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Neuroimaging Features of Neurenteric Cysts: Analysis of Nine Cases and Review of the Literature

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PURPOSE: To gain a better understanding of neurenteric (NE) cysts via correlation of imaging findings and surgical and pathologic data. **METHODS:** The medical records, imaging studies, surgical information, and pathologic material were retrospectively reviewed in nine patients with NE cysts, including seven proved and two very probable cases. **RESULTS:** NE cysts occurred in the cerebellopontine angle in one case and extended from the cerebellopontine angle to the C2 level in a second. In the latter patient and the remaining seven with intraspinal lesions, the NE cyst was always located anterior to the spinal cord. The most common myelographic and CT myelographic appearance was that of a lobulated intradural extramedullary (IDEM) mass. Two patients had an intramedullary NE cyst with a somewhat unusual appearing exophytic IDEM-appearing expansion that can be a characteristic feature of these lesions. MR imaging demonstrated the NE cyst to be isointense to hyperintense relative to cerebrospinal fluid on long TR sequences and isointense or slightly hyperintense to cerebrospinal fluid on T1-weighted images. These signal characteristics correlate with the high-protein-content fluid within the cysts, usually described at surgery as milky or mucinous in character. **CONCLUSION:** The diagnosis of NE cyst should be considered when imaging studies reveal the presence of a lobulated IDEM or an exophytic intramedullary cystic mass, especially in association with anterior spina bifida or other vertebral anomalies. MR can uniquely confirm the cystic nature of these masses and is the method of choice for their imaging investigation. Because cyst recurrence can occur, MR should also be used for long-term patient follow-up.

Index terms: Spinal cord, cysts; Brain, cysts; Spine, magnetic resonance; Spine, computed tomography; Brain, magnetic resonance

Neurenteric (NE) cysts are relatively rare lesions that result from development during or shortly after the third week of embryogenesis, the period of notochordal development. To date, the magnetic resonance (MR) imaging findings of NE cysts have been described in only a few individual cases (1–4) and in one report of two cases (5). We reviewed the myelographic, computed tomography (CT) myelographic, and MR studies of nine patients with NE cysts, together with a review of the literature to gain a better understanding of this uncommon entity.

Materials and Methods

The nine cases were collected from the teaching files of four neuroradiologists. The medical records, imaging examinations, and surgical pathologic material were retrospectively reviewed.

The patients included six males and three females ranging in age from 22 months to 43 years. Myelography and/or CT myelogram studies were available in seven of the eight patients with intraspinal lesions. Four patients had MR examinations including the single case of a cerebellopontine angle (CPA) NE cyst, and three other patients with intraspinal lesions. The MR studies were performed at 0.5 T in two, and at 1.5 T in the other two patients. Final

diagnosis was based upon description of the surgical findings and review of the histologic material with a neuropathologist in seven patients. In one patient (case 3), the pathologic material is not available for review but the clinical history, myelographic, CT, and MR findings all appear classical for NE cyst and were regarded as sufficiently characteristic for definitive diagnosis. In a final patient (case 6), there is no indication for surgery, but the combination of clinical and imaging features makes NE cyst a very probable diagnosis. The clinical and surgical pathologic data and the neuroimaging features are summarized in Tables 1 and 2.

Results

An NE cyst occurred in the CPA in one patient in this series (Fig. 1), and extended from the CPA to the C2 level in another case. The remaining NE cysts were entirely intraspinal in location, including four cervical and two thoracic lesions, and one located at the L1 level in association with a diastematomyelia. Two large cervical NE cysts were intramedullary (Figs. 2 and 3). The rest of the cases all had extraaxial lesions. The NE cyst had an anterior relationship to the spinal cord in each of the eight patients with intraspinal lesions.

Associated vertebral anomalies were present in six patients. In one patient with a high cervical NE cyst, the only abnormality was a cleft in the posterior arch of C1 (case 8). There was enlargement of the anteroposterior and interpediculate dimensions of the cervical spinal canal in a patient with a large lobulated exophytic intramedullary NE cyst in the midcervical region (Fig. 2). In a patient with Chiari III malformation and a probable small midcervical NE cyst by MR, segmentation fusion anomaly (Klippel-Feil) of the upper and midcervical vertebrae was present. In a patient with an NE cyst at the T1-T3 level, fusion of several upper thoracic vertebral bodies was found. One patient had a right posterior mediastinal mass with thoracic kyphosis, multiple thoracic vertebral segmentation fusion anomalies, and anterior spina bifida at T4-T5. A patient with diastematomyelia and an NE cyst at the L1 level also had scoliosis and a posterior spina bifida defect. No vertebral abnormalities were identified in the patients with CPA and CPA to high cervical NE cysts (cases 1 and 7), and in one patient with a midcervical intramedullary NE cyst (case 3).

Myelography and/or CT myelography showed ovoid or lobulated anterior intradural extramedullary (IDEM) masses in three patients. In one patient with a midcervical intramedullary NE cyst, the myelogram and CT myelogram images

demonstrated spinal cord expansion terminating in an exophytic-appearing bilobulated mass, with displacement of an atrophic cord posteriorly below the level of the mass (Fig. 2). A midcervical ovoid expanding intramedullary cyst was found at myelography in another case (Fig. 3). In one patient diagnosed at 5 months of age with a large mediastinal mass and multiple thoracic vertebral segmentation fusion anomalies (Fig. 4), myelography and CT revealed the cord to be ventrally displaced occupying a trough created by an anterior spina bifida defect in dysplastic fused T4-T5 vertebral bodies. An eccentric collection of contrast media ventral to the cord was noted (Fig. 4A) consistent with extension of a diverticulum of the subarachnoid space in relation to the connecting stalk between the spinal cord and the mediastinal cyst. This patient had thoracotomy and resection of the mediastinal cyst. At the time of surgery, a stalk was found extending from the cyst through the anterior spina bifida and through a defect in the dura into the subarachnoid space. After opening the dura, the stalk could be identified attached to the anterior aspect of the cord over a length of approximately 2 cm, and forming a shallow groove in the cord from which it was separated by thickened arachnoid. In a final patient, CT myelography was initially performed because of a thoracic vertebral body compression fracture after motor vehicle accident trauma, and disclosed a diastematomyelia at the L1 level. There was a cartilaginous or bony septum extending from posteriorly between the two hemicords, and a round mass anterior to the divided cord in the midline that had attenuation characteristics similar to those of the spinal cord (Fig. 5).

