Intradural spinal bronchiogenic cyst

Case report

Department of Surgical Neurology, Royal Infirmary and Western General Hospital, and Department of Neuropathology, University of Edinburgh, Edinburgh, Scotland

The successful treatment of an intradural cervical cyst of apparent bronchial origin in a 14-year-old girl is recorded. Its possible relationship to enterogenous cysts and the split notochord syndrome is discussed.

KEY WORDS spinal cord · bronchiogenic cyst · enterogenous cyst · split notochord syndrome · teratoid of cord · spina bifida

The occurrence of intradural spinal non-neoplastic cystic lesions is exceedingly rare. Many of those described have been considered to be enterogenous and have been distinguished from cystic teratomas. The purpose of this paper is to present a case of intradural cervical cyst which was considered to be of bronchial origin and was associated with vertebral anomalies. So far as we know, a bronchiogenic cyst in this location has not been described previously.

Case Report

A 14-year-old girl was first admitted to the Western General Hospital on November 17, 1969, with a 2-year history of intermittent neck pain, and pain and stiffness of the left arm. The pain was aggravated by physical exercise and relieved by analgesics and rest. It extended into the fingers but not the thumb and was associated with slight numbness of the fingers. Her previous health had been satisfactory and there was nothing of note in the family history.

First Examination. The patient was found to have severe limitation of all neck movements, especially of flexion to the right side. The base of her neck was very broad (Fig. 1). There was a cutaneous capillary hemangioma about 4 cm in diameter in the midline in the neck and local tenderness over the base of the neck posteriorly. Radiographs of the cervical spine (Fig. 2) revealed spina bifida occulta of the laminae of C-5 to T-1, congenital fusion of the bodies of the C-6 and C-7 vertebrae and their neural arches, and narrowing of the C5-6 disc space. Her symptoms disappeared without treatment, and she was discharged without further investigation.

In October, 1970, and May, 1971, the neck pain returned.

Second Examination. The patient was readmitted on September 27, 1971, because of symmetrically increased deep reflexes.
Intradural spinal bronchiogenic cyst

noted in both legs. A myelogram (Fig. 3) showed a partial hold-up of the Myodil column in the lower cervical region opposite C-6 and C-7 where a rounded filling defect to the left of the midline, suggestive of an intradural mass, was seen.

Operation. A lower cervical laminectomy was carried out on October 25, 1971. There was no connection between the cutaneous hemangioma and the dura. Opening of the dura revealed a pearly, bilobed cystic lesion; this was partially aspirated. The cystic fluid was a viscous, opaque, white fluid which was later reported as being amorphous, devoid of cells, and faintly eosinophilic; it stained strongly with mucous stains. The cyst was deeply embedded in the substance of the cord, lay slightly more to

Fig. 1. Photograph showing broad neck.

Fig. 2. Preoperative radiographs showing anteroposterior (left) and lateral (right) views of spina bifida occulta of the laminae of C-5 to T-1, congenital fusion of the bodies of the C-6 and C-7 vertebrae and their neural arches, and narrowing of the C5-6 disc space.
Junkoh Yamashita, Anthony F. J. Maloney and Phillip Harris

FIG. 3. Myodil myelogram showing anteroposterior (left) and lateral (right) views of a rounded filling defect suggestive of an intradural mass at the level of C-6 and C-7.

the left of the midline than to the right, and had virtually divided the cord into two parts, each half being displaced laterally and forward. The cyst was easily dissected off the cord. The ventral aspect of the cord was not examined to avoid damaging the already thinned neural tissue.

Postoperative Course. Recovery was uneventful. Plain radiographs were repeated but no evidence of an anterior spina bifida was detected (Fig. 4). When examined 11 months later, the patient had a full range of movements of the cervical spine, no motor or sensory symptoms, and no disturbance of micturition. Deep reflexes in the legs remained hyperactive.

Pathological Examination. The specimen (Fig. 5) measured 2.0 x 1.5 x 1.4 cm; it contained two fairly large cysts, one of which was collapsed, and several much smaller cysts. Microscopically the cysts were caricatures of small bronchi (Fig. 6). They were lined by epithelial cells ranging from pseudo-stratified ciliated columnar through cuboidal to flattened. The epithelium rested on fibrous tissue in which unstriped muscle fibers, quite large plates of cartilage, and mucous glands were seen. A mild chronic inflammatory reaction was noted in the connective tissue in some places.

