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pertension associated with these AV fistulas of the brain and the spinal cord.

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Neurenteric Cyst or Teratomatous Cyst

To THE EDITOR: I read the recent article by LeDoux, et al. (LeDoux MS, Faye-Petersen OM, Aronin PA, et al: Lumbosacral neurenteric cyst in an infant. Case report. J Neurosurg 78:821–825, May, 1993) and disagree with their diagnosis. It is incomprehensible why they diagnosed their case as a neurenteric cyst. It appears to me that they made their diagnosis only from the clinical appearance of an intra- and extraspinal cyst and then from the histological examination. Indeed, the term “neurenteric cyst” used to be employed for an intraspinal cyst-forming lesion of maldevelopmental origin, but nowadays it has been applied to a cyst composed only of epithelium and connective tissue.1 It is a synonym for enterogenous cyst.2 The epithelium contains ciliated cells and goblet cells. Squamous cells may be present, but they are usually minor components and interpreted as a result of squamous metaplasia.2–5

In the case described by LeDoux, et al., the lesion was composed of nonkeratinized stratified squamous epithelium and foci of smooth-muscle and adipose tissue. It was made up of endodermal and mesodermal components. Such a lesion should be distinguished from a cyst composed only of epithelium of endodermal origin and connective tissue.1–3 It should be classified as a teratomatous cyst; thus, I suggest the case presented was one of a teratomatous cyst. Le Doux, et al., stated that the lesion in their case did not conform to previously reported teratomatous cysts, because such cysts had not been described as having both intra- and extraspinal components. It is not reasonable to deny a teratomatous cyst based on the distribution of the lesion. They also stressed the absence of Barr bodies in the specimen. The presence of Barr bodies in the epithelial cells of cyst wall of a male patient may support the diagnosis of teratomatous cyst, but their absence cannot be used to deny the diagnosis. Rewcastle and Francoeur5 stated that two of four teratomatous cysts of male patients had no sex chromatin.

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References


To THE EDITOR: I read the paper by LeDoux, et al., describing a “lumbosacral neurenteric cyst in an infant” with great pleasure (LeDoux MS, Faye-Petersen OM, Aronin PA, et al: Lumbosacral neurenteric cyst in an infant. Case report. J Neurosurg 78:821–825, May, 1993). I do not, however, agree completely that the term “teratomatous cyst” is controversial. Rather, I contend that embryological theories are inconsistent and all seem to explain only parts of the clinical spectrum from normal spinal anatomy to extreme spinal dysraphism as well as from teratomatous to neurenteric/enterogenous cysts.

To conform to the definition of neurenteric cyst, some connection of the lesion to the abdominal or thoracic cavity should be present.4 This means that either a connection of the cyst itself to the abdominal and/or thoracic cavity or some form of anterior rachischisis must always exist. If no spinal dysraphism is present, one should conclude that a displacement of primordial germ cells6 or the outgrowth of an early embryonic tissue rests has occurred to explain any tergalinal intradural (cystic) tumor. On the other hand, if spinal dysraphism is found, a concurrent intraspinal trigerminal tumor can also be explained by persistence of the neurenteric canal7 or the presence of an accessory neurenteric canal.1 In this case, both teratomatous and neurenteric cysts can occur, but differentiation between the two can only be made by pathological examination. Pathological studies of neurenteric cysts frequently show a more differential gastrointestinal-type epithelial lining (with a mucosal, a muscular and a stromal layer) with or without ciliae and, in most cases, an abundance of mucin-producing cells is present.3 On the contrary, in teratomatous cysts the growth of the cyst wall is less differentiated; that is to say, the growth of the three different germinal layer-derived elements is more haphazard.6 Also, different types of epithelial lining of the cyst wall can be present in one specimen.

Taking into account these pathological and embryological considerations, the case described can well be classified as a teratomatous cyst instead of a neurenteric cyst. The origin at the conus medullaris with further caudal outgrowth to the presacral region can probably be explained by tumor growth following the path of least resistance. There was no accompanying anterior (or posterior) rachischisis. Pathological examination is inconclusive and cannot help in distinguishing between a neurenteric and a teratomatous cyst. Mucin-producing cells or a well-defined gastrointestinal pattern of the entire cyst wall were not present.