MR was obtained in four patients, including three with intraspinal lesions and a single patient with a CPA NE cyst. Two patients had MR findings of an intramedullary cystic lesion: on T1-weighted images it appeared hypointense to cord and was mildly hyperintense to cerebrospinal fluid (CSF) in one patient with a residual cyst postoperatively (Fig. 2D); it appeared more prominently hyperintense to CSF in the second patient, whose clinical history strongly suggested cyst recurrence at age 17 after original surgery at 9 years of age. Surgery at age 24 in the latter case failed to identify or drain the recurrent NE cyst that was shown by MR (Figs. 3B and 3C), and the patient has not had another operation to date. In the third patient, MR findings included tectal deformity consistent with Chiari malformation,

TABLE 1: Clinical and surgical-pathologic findings in nine patients with neurenteric cysts

Case No.	Age (years)/ Sex	Clinical History	Surgical Description	Pathologic Findings
1	43/F	Headache; normal neurologic exam	Gelatinous tumor was removed in one piece	Ciliated columnar epithelium with goblet cells reminiscent of respiratory epithelium; electron microscopy confirmed diagnosis of NE cyst
2	19/M	Progressive LE stiffness and weakness for 6 months. UE weakness for 1 week. Spastic quadriparesis on admit exam. Significant improvement UE motor strength postoperatively	Cyst puncture yielded cloudy fluid; cord biopsy obtained followed by resection of exophytic IM cysts	Cyst wall of tall columnar epithelium with abundant mucus
3 Presumed NE cyst	26/F	Severe interscapular pain relieved by drainage of "spinal cord cyst" at age 9; progressive right hemiparesis since age 17; repeat surgery at age 24; no functional improvement postoperatively	Atrophic cord trapped by adhesive bands within pseudomeningocele reported at second operation; no cord tumor or cyst was said to be able to be identified	Not available
4	22 weeks/M	Spastic paraplegia and thoracic kyphosis noted at birth	Resection of right posterior mediastinal cyst containing mucinous fluid; cyst stalk extended through anterior spina bifida and dural defect attaching to anterior spinal cord	Mediastinal mass contained respiratory epithelium; connecting stalk was fibrous with no mucous glands present
5	31/F	Patient was evaluated because of thoracic vertebral compression fracture status post MVA	Diastematomyelia without dural septum at L1; resection of midline cystic mass anterior between split spinal cord	Cyst wall of stratified squamous epithelium
6 Chiari III presumed NE cyst	16/M	Surgery as infant for occipital encephalocele/shunt for hydrocephalus; recent behavioral outbursts; increasing frequency of seizures	No clinical indications for surgery	Not available
7	19/M	Neck pain with torticollis for 3 weeks; quadriparesis for 3 days; ambulating with minimal assistance postoperatively	Anterior IDEM cystic mass with aspiration of milky fluid; cord appeared decompressed status postremoval of cyst capsule	Cyst wall of tall simple columnar epithelium with goblet cells and occasional ciliated cells
8	13/M	Neck stiffness and UE weakness 1 month; PTA; flaccid UE paresis and spastic paraparesis on admission; marked improvement of UE strength, normal gait postsurgery	Membranous cyst anterior to cord at C1-C2; slightly opaque fluid on cyst aspiration; return of normal cord pulsation postcyst removal	Cyst wall of cuboidal and low columnar epithelium with some goblet cells

Note.—NE = neureuteric; LE = lower extremity; UE = upper extremity; MVA = motor vehicle accident; IM = intramedullary; IDEM = intradural extramedullary; NA = not available; PTA = prior to admission.

TABLE 1: Continued

Case No.	Age (years)/ Sex	Clinical History	Surgical Description	Pathologic Findings
9	3/M	Tonsilitis, neck stiffness for 2 weeks followed by left hemiparesis; spastic quadriparesis on admission; complete recovery with no residual weakness post-surgery	Upper thoracic anterior cyst extramedullary mass displacing spinal cord posteriorly Mass was excised in toto	Cyst wall of ciliated columnar epithelium with few scattered low cuboidal cells

Note.—NE = neureuteric; LE = lower extremity; UE = upper extremity; MVA = motor vehicle accident; IM = intramedullary; IDEM = intradural extramedullary; NA = not available; PTA = prior to admission.

TABLE 2: Neuroimaging findings in nine patients with neurenteric cysts

Case No.	Myelography	CT Myelography	MR
1	ND	ND	Extraaxial right CPA mass; heterogeneous on T1-WI; hyperintense, to gray matter on spin density-WI. Isointense to CSF on T2-WI; mass did not enhance with Gd-DTPA
2	Bilobed IDEM appearing masses anterior to cord at C4-C5	Lobular IM masses from anterior cord; cord atrophy posteriorly and caudally	Postoperatively defined residual IM cord cyst with associated cord atrophy and resection of bilobulated exophytic cyst components
3 Presumed NE cyst	Bulbous IM anterior cord expansion C3-C5	IM cord enlargement with cord atrophy cephalad and caudad	Ovoid IM mass hypointense to cord, hyperintense to CSF on T1 WI
4	Right posterior mediastinal mass; multiple vertebral segmentation fusion anomalies; midthoracic kyphosis; anterior spina bifida T4-T5; ventrally displaced cord T4-T6 with anterior crescentic collection of contrast material	Cord occupying V-shaped trough of dysplastic T4-T5 with apparent connection to right posterior cystic mediastinal mass through anterior spina bifida	ND
5	ND	Diastematomyelia at L1; round anterior midline mass between divided cord; CT attenuation same as spinal cord	ND
6 Chiari III presumed NE cyst	ND	ND	Multiple upper cervical segmentation fusion anomalies; tectal deformity of Chiari malformation; upper cervical IDEM 5-mm mass anterior to cord, mildly hyperintense to cord on T1 WI, prominently hyperintense to cord on spin density WI, hyperintense to CSF on T2 WI
7	Multilobular anterior IDEM mass CPA to C2 level	ND	ND
8	Ovoid anterior IDEM mass at C1-C2 level	ND	ND
9	Upper thoracic vertebral body fusion with IDEM obstructing mass anterior to cord at T1-T3	ND	ND

