excised later on mediastinal	Type II with columnar lining
7	Male	7 months	Bleeding per rectum, failure to thrive	Left side Cyst communicating with ileum, scoliosis	First communication excised later on mediastinal	Type II with columnar lining

One of the patients was diagnosed as having neurenteric cyst on the prenatal ultrasonographic examination while others were diagnosed after birth. The main presenting symptom was recurrent chest infection however two cases also had additional history of dysphasia. One of our cases presented with rectal bleeding, failure to thrive and recurrent chest infection.

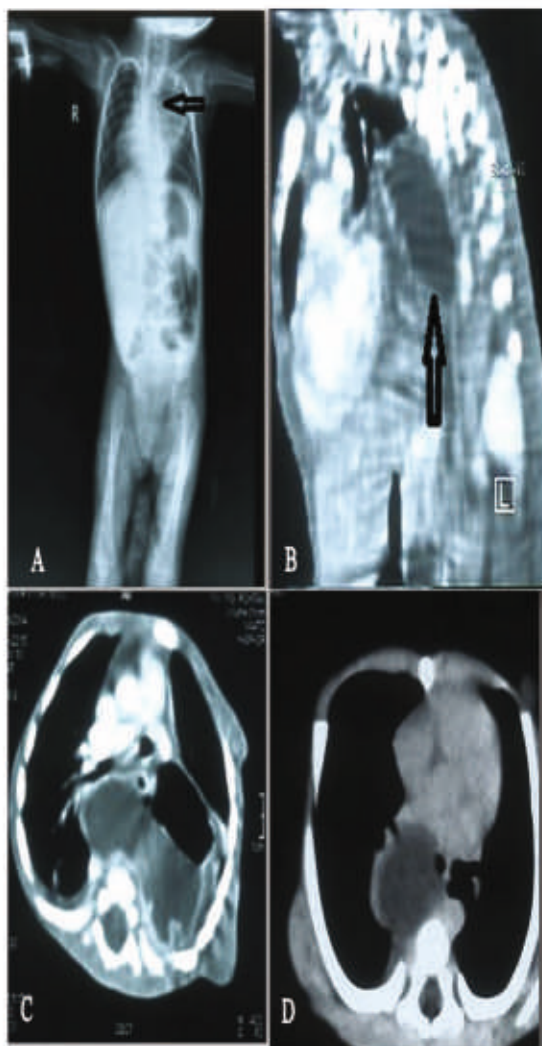


Figure 1: Chest radiograph (postero-anterior view) showing scoliosis and upper hemi vertebrae (A). CT scan showing posterior mediastinal cysts (B; sagittal view, C & D; left and right side cyst)

In 5 cases the cysts were located on the right side while in two cases on the left side in posterior mediastinum. In 3 cases (all right sided) there were pure mediastinal cysts without any abdominal component while in two cases (one right and one left sided) the cysts were thoraco-abdominal. In one case the right sided mediastinal cyst was associated with ileal duplication cyst while one patient who presented with rectal bleeding had left mediastinal cyst which was communicating with ileum through the diaphragm. This was misdiagnosed on CT scan as diaphragmatic hernia because of the presence of air fluid level in thorax. Six out of the seven cysts were non communicating type and all were associated with vertebral anomalies mainly hemi and butterfly vertebrae. Three cases (two right sided and one left sided) were associated with scoliosis. The spinal cord was normal and there was no intraspinal communication with the cyst in any of our patients. Complete excision of the cyst through a

posterolateral thoracotomy was done. In one case with associated duplication cyst, firstly the abdominal cyst was excised and then the mediastinal cyst was excised through thoracotomy. In thoraco-abdominal cysts the inner lining of abdominal part was excised along with the mediastinal cyst leaving behind the outer one because it was not possible to excise the complete cyst through a thoracic approach. In the patient who presented with rectal bleeding and underwent emergency laparotomy, resection of the communication of neurenteric cyst with ileum was done at first and later on the thoracic part was removed through thoracotomy, which was removed on the 5th post operative day. The excised masses were sent for histopathology which on gross examination revealed cystic structures filled with mucinous to straw coloured fluid (**Figure 2**).

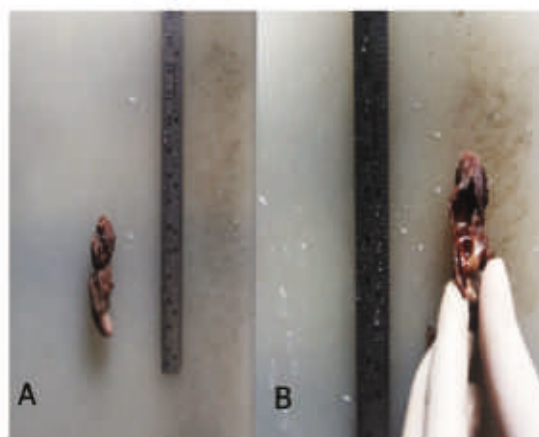


Figure 2: (A&B). Gross appearance of the duplication cyst revealing gut wall like lining on cut section.

Microscopy showed cysts lined by tall columnar to transitional to cuboidal epithelium along with muscularis mucosae. Outside this there were submucosa and bilayered muscularis layer (**Figure 3**).

