

A CASE OF NEURENTERIC CYST*

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An eight-month-old female infant with a neurenteric cyst situated in the lumbar region is presented because of its rarity and because it can mimic other neurological malformations which occur on the same site. Microscopic examination of the surgically excised cyst revealed Water-Paccini corpuscles, lymphoid follicles, intestinal mucosa and meningeal tissue which originated from two ectoderm and endoderm-germ layers. *Key words:* neurenteric cyst, enterogenous cyst, Intestinoma, teratoid cyst.

Neurenteric cysts are rare malformations caused by the disturbance in closure of the neurenteric canal occurring between the 18th - 21st day following conception. The exact etiology of this complex malformation remains unknown¹. It is suggested that the failure of interposition of the mesoderm which forms the normal covering around the spinal cord and persistent opposition of endodermal and ectodermal derivatives give rise to these cysts². Smith³ called these malformations "dorsal fistula remnants", but they have also been termed "teratoma-like-cyst", "enterocystoma", "foregut cyst", "gastrogenic cyst", "intestinoma", "enterogenous cyst", etc.^{1, 4}. These cysts are always situated between the gastrointestinal tract and the skin of the back. They may be prevertebral, postvertebral and occasionally intraspinal in type. It is reported that intracranial localization is very rare. The pons and medulla oblongata are the sites of localization of this malformation¹.

Neurenteric cysts are occasionally associated with gastrointestinal, vertebral and central nervous system malformations¹⁻⁵.

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In this study, we wish to report a case of neurenteric cyst because it is a rare entity and resembles other neurological malformations.

Case Report

An eight-month-old female infant was admitted to the Karadeniz Technical University Farabi Hospital because of a swelling in the lumbar region. Physical examination revealed a soft, tumoral mass located in the lumbar region and covered by skin. The remainder of the physical examination was within normal limits. On computerized tomography posterolateral laminar defects were detected at the 2nd and 3rd lumbar vertebrae. After a diagnosis of meningocele was established, the mass was excised. Macroscopic examination of the excised biopsy material revealed an off-white nodular mass, 5 x 4 x 2 cm in diameter. Microscopic examination of the mass revealed lymphoid follicles, intestinal mucosa, lipid tissue and Water-Paccini sensitive nerve endings covered by meningeal tissue and skin, which were regularly organized (Figs. 1a, b).

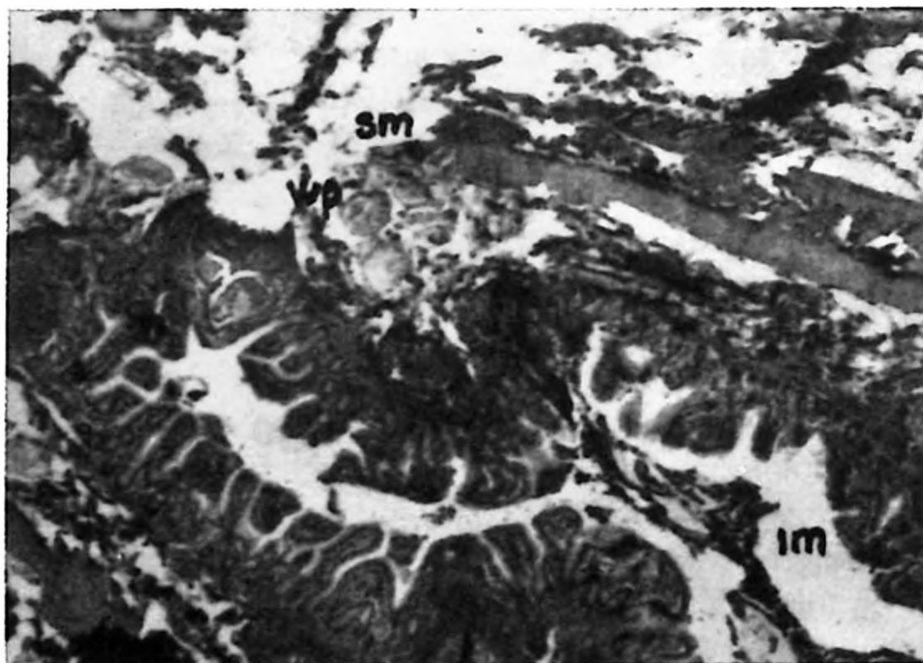


Fig. 1a: Microscopic examination of excised cyst.

Discussion

Neurenteric cysts are rare congenital malformations and the largest well documented series (19 cases) has been published by The Hospital For Sick Children, Toronto⁵.