Note.—CPA = cerebellopontine angle; IDEM = intradural extramedullary; IM = intramedullary; WI = weighted images; CSF = cerebrospinal fluid; ND = not done.
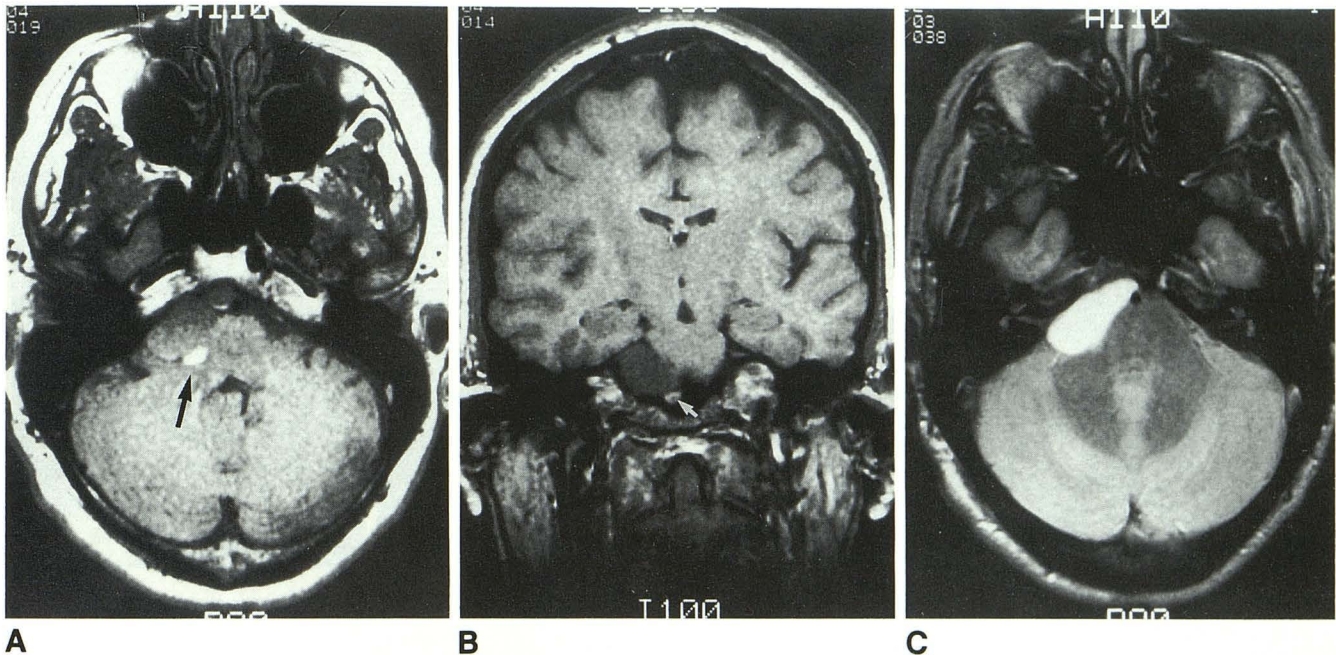
A   **B**   **C**

Fig. 1. A 43-year-old woman with right CPA and prepontine cistern NE cyst.

A and *B,* Transaxial (*A*) and coronal (*B*) T1-weighted (600/20 (TR/TE)) precontrast images at 1.5 T. The mass shows heterogeneous signal and flattens the pons. Note increased signal intensity in posteroinferior portion of the mass (*arrow*). This may represent hemorrhage or increased protein in dependent portions of the cyst. There was no abnormal enhancement with Gd-DTPA.

C, Transaxial spin-density image (3000/30) shows prominent hyperintensity of the mass relative to gray matter with extension anteromedially adjacent to the basilar artery.

and the patient had Klippel-Feil anomaly and surgery in infancy for occipital encephalocele and hydrocephalus (Chiari III). In this patient, a small high cervical IDEM mass anterior to the cord appeared mildly hyperintensive to cord on T1-weighted images and prominently hyperintense on spin-density images, and was hyperintense to CSF on T2-weighted images. Finally, case 1 of this series had MR of an approximately 2 × 3.5 × 2 cm (T × AP × SI) CPA and prepontine cistern mass that appeared moderately hyperintense to CSF and hypointense to cerebellum on T1-weighted images, except for a small posteroinferior bright signal component. Spin-density images showed prominent hyperintensity relative to brain and CSF, and the mass was iso- to slightly hyperintense to CSF on T2-weighted images. There was no enhancement with Gd-DTPA. At surgery, a gelatinous tumor that was able to be removed in one piece was described.

Discussion

NE cysts result from dysgenesis occurring during the third embryonic week, at the time of notochordal development and the transitory existence of the neurenteric canal. The treatment

of this subject in the English radiologic literature up to 1986 has been rather limited (1, 2, 6–10). Several more recently published manuscripts (3–5, 11) have been, with one exception (5), isolated case reports. NE cysts are known to occur in the posterior fossa, spinal canal, posterior mediastinum, and abdomen, and occurrence in more than one of these locations in the same patient has also been reported (6, 12–15).

During the third week of gestation, invagination and migration of cells at the primitive streak and node of the embryonic disk result in the formation of the mesodermal germ layer and the notocord, producing separation of the ectoderm and endoderm layers of the embryo (Figs. 6A and 6B). The definitive notochord detaches from its underlying endoderm and forms the midline axis that will serve as the basis for future development of the axial skeleton and, in turn, has an inductive influence on formation of the neural tube from the overlying ectoderm. During this time of notochordal development, the small canal at the level of the primitive pit that transiently connects the yolk sac, or future alimentary tract of the embryo, with the amniotic cavity is termed the "neurenteric canal." The neurenteric canal allows temporary contact between endoderm and the
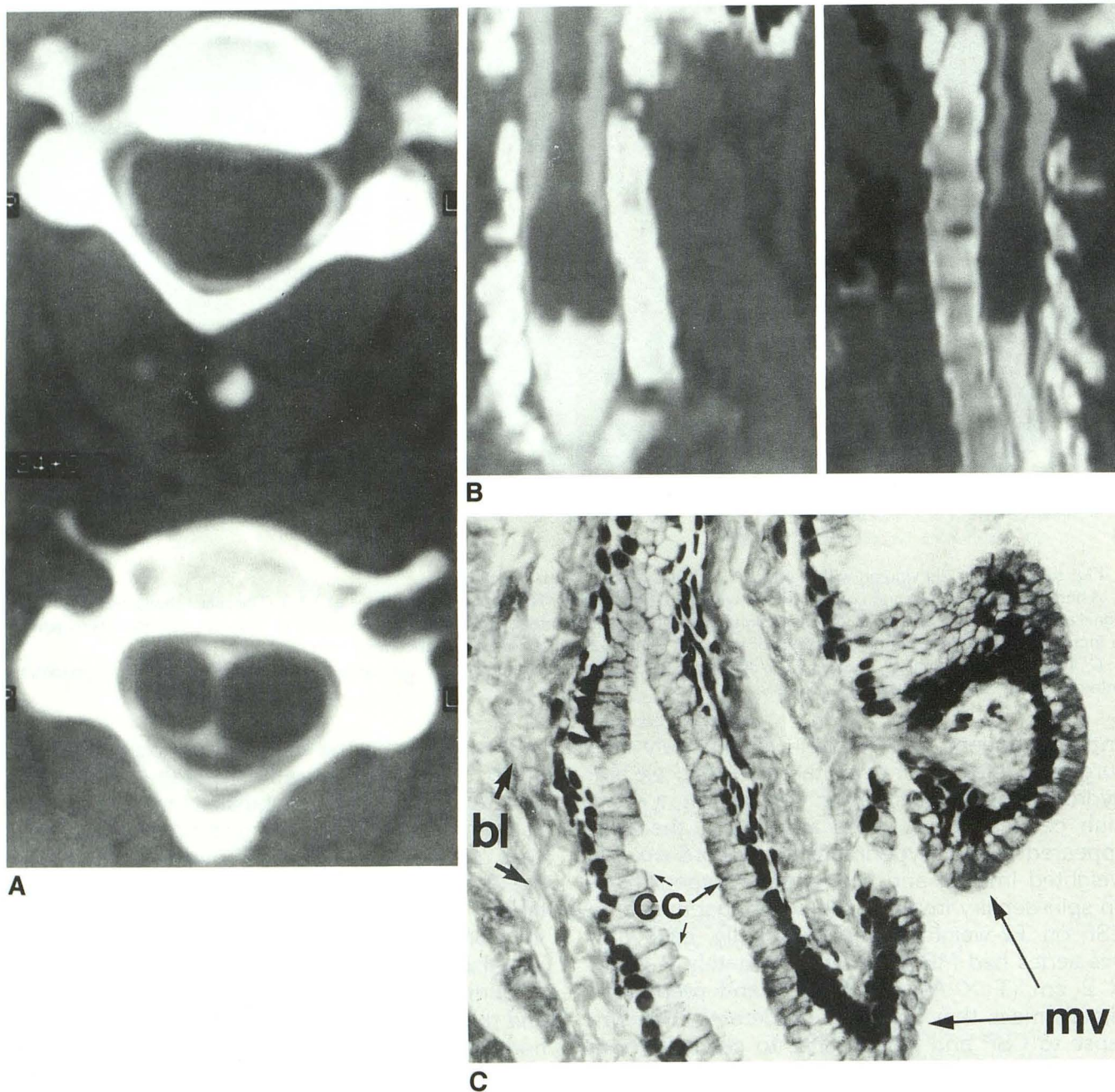
Fig. 2. Cervical cord NE cyst in a 19-year-old man.