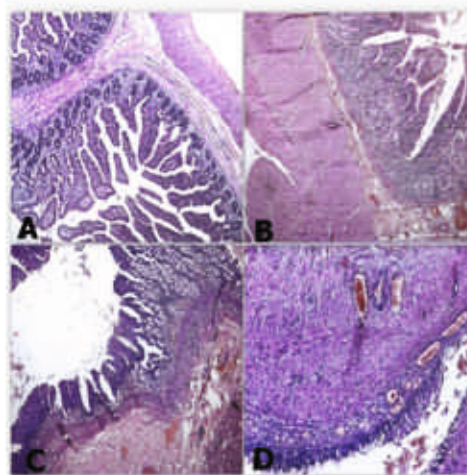


Figure 3: Photomicrograph showing intestinal (A & B), gastric (C) and respiratory lining (D) of wall of various duplication cysts along with underlying muscularis layer (H&E; 40X).

Based on Wilkins and Odome histological classification 5 of the cases were Type II and the other 2 cases were Type I.

The patients were discharged on the 10th postoperative day and are doing well since.

Discussion

Neurenteric cysts are endodermal cysts resulting from failure of separation of ectoderm from the endoderm during the third week of embryonic life. This attachment may prevent fusion of the vertebral bodies which leads to spinal anomalies.⁶ Persistence of only a part of the tract can produce isolated intraspinal cyst.

The most common location is the cervicothoracic region and the mediastinal cysts are reported mainly at right paravertebral location on ventral aspect of spinal cord.⁷ In our study 3 cases had pure mediastinal cyst, 2 thoraco-abdominal, one mediastinal along with a separate ileal duplication cyst and another one with a mediastinal cyst communicating with the ileum near the ileo-caecal valve. Five of the cysts were on the right side at ventral aspect of spinal cord while two were on the left side in posterior mediastinum. According to the literature patients age can range from 5 weeks to 52 years.⁸ Our patients had an age range from 4 months to 8 years with one case diagnosed antenatally.

Neurenteric cysts in mediastinum can compress adjacent structures like lungs, airways, heart and great vessels, and those with coexisting intraspinal lesions may give rise to central nervous system (CNS) abnormalities. The clinical course depends mainly on the extent of displacement and functional impairment of the adjacent viscera, and on the associated CNS complications. They may cause pulmonary hypoplasia and severe postnatal respiratory distress requiring prolonged ventilatory support.⁹ Our patients presented

with recurrent chest infections along with dysphasia in two cases and none of our case required pre or postoperative ventilator support.

X-ray and CT scan can demonstrate congenital anomalies of the spine like spina bifida, butterfly vertebra, lack of segmentation, partial fusion, scoliosis along with the cyst. MRI shows exact site/extent of the lesion along with its relationship with the spinal cord or any intraspinal component.

The most common vertebral anomalies were hemi vertebra and butterfly vertebra. There was no intraspinal component. Surgical excision is advised even in asymptomatic cases to avoid the complications like compression of surrounding visceral structures and vessels. The approach is through posterior thoracotomy route. If there are two components of neurenteric cyst like in one of our case where there was thoracoabdominal involvement, total excision in a single operation can be carried out.

Histologically although these are endodermal lined cysts, variation in constituting components also occur which is divided in to three groups by Wilkins and Odome:

Type I is characterized by a single lining of pseudostratified, cuboidal or columnar epithelium on the basement membrane which could also have a cilia lying. Type II is similar to type I along with the addition of nerve ganglion, mucous or serous glands, lymphoid tissue, cartilage, bone, fat, elastic fibres or smooth muscle. Type III is similar to type II along with ependymal or glial tissue.

Though rare but neurenteric cyst should be kept in mind while dealing with mediastinal cysts of paediatric age group as prognosis is excellent after complete surgical excision.

References

1. Fernandes ET, Custer MD, Burton EM, et al: Neurenteric cyst: surgery and diagnostic imaging. *J Pediatr Surg* 1991; 26(1):108-10.
2. Strollo DC, Rosado-de-Christenson ML, Jett JR: Primary mediastinal tumors: part II. Tumors of the middle and posterior mediastina. *Chest* 1997; 112: 1344-2.
3. Wilkinson CC, Albanese CT, Jennings RW et al: Fetal neurenteric cyst causing hydrops: Case report and review of the literature. *Prenat Diagn* 1999; 19:118-21.
4. Sreedhar M, Menon S, Varma G et al: Cervico-thoracic neurenteric cyst – a case report. *Indian Journal of Radiology and Imaging* 2006; 16(1):99-102.
5. Agarwal N, Kaur N, Miglani R et al: Thoraco-abdominal neuroenteric cyst: a case report and review of the literature. *SAJCH* 2011; 5:55-7.
6. Bourne A: Congenital and developmental abnormalities. In: Whitehead R (ed): *Gastrointestinal and Esophageal Pathology*. New York: Churchill Livingstone, 1995:275-301.
7. Kumar R, Jain R, Rao KM, et al: intra spinal neurenteric cysts: report of three paediatric cases. *Childs Nerv Syst* 2001;17(10):584-8.
8. Rauzzino MJ, Tubbs RS, Alexander E 3rd et al: Spinal neurenteric cysts and their relation to more common aspects of occult spinal dysraphism. *Neurosurg Focus* 2001;10:1.
9. Adzick NS, Harrison MR, Crombleholme TM et al: Fetal lung lesions: Management and outcome. *Am J Obstet Gynecol* 1998; 179:884-9.