Discussion

According to Willis, a teratoma is a true tumor or neoplasm composed of multiple
Intradural spinal bronchiogenic cyst

is often difficult to tell from its description whether a lesion is a malformation or a benign teratoma. It seems mandatory to consider some of the pathogenic mechanisms that have been proposed to explain the occurrence of enterogenous cysts and other similar anomalies.

Brun and Saldeen described an intraspinal enterogenous cyst associated with an Arnold-Chiari malformation and came to the conclusion that enterogenous cysts are of developmental origin and can be differentiated from teratomatous cysts if they have the following characteristics: 1) the wall completely duplicates a gastrointestinal wall but is devoid of nonintestinal tissue components; 2) the cervical or thoracic lesion is either intramedullary or ventral to a normal cord, or ventral to and between the components of a split cord; and 3) the lesion is associated with some kind of vertebral anomaly.

Fallon, et al., described two cases of mediastinal cyst of foregut origin associated with vertebral anomalies. They regarded these cysts as the end result of an ectoendodermal adhesion in the early stages of embryonic life. McLetchie, et al., reported a case in which an accessory stomach extended upward through the diaphragm and was connected by "neuro-enteric" strands to the vertebral column, which showed a fused hemivertebra of C-6. They expressed a similar view as to its mechanism of origin and stated that this condition is most frequent in the cervical and thoracic regions because the cephalic end of the notochord develops first and any organ is most sensitive to disturbance in development at its moment of inception.

Rhaney and Barclay reported three cases that exhibited deformities of the gut and vertebral column; in two, enterogenous cysts were present, one of which lay in the cord. They re-evaluated the theory of abnormal separation of germ layers in early embryonic development and concluded that the origin of intraspinal enterogenous cysts located ventral to the spinal cord can be explained by this theory. They suggested that enterogenous cysts lying dorsal to the spinal cord may have originated because the ectoderm of the primitive streak is probably capable...
Fig. 6. Photomicrographs of cyst.  
Upper Left: Whole thickness of cyst wall showing from above downward mucous membrane, cartilage, and connective tissue. H & E, x 60.  
Upper Right: Bronchial type of epithelium (at the right) resting on cartilage (at the left). Note mucous glands beneath epithelium (central). H & E, x 120.  
Lower Left: Ciliated columnar pseudostratified epithelium is seen lining the cyst wall. H & E, x 700.  
Lower Right: Part of cyst wall incompletely lined by a single layer of epithelium (above) resting on fibrous tissue containing a little plain muscle, H & E, x 110.
Intradural spinal bronchiogenic cyst of forming both endoderm and paraxial mesoderm.

Recently, Silvernail and Brown\(^{6}\) reported a case in which an intramedullary cervical enterogenous cyst was associated with multiple anomalies of the vertebral bodies, a mediastinal enterogenous cyst, malrotation of the bowel, and a diverticulum of the small intestine.

Bentley and Smith\(^{1}\) have developed the concept of the split notochord syndrome. They postulated that an embryo may develop with partial duplication and separation of the notochord, through which the ventrally situated yolk sac or gut anlage endoderm may herniate and rupture, with a resultant fistula between the yolk sac and the amniotic cavity that passes through and divides the future cord and spine. Subsequent differential growth of the embryo tends to close the fistula, and the site and size of the resultant lesion depends on the degree of obliteration attained by this process. Persistent remnants of yolk sac may differentiate into tissues characteristic of any part of the gut or its embryological derivatives. This theory is more satisfactory than the ectoendodermal adhesion theory because it explains a wider variety of congenital anomalies, particularly enterogenous cysts located dorsal to the spinal cord. The cutaneous capillary hemangioma in our case is also better understood in the light of this theory.

Embryologically\(^{3}\) the respiratory system is developed from the primitive foregut. Mediastinal cysts of foregut origin\(^{3}\) are classified as bronchiogenic, esophageal, and enterogenous. Bronchiogenic cysts are formed of tissues proper to the respiratory tract. Cysts in the wall of the esophagus are also of respiratory origin.

In the present case the lesion is comprised only of cystic structures resembling mature bronchi. Since there are no tissues from other germ layers it cannot be termed a teratoma. If the intimate relationship between the respiratory and alimentary systems in the early stages of embryonic development is taken into consideration, one can readily interpret the present case as a variant of the so-called spinal enterogenous cyst.

References
5. Rhaney K, Barclay GPT: Enterogenous cysts and congenital diverticula of the alimentary canal with abnormalities of the vertebral column and spinal cord. *J Path Bact* 77:457–471, 1959

Address reprint requests to: Phillip Harris, Esq., F.R.C.S.E., Department of Surgical Neurology, Western General Hospital, Crewe Road, Edinburgh EH4 2XU, Scotland.