A and *B*, CT myelographic transaxial images (*A*) and coronal and sagittal reformatted views (*B*) demonstrate spinal cord enlargement with termination in a bilobulated mass. The ribbonlike atrophic cord posterior to the lobular masses is apparent on the sagittal reformatted view.

C, Histologic section. Light microscopy demonstrates a single layer of columnar cells (*cc*) resting on a basal lamina (*bl*). Abundant mucin droplets are present in the apical portions of the cells. Microvilli (*mv*) and occasional cilia are seen.

developing neuroectoderm. Its persistence may interfere with notochordal development and result in formation of an NE cyst or, alternatively, persistent endoectodermal adhesions or adhesions between notochord and endoderm may produce notochordal dysgenesis and a resultant NE cyst (Figs. 6C and 6D). Depending on the ability to repair the notochordal derangement completely or incompletely, the NE cyst may then be associated with coexistent spinal abnormalities.

The previous literature has been confusing with regard to terminology of these lesions, variously referred to as neurenteric cysts (2, 4–6, 11,
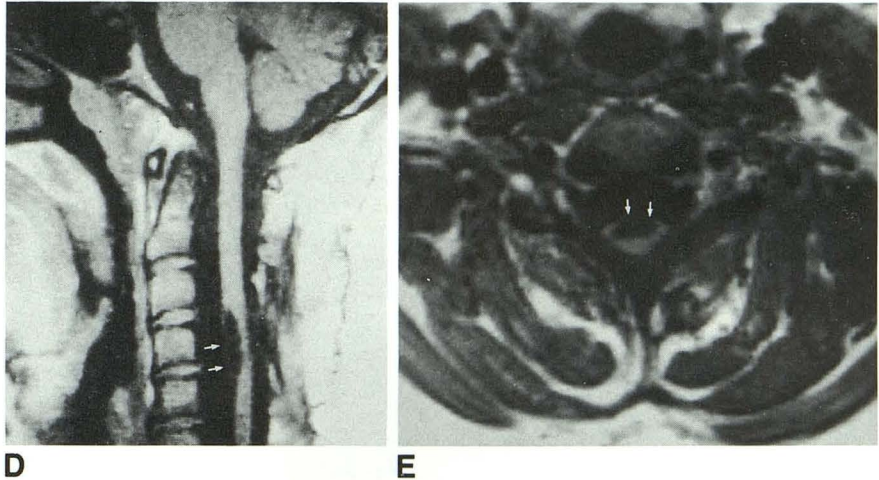
Fig. 2. —*Continued*. *D* and *E*, Spin-density (3300/20) sagittal (*D*) and T1-weighted (700/26) transaxial (*E*) images obtained 2 years after surgery. The exophytic lobulated component of the NE cyst has been resected, and the residual intramedullary component is still seen in both sections (*arrows*).
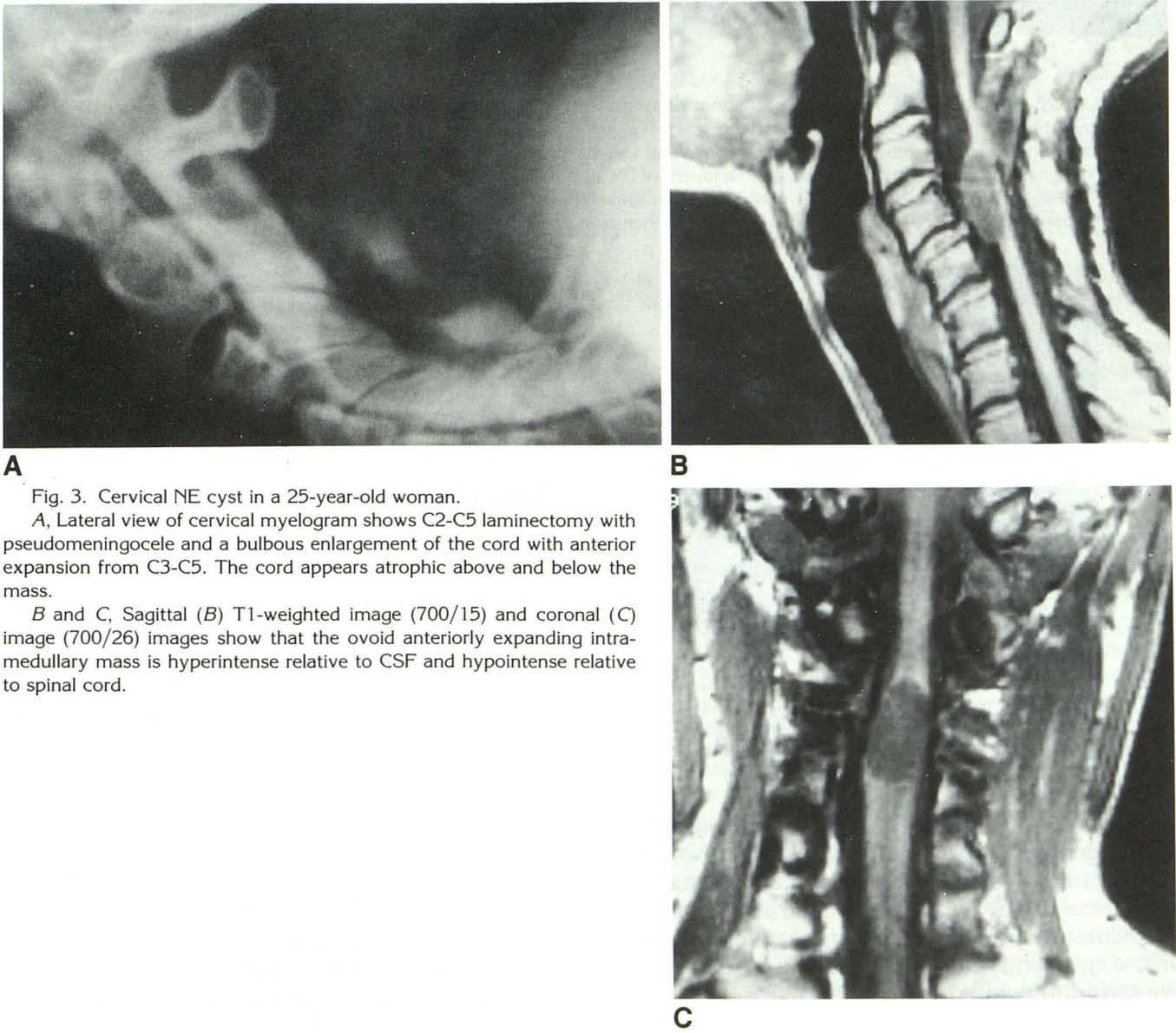

