Intradural bronchogenic cysts

N Wilkinson, H Reid, D Hughes

Abstract

The pathological findings of an intradural and extramedullary cyst in the mid cervical spinal canal are described in a 55 year old woman who presented with a short history of pain and paraesthesia of the right arm. Intradural well defined solitary cystic lesions in the spinal canal are uncommon, their pathogenesis is poorly understood, and their nomenclature is confusing. In this case the cyst was a bronchogenic cyst; these are a rare form of such cysts and they are thought to be a malformation arising from a split notochordal syndrome and not a teratoma.

(*J Clin Pathol* 1992;45:1032-1033)

Case report

A 55 year old woman presented with a two week history of increasing pain and paraesthesia in her right arm. She also had neck stiffness. Other relevant history included a complete duplex urinary system on the right side and partial duplex on the left side. There

was also a hairy naevus in the midline over the cervical spine posteriorly at the level of C7/T1. A myelogram showed an intradural, extramedullary tumour located posteriorly at C3/C4 disc level. A dorsal scoliosis convex to the left with some degenerative changes were also noted, but no other vertebral abnormalities were seen (Figs 1A and B).

A C3/C4 laminectomy was performed and the lesion, which was adherent to the posterior columns of the spinal cord, was partly decompressed by aspiration of its contents and then removed. The cyst contents were viscous, opaque, and white.

After surgery the pain disappeared and at one year follow up there were no symptoms of pain nor paraesthesia, and neurological signs were not evident.

Pathological findings

An oval cystic nodule measuring $1.0 \times 0.8 \times 0.4$ cm suspended on a short pedicle was excised. On histological examination it was found to be a unilocular, thin walled cyst. The latter was lined by pseudostratified ciliated columnar epithelium with interspersed goblet cells and an underlying basement membrane (fig 2). The cyst wall included an oval mass of hyaline cartilage, strands of smooth muscle, and seromucinous glands all lying within a fibrovascular connective tissue stroma. The pedicle contained nerve fibres, ganglion cells and mature adipose tissue. No pacinian corpuscles were identified.

Immunoperoxidase stains showed that there was positivity for epithelial membrane antigen (EMA) but not for glial fibrillary acidic protein (GFAP).

Discussion

The histopathological appearances of intradural extramedullary cysts help differentiate the various types: arachnoid, ependymal, epidermoid and dermoid cysts. All of these have an inner lining of cells on a connective tissue wall without a basement membrane. The lining cells of arachnoid cysts are negative for GFAP¹; those of ependymal cysts are positive for GFAP, but negative for keratin and glycoprotein. In contrast, epidermoid and dermoid cysts are lined by stratified squamous epithelium and are positive for keratin; in the dermoid cysts there are hair follicles and sebaceous glands in the wall.

Those cysts with basement membrane underlying their epithelium have been called by a plethora of names and the literature on

Department of
Pathological Sciences,
University of
Manchester, Oxford
Road, Manchester
M13 9PT
N Wilkinson
H Reid

Department of
Radiology, Hope
Hospital, Eccles Old
Road, Salford
D Hughes

Correspondence to:
Dr H Reid

Accepted for publication
14 May 1992

Figure 1 Myelogram. The oval well defined mass is outlined by the contrast medium in the upper cervical spinal canal (arrows). (A) antero-posterior view; (B) lateral view.

Figure 2 The cyst wall is lined by ciliated pseudostratified epithelium. The wall has the organoid structure of a bronchus with lamina propria, smooth muscle, seromucinous glands, a nerve bundle and cartilage (haematoxylin and eosin).

these cysts is confusing.² They have been termed enterogenous, neurenteric, epithelial, bronchogenic, teratoid and teratomatous. Wilkins and Odom¹ placed them into three categories—type A, B, and C.

Category A

A simple cyst lined by epithelium on a basement membrane with a thin wall of connective tissue.

Category B

An epithelial lined cyst with a wall containing tissues found along the gastrointestinal or tracheobronchial tree.