I would like to suggest that it is too early to reject the term “teratomatous cyst,” as further embryological
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References

Response: We appreciate the great interest in our article shown in the letters from Drs. Morita and Hes (Dr. Morita's article was not included in our reference list because of space limitations). Their letters are useful because they help to highlight the many problems associated with the term "teratomaous cyst" and will certainly aid in excluding this term from the neurosurgical and neuropathological literature until more definitive data supporting its use become available.

These two letters support our contention that the term "teratomaous cyst" is controversial and provide disparate views on the appropriate use of the term "neuroneentric cyst." The body of literature on neuroneentric cysts clearly does not support Dr. Morita's statement that the term is "nowadays... applied to a cyst composed only of epithelium and connective tissue." He states that lesions with endodermal and mesodermal components should be classified as "teratomaous cysts." In this regard, it is important to review the relationship of neuroneentric cysts to the development of the enteric canal. The derivatives of the primitive gut include the pharynx, lower respiratory tract, esophagus, stomach, small intestine, pancreas, liver, gallbladder, extrahepatic biliary tree, colon, and rectum. The gastrointestinal tract has common histological mural characteristics consisting of mucosa, submucosa, smooth muscle, and serosa. The endoderm of the primitive gut gives rise to most of the epithelium of the digestive tract. The muscular and fibrous elements are derived from splanchnic mesenchyme. The epithelial lining of the mucosal layer is composed of various combinations of several different cell types including stratified squamous, columnar mucus-secreting, columnar, absorptive, and enterochromaffin cells. In early development, the esophagus is lined by ciliated epithelium. Therefore, the characteristics of individual neuroneentric cysts can vary according to their degree of differentiation. The neuroneentric cyst is typically lined by pseudostratified columnar or cuboidal epithelium which may be ciliated or show mixtures of intestinal, gastric, pancreatic, and/or squamous epithelium. The neuroneentric cyst may have a simple connective-tissue capsule, a connective-tissue wall containing varying amounts and types of mesenchymal tissue, or a multilayered enteric wall. Intraspinal and extraspinal components are frequently connected through anterior vertebral defects. According to Dr. Morita's letter, an intraspinal cystic structure with focci of muscle in its wall which is connected to an intraspinal duplication cyst through an anterior vertebral defect would be classified as a "teratomaous cyst." Obviously, the term has little descriptive value in this context.

The term "teratomaous cyst" also poses particular difficulty when attempting to separate it from "cystic teratoma." Drs. Morita and Hes do not address this problem. Another problem is the observation that neither the presence nor absence of sex chromatin allows for the separation of "teratomaous cysts" from neuroneentric cysts or cystic teratomas.

Dr. Hes inappropriately relies on Lerma, et al., to support his statement that "To conform to the definition of neuroneentric cyst, some connection of the lesion to the abdominal or thoracic cavity should be present." The publication by Lerma, et al., reports the case of a 5-year-old boy with an entirely intraspinal neuroneentric cyst who had no associated vertebral anomalies. Although anterior vertebral defects and intrathoracic and/or intra-abdominal cysts are frequently associated with intraspinal neuroneentric cysts, they need not be present for an intraspinal cyst to be diagnosed as a neuroneentric cyst. Dr. Hes' letter contains other errors and inconsistencies. In his second paragraph, he explains how neuroneentric cysts can be differentiated from "teratomaous cysts" by pathological examination. However, in the next paragraph of his letter, he states that "Pathological examination is inconclusive and cannot help in distinguishing between a neuroneentric and a teratomaous cyst." Finally, he believes that the path taken by the cyst in our patient through the S-4 vertebral body can "probably be explained by tumor growth following the path of least resistance." A much more plausible explanation is that the cyst in our patient and the associated defect in the S-4 vertebral body were developmental in origin.

In conclusion, there is little evidence at present to support the use of the term "teratomaous cyst" since it requires the application of poorly substantiated assumptions and theories. Given the embryopathogenesis of neuroneentric cysts, our histopathological and clinical findings, and the commonly accepted use of the term "neuroneentric cyst," the diagnosis of neuroneentric cyst was made in our case. We await with interest clinical and experimental data that conclusively support use of the term "teratomaous cyst," with the understanding that this information may not change the clinical management of patients with intraspinal neuroneentric or "teratomaous" cysts.

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