Fig. 3. Cervical NE cyst in a 25-year-old woman.

A, Lateral view of cervical myelogram shows C2-C5 laminectomy with pseudomeningocele and a bulbous enlargement of the cord with anterior expansion from C3-C5. The cord appears atrophic above and below the mass.

B and *C*, Sagittal (*B*) T1-weighted image (700/15) and coronal (*C*) image (700/26) images show that the ovoid anteriorly expanding intramedullary mass is hyperintense relative to CSF and hypointense relative to spinal cord.

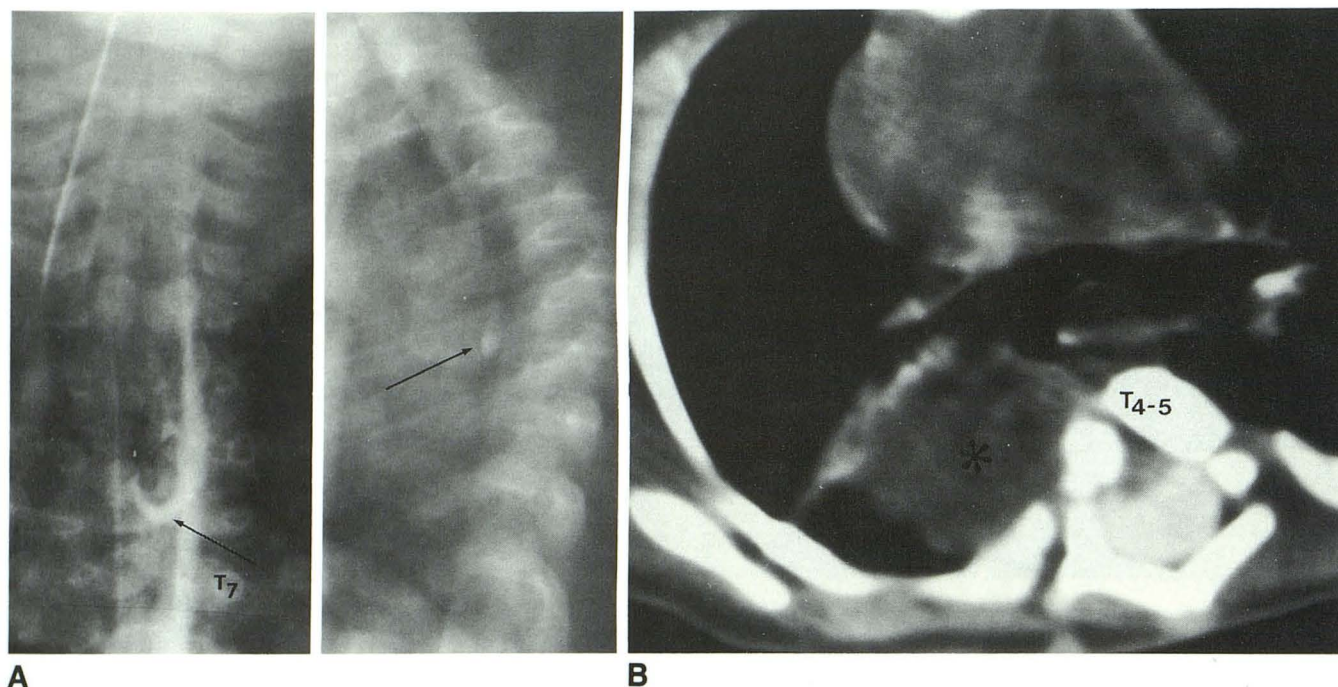**A**   **B**

Fig. 4. A 22-month-old male infant with posterior mediastinal cyst connecting to arachnoid of anterior spinal cord via extension of fibrous stalk through T5 anterior spina bifida defect.

A, Thoracic myelogram anteroposterior supine and lateral views shows multiple vertebral segmentation anomalies and hook-shaped contrast collection partially surrounding site of attachment of NE cyst stalk to meninges (*arrows*). Note thoracic kyphosis.

B, CT myelogram transaxial section at level of T5 anterior spina bifida shows posterior mediastinal cyst (*asterisk*), defect in dysplastic fused vertebral bodies (*T4-T5*), and spinal cord occupying triangular expansion of the anterior spina bifida.
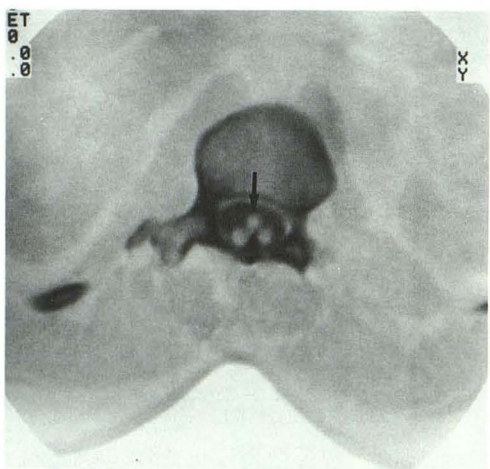

Fig. 5. A 31-year-old woman with NE cyst projecting anteriorly from cleft in a split spinal cord at the L1 level. Transaxial video reversed CT myelogram at L1 shows a round structure anterior to the split cord (*arrow*) which proved to be cystic.