Neurenteric canal closure defect is the main cause of the disease⁵. The presence of female sex chromosomes in the cysts in male patients strongly suggests their

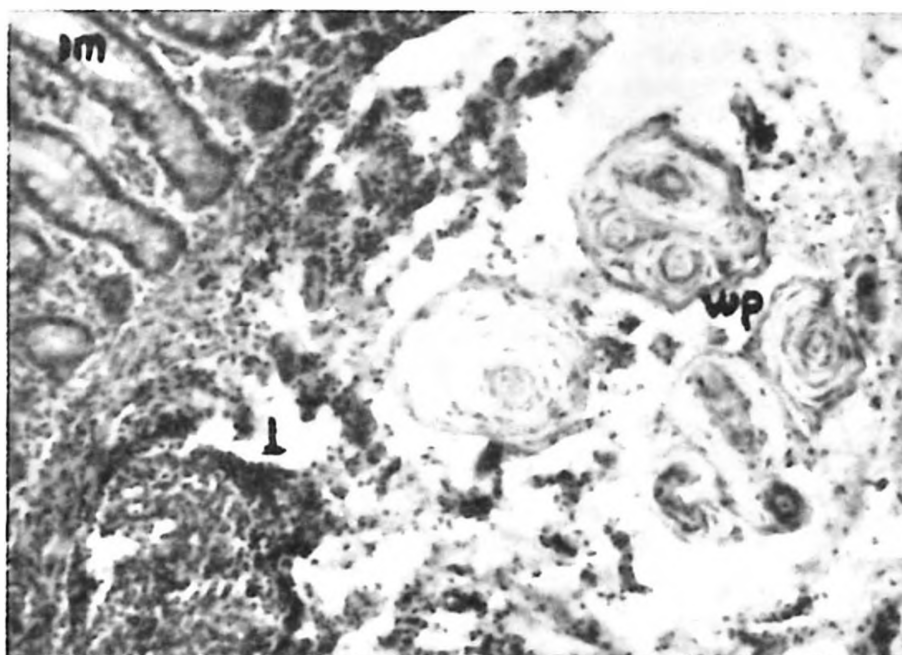


Fig. 1b: Microscopic examination of excised cyst.

teratomatous origin. There is a moderate male preponderance³. Their most common site is the cervico-thoracic vertebral region. However, they may also be situated in the medulla oblongata, pons, posterior mediastinum, and in the pelvis². In our case, the cyst was located in the lumbar region. Localization is usually extramedullary, as in our patient¹. These cysts rarely mimic multiple sclerosis, other cerebromotor dysfunctions, mediastinal, vertebral and gastrointestinal malformations or hydrocephalus. A preoperative diagnosis of meningocele was made in our patient. Other authors have also established such a diagnosis in these cases^{3,6}. Ciliated pseudostratified or cubic Periodic-Acid-Schiff positive gastrointestinal-type epithelial covering of the cysts and neuroglial elements are the main elements^{1,7}. As was described by Hecker⁸, we also observed Water-Paccini corpuscles in our patient. In addition to these elements, we detected lymphoid follicles. The main therapeutic approach is surgical excision of the cyst, which was carried out in our patient.

REFERENCES

1. Zalatnai A. Neurenteric cyst of medulla oblongata a curiosity. *Neuropediatrics* 18:40, 1987.
2. Moore KL. *The Developing Human: Clinically Oriented Embryology* (2nd ed). Philadelphia: WB. Saunders, 1967, pp 113-118.
3. Smith JR. Accessory enteric formations: a classification and nomenclature. *Arch Dis Child* 35:87, 1960.
4. Vinter HV, Gilbert JJ. Neurenteric cysts of the spinal cord mimicking multiple sclerosis. *Can J Neurol Sci* 8:159, 1981.

5. Superina RA, Ein SH, Humphreys RP. Cystic duplications of the esophagus and neurenteric cyst. *J Pediatr Surg* 19:527, 1984.
6. Hashimoto T, Nakamura N, Yasue M, et al. Intraspinal neurenteric cyst associated with increased intracranial pressure (author's transl). *Neurol Med Chir (Tokyo)* 21:1183, 1981.
7. Marchiori PE, Scaff M, Callegaro D, et al. Cisto neuroentérico: registro de un caso. *Arq Neuropsiquiatr* 39:478, 1981.
8. Hecker WC, Hollmann G, Stück B. [Persistent neuroenteric canal and its relation to the development of coccygeal teratoma, prevertebral and postvertebral enterogenous cysts and spina bifida]. *Langenbecks Arch Klin Chir (Ger)* 302:662, 1963.