16–21), enteric cysts (22), enterogenic (23) and enterogenous cysts (3, 7, 9, 12, 15, 24–35), endodermal cysts (36), gastrogenic (37) and gastroenterogenous cysts (38), gastrocytoma (39), archenteric cysts (40), cyst of foregut origin (13, 14, 41), ependymal cyst (42, 43), teratomatous cysts (44, 45), and teratoma (46). For the sake of

clarity and because it properly emphasizes the embryologic origins of this entity, we regard neurenteric cyst as the preferred term.

The histology of NE cysts is indicative of their origin from some portion of the alimentary tract, most often the foregut. The lining mucosa may have features of esophagus, stomach, small bowel, or some combination of these. A less differentiated NE cyst with a more embryonic type of epithelium will have a basement membrane and a thin cyst wall composed of cuboidal or columnar cells that tend to contain mucin and to have a basal orientation of nuclei (26, 36, 38). In some cases, the cyst epithelium may be reminiscent of respiratory epithelium (2, 4, 36). These histologic features were present in the seven patients for whom surgical pathologic material was available for review in this series (Table 1 and Fig. 2C).

Anterior spina bifida is the most helpful of the osseous developmental anomalies that can accompany an NE cyst. It has long been recognized that in the presence of an anterior spina bifida, a connection between the spinal cord or meninges and some portion of the alimentary tract should be suspected (47). The defect in the vertebral
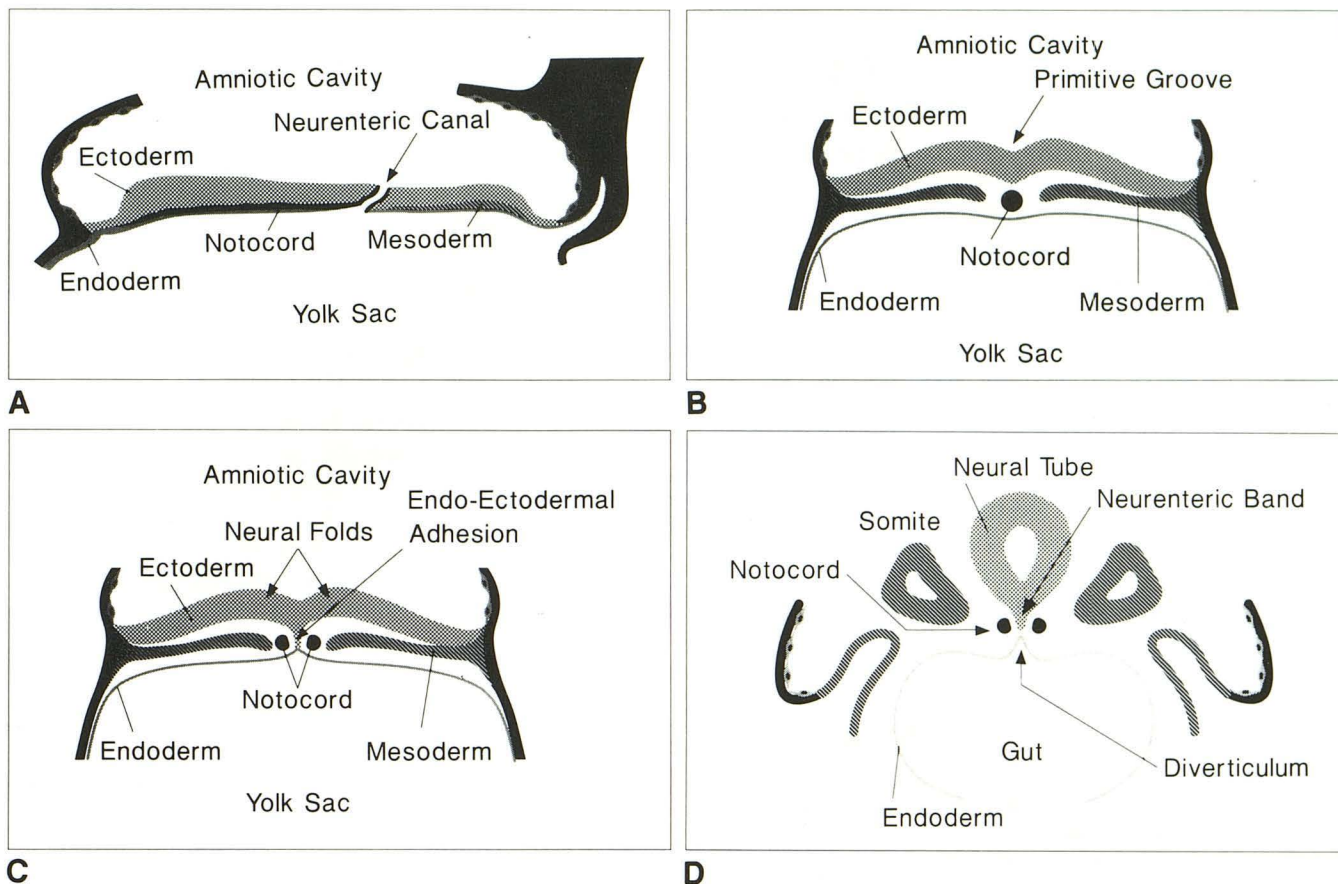
Fig. 6. Normal embryologic anatomy, third gestational week (*A* and *B*) and postulated mode of formation of an NE cyst (*C* and *D*).

A and *B*, Midsagittal (*A*) and coronal (*B*) sections show transient connection of the amniotic cavity and yolk sac via the neurenteric canal.

C and *D*, Persistence of the neurenteric canal, failure of complete detachment of notochord from endoderm, or persistent ectoendodermal adhesions may result in notochordal dysgenesis and formation of traction diverticulae and/or an NE cyst. The notochrodal derangement may or may not undergo later repair.

body or bodies may be variable in size and in some cases may consist of only a small cleft or a vertebral body tunnel that may require CT or tomography for its demonstration (37). The connection may consist of a tube composed of nervous tissue and smooth muscle, or of a fibrous cord. In the case of a tubular connection, the lumen extends throughout only infrequently, and more often exists as a diverticulum extending from either the alimentary or the meningeal end of the connection (Figs. 4 and 6D). Patient 4 of this series is a classic example of the situation in which a posterior mediastinal NE cyst is connected by a stalk or "neurenteric band" to the meninges and cord via an anterior spina bifida and dural defect. A small meningeal diverticulum partially surrounded the attachment of the NE cyst stalk to the pia-arachnoid and cord (Fig. 4). A number of similar cases have been reported (12, 14, 48, 49). In addition to anterior spina

bifida and vertebral body tunnels, other spinal abnormalities that have been found in association with NE cyst include Klippel-Feil anomaly (present in case 6), other segmentation fusion anomalies (present in cases 4 and 9), spinal canal enlargement (case 2), diastematomyelia (case 5), and posterior spina bifida defects (cases 5 and 8). NE cyst therefore should always be considered in the differential diagnosis when vertebral developmental abnormalities are present in association with a posterior mediastinal or abdominal cyst or an intraspinal cystic mass.

In the eight patients having intraspinal lesions, myelography demonstrated a multilobulated type of IDEM NE cyst in three cases (cases 7–9). This is a common appearance of NE cyst and has been described in numerous reports. Less common are the imaging features demonstrated by cases 2 and 3, in which an exophytic intramedullary NE cyst produces a prominent lobulated or

bulbous IDEM-appearing anterior cord expansion (Figs. 2 and 3). This unusual imaging appearance, when present, is sufficient to include NE cyst in the differential diagnosis, especially if vertebral body dysplasia is also present. In previous descriptions of similar cases, it has sometimes been suggested that this appearance resulted from invagination of an IDEM NE cyst into the spinal cord (1, 2). However, in our cases, the merging of the cephalic extent of the cyst into the cord was very suggestive of an intramedullary origin. In both types of intraspinal lesions, the NE cyst was always anterior to the spinal cord. Posterior location of an intraspinal cystic mass should lead one to favor other diagnoses such as cystic teratoma, arachnoid cyst, or ependymal cyst, rather than an NE cyst. In the previous literature, intraspinal NE cysts have almost always been reported to be anterior in location with only rare exceptions (36).

Finally, one NE cyst occurred in the CPA in this series (case 1) and another extended from the CPA to the C2 level (case 7). The posterior fossa is a known site of occurrence of NE cysts (9, 30–32), with NE cysts also having been reported intraaxially within the medulla (21), the fourth ventricle (33), and the cisterna magna (34).

MR evaluations in our cases and in previous reports have shown NE cysts to be isointense to hyperintense relative to CSF and hyperintense to spinal cord on long TR sequences. On T1-weighted sequences, most are isointense or mildly hyperintense to CSF and hypointense relative to spinal cord. Occasionally, homogeneous very bright signal intensity within a small NE cyst may be seen on both short and long TR images. This occurred in case 6 and, although not surgically proven in this series, the MR findings were virtually identical to a proven high thoracic NE cyst illustrated by DiChiro et al (1). We believe the homogeneous high signal intensity within these smaller lesions to be caused by their high protein content rather than hemorrhage. In patients having cyst puncture, aspirated cyst contents are usually described as cloudy, milky, opaque, or mucinous, as occurred in four patients in this series (cases 2, 4, 7, and 8). The CPA NE cyst in case 1 was described as gelatinous in character. Variation in cyst protein content (30) and hemorrhage into NE cysts have both been reported (13, 27), and the small portion of higher signal intensity seen in the mass in case 1 (Figs. 1A and 1B) was thought most likely due to either an area of hemorrhage or to higher protein content within

a loculated portion of the cyst. No enhancement occurred in our patients 1 and 3, and lack of enhancement was also reported in the two cases discussed by Pierot et al (2).

Our case material and review of the literature indicate that the clinical course of older children and adults with NE cyst may be either rapidly or intermittently progressive. In infants and young children, an acute onset and progression seems to be more frequent (6, 9, 15, 19, 23, 38). An episode of trauma may initiate or exacerbate symptoms in some patients (9, 15, 16, 22, 25, 26). Numerous reports have documented resolution of severe neurologic deficits including paraplegia and quadriplegia after surgical decompression of an NE cyst (6, 12, 15, 19, 23–25, 36, 45). This was true for cases 7–9 of this series. Other patients have had limited neurologic improvement after surgery (14, 36, 45, 50, 51); case 2 of this series, who had cord atrophy, would be included in this latter group. Most previous reports do not include long-term follow-up patients, precluding accurate estimation of the incidence of cyst recurrence. However, recurrence has been documented in a number of instances (9, 14, 28, 29); case 3 of this series also appears to represent an example of a recurrent cervical intramedullary NE cyst. Awareness of this possibility is important both from the point of view of patient education and management, inasmuch as long-term follow-up examination would appear advisable.

NE cysts represent an uncommon entity. In the CPA, other more commonly occurring lesions including arachnoid cyst; neuroepithelial cyst; epidermoid, dermoid, and teratomatous cystic tumors; and cystic meningioma would all have to be considered first. Cystic metastases and abscesses usually should be able to be distinguished on the basis of their enhancement characteristics and thickness of their marginal rim of solid tissue. Parasitic cysts are an additional possibility, especially in endemic regions, but in some cases, multiplicity of lesions would be helpful.

Differential diagnosis of intraspinal cystic masses includes consideration of arachnoid cysts (52) and postinflammatory or posttraumatic subarachnoid cyst (53) as well as, again, cystic dermoid, epidermoid and teratoma masses, parasitic cysts, and ependymal cysts, as above. In addition, an intramedullary NE cyst would have to be distinguished from cystic cord tumors (54), syringomyelia, IDEM masses with an associated syrinx (55), and myelomalacia.

NE cysts should be considered in the differential diagnosis of intraspinal cystic masses in particular when osseous spinal developmental defects are present in association with a multilobulated IDEM or an exophytic intramedullary mass. The additional presence of a mediastinal or abdominal cyst will be helpful in some cases. We hope that the case material forming the basis of this report and review of the literature will serve to improve the awareness and enhance the understanding of the embryology and the clinical and imaging features of these lesions.

References

1. Di Chiro G, Doppmann JL, Dwyer AJ, et al. Tumors and arteriovenous malformations of the spinal cord: assessment using MR. Radiology 1985;156:689–697
2. Kantrowitz LR, Pais MJ, Burnett K, et al. Intraspinal neurenteric cyst containing gastric mucosa: CT and MRI findings. Pediatr Radiol 1986;16:324–327
3. Aoki S, Machida T, Sasaki Y, et al. Enterogenous cyst of cervical spine: clinical and radiological aspects (including CT and MRI). Neuroradiology 1987;29:291–293
4. Geremia GK, Russell EJ, Clasen RA. MR imaging characteristics of a neurenteric cyst. AJNR: Am J Neuroradiol 1988;9:978–980
5. Pierot L, Dormont D, Queslati S, et al. Gadolinium-DTPA enhanced MR imaging of intradural neurenteric cysts. J Comput Assist Tomogr 1988;12:762–764
6. Neuhauser EBD, Harris GBC, Berrett A. Roentgenographic features of neurenteric cysts. AJR: Am J Roentgenol 1958;79:235–240
7. Gleeson JA, Stovin PGI. Mediastinal enterogenous cysts associated with vertebral anomalies. Clin Radiol 1961;12:41–48
8. Gimeno A, Lopez F, Figuera D, Rodrigo L. Neuroenteric cyst. Neuroradiology 1972;3:167–172
9. Chavda SV, Davies AM, Cassar-Pullicino VN. Enterogenous cysts of the central nervous system: a report of eight cases. Clin Radiol 1985;36:245–251
10. Woodring JH, Daniel TL. Mediastinal analysis emphasizing plain radiographs and computed tomograms. Med Radiogr Photogr 1986;62:2–49
11. Rodacki MA, Teixeira WR, Boer VHT, et al. Intradural, extramedullary high cervical neurenteric cyst. Neuroradiology 1987;29:588
12. McLetchie NGB, Purves JK, Saunders RLdeCH. The genesis of gastric and certain intestinal diverticula and enterogenous cysts. Surg Gynecol Obstet 1954;99:135–141
13. Fallon M, Gordon ARG, Lendrum AC. Mediastinal cysts of fore-gut origin associated with vertebral abnormalities. Br J Surg 1954;41:520–533
14. Dorsey JF, Tabrisky J. Intraspinal and mediastinal foregut cyst compressing the spinal cord: report of a case. J Neurosurg 1966;24:562–567
15. Laha RK, Huestis WS. Intraspinal enterogenous cyst: delayed appearance following mediastinal cyst resection. Surg Neurol 1975;3:67–70
16. Vinters HV, Gilbert JJ. Neurenteric cysts of the spinal cord mimicking multiple sclerosis. Can J Neurol Sci 1981;8:159–161
17. Holcomb GW, Matson DD. Thoracic neurenteric cyst. Surgery 1954;35:115–121
18. Case records of the Massachusetts General Hospital (case #26-1975). N Engl J Med 1975;293:33–38
19. Klump TE. Neurenteric cyst in the cervical spinal canal of a 10-week old boy. J Neurosurg 1971;35:472–476
20. D'Almeida AC, Stewart DH Jr. Neurenteric cyst: case report and literature review. Neurosurgery 1981;8:596–599
21. Zalatnai A. Neurenteric cyst of medulla oblongata: a curiosity. Neuropediatrics 1987;18:40–41
22. Case records of the Massachusetts General Hospital (case #46122). N Engl J Med 1960;262:623–627
23. Piramoon AM, Abbassioun K. Mediastinal enterogenic cyst with spinal cord compression. J Pediatr Surg 1974;9:543–545
24. Mohanty S, Rao CJ, Shukla PK, et al. Intradural enterogenous cyst. J Neurol Neurosurg psychiatry 1979;42:419–421
25. Fabinyi GCA, Adams JE. High cervical spinal cord compression by an enterogenous cyst. J Neurosurg 1979;51:556–559
26. Harriman DGF. An intraspinal enterogenous cyst. J Pathol Bacteriol 1958;75:413–419
27. Rhaney K, Barclay GPT. Enterogenous cysts and congenital diverticula of the alimentary canal with abnormalities of the vertebral column and spinal cord. J Pathol Bacteriol 1959;77:457–471
28. Holmes GL, Trader S, Ignatiadis P. Intraspinal enterogenous cysts. Am J Dis Child 1978;132:906–908
29. Mizuno J, Fiandaca S, Nishio S, O'Brien MS. Recurrent intramedullary enterogenous cyst of the cervical spinal cord. Childs Nerv Syst 1988;4:47–49
30. Kak VK, Gupta RK, Sharma BS, Banerjee AK. Craniospinal enterogenous cyst: MR findings. J Comput Assist Tomogr 1990;14:470–472
31. Small JM. Preaxial enterogenous cysts: Proceedings of the Society of British Neurological Surgeons. J Neurol Neurosurg Psychiatry 1962;25:184
32. van der Wal AC, Troost D. Enterogenous cyst of the brain stem: a case report. Neuropediatrics 1988;19:216–217
33. Afshar F, Scholtz CL. Enterogenous cyst of the fourth ventricle. J Neurosurg 1981;54:836–838
34. Giombini S, Lodrini S, Migliavacca F. Intracranial enterogenous cyst. Surg Neurol 1981;16:271–273
35. Miyagi K, Mukawa J, Mekaru S, et al. Enterogenous cyst in the cervical spinal canal. J Neurosurg 1988;68:292–296
36. Palma L, Di Lorenzo N. Spinal endodermal cysts without associated vertebral or other congenital abnormalities. Acta Neurochir 1976;33:283–300
37. Veeneklaas GMH. Pathogenesis of intrathoracic gastrogenic cysts. Am J Dis Child 1952;83:500–507
38. Bale PM. A congenital intraspinal gastroenterogenous cyst in diastematomyelia. J Neurol Neurosurg Psychiatry 1973;36:1011–1017
39. Knight G, Griffiths T, Williams I. Gastrocystoma of the spinal cord. Br J Surg 1955;42:635–638
40. Hutchinson J, Thompson JD. Congenital archenteric cysts. Br J Surg 1953;41:15–20
41. Olenik JL, Tandatnick JW. Congenital mediastinal cysts of foregut origin. Am J Dis Child 1946;71:466–476
42. Hyman I, Hamby WB, Sanes S. Ependymal cyst of the cervicodorsal region of the spinal cord. Arch Neurol Psychiatry 1938;40:1005–1012
43. Moore MT, Book MH. Congenital cervical ependymal cyst: report of a case with symptoms precipitated by injury. J Neurosurg 1966;24:558–561
44. Kubie LS, Fulton JF. A clinical and pathological study of two teratomatous cysts of the spinal cord containing mucus and ciliated cells. Surg Gynecol Obstet 1928;47:297–311
45. Rewcastle NB, Francoeur J. Teratomatous cysts of the spinal canal with "sex chromatin" studies. Arch Neurol 1964;2:91–99
46. Bucy PC, Buchanan DN. Teratoma of the spinal cord. Surg Gynecol Obstet 1935;60:1137–1144
47. Bell HH. Anterior spina bifida and its relation to a persistence of the neurenteric canal. J Nerv Ment Dis 1923;57:445–462

48. Saunders RL de CH. Combined anterior and posterior spina bifida in a living neonatal human female. *Anat Rec* 1943;87:255–278

49. Bentley JFR, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. *Arch Dis Child* 1960;35:76–88

50. Hosoi K. Intradural teratoid tumors of the spinal cord. *Arch Pathol* 1931;11:875–883

51. Adams RD, Wegner W. Congenital cyst of the spinal meninges as cause of intermittent compression of the spinal cord. *Arch Neurol Psychiatry* 1947;58:57–69

52. Kendall BE, Valentine AR, Keis B. Spinal arachnoid cysts: clinical and radiological correlation with prognosis. *Neuroradiology* 1982;22:224–225

53. Sklar E, Quencer RM, Green BA, et al. Acquired spinal subarachnoid cysts: evaluation with MR, CT myelography, and intra-operative sonography. *AJNR: Am J Neuroradiol* 1989;10:1097–1104

54. Williams AL, Haughton VM, Pojunas KW. Differentiation of intramedullary neoplasms and cysts by MR. *AJR: Am J Roentgenol* 1987;149:159–164

55. Quencer RM, El Gammal T, Cohen G. Syringomyelia associated with intradural extramedullary masses of the spinal canal. *AJNR: Am J Neuroradiol* 1986;7:143–148