Category C

As B, but with ependymal and glial tissues as an intrinsic part of the lesion and not from the adjacent spinal cord.

The histopathological features of this cyst with an epithelium containing both ciliated and goblet cells lying on a basement membrane and the wall containing smooth muscle and cartilage having an organoid appearance are those of a bronchogenic cyst and are similar to those described by Raney and Barclay,³ Yamashita,⁴ and Ho.⁵ These are of similar histological appearances to the bronchogenic cysts described in the mediastinum of children. These intradural bronchogenic cysts are found on the posterior aspect of the cord.

Using the definition of a teratoma that Bale⁶ used, which is based on that of Willis,⁷ a teratoma is a neoplasm with the power of autonomous growth which is also a malformation composed of multiple tissues foreign to the part and lacking organ specificity. We consider this cyst to be a malformation and not a teratoma, because it shows organ specificity.

Electron microscopic examination by Ho⁵

showed that the epithelial lining is of respiratory type and includes Kulchitsky cells as well as ciliated, goblet cells, and undifferentiated cells. Hirano also reported finding an epithelial cyst and came to the conclusion that it was respiratory in type.⁸ Other reports of the cysts of gastrointestinal type containing specialised gastric cells have been described.

The cyst described here has the characteristics of a bronchogenic cyst, a term we would prefer to use to differentiate this from an enterogenous cyst as defined above. We would also distinguish it from an epithelial cyst which is a cyst with a connective tissue wall lined by epithelium. Teratoid or teratomatous are confusing terms to use and should be avoided.

There are three main hypotheses for the occurrence of these malformations.³⁻⁹ First, as a result of ectoendodermal adhesion in the early stages of embryonic life with abnormal separation of germ cell layers. Secondly, an ectodermal origin as the ectoderm of the primitive streak is capable of forming both endoderm and paraxial mesoderm. Thirdly, there is partial duplication and separation of the notochord through which the ventrally placed 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 differentiated 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. The latter process is called the split notochord syndrome,⁹ and although there was no vertebral malformation the hairy naevus may be indicative of an ectodermal abnormality.

Hirano found tight junctions between the epithelial cells even with the presence of goblet cells, and these may limit the amount of expansion that occurs within the cyst and they are therefore slow growing.⁸ These cysts, when excised, usually relieve the patient's symptoms and full recovery occurs. There are no instances of malignancy developing within them.

We thank Mr P L Richardson for allowing us to publish the clinical details of this case.

- 1 Wilkins RH, Odom GL. Spinal intradural cysts: Epidermoid and dermoid cysts. In: Vinken PJ, Bruyn GW eds. *Handbook of Clinical Neurology*. Amsterdam, North-Holland Publishing Co, 1976:73-82.
- 2 Kosenbach R, Godersky JC, Traynelis VC, Schelper RD. Intradural extramedullary cysts of the spinal canal: clinical presentation, radiographic diagnosis and surgical management. *Neurosurgery* 1992;30:35-42.
- 3 Raney 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;77:457-71.
- 4 Yamashita J, Maloney AFJ, Harris P. Intradural spinal bronchiogenic cyst. Case report. *J Neurosurg* 1973;39:240-5.
- 5 Ho K-L, Tiel R. Intraspinal bronchogenic cyst: ultrastructural study of the lining epithelium. *Acta Neuropathol (Berlin)* 1989;78:513-20.
- 6 Bale PM. Sacrococcygeal developmental abnormalities and tumors in children. *Perspect Pediatr Pathol* 1984;1:9-56.
- 7 Willis RA. *The borderland of embryology and pathology*. London: Butterworths, 1958:410-11.
- 8 Hirano A, Ghatak NR, Wisoff HS, Zimmerman HM. An epithelial cyst of the spinal cord. An electron microscopic study. *Acta Neuropathol (Berlin)* 1971;77:214-23.
- 9 Bentley JFR, Smith JR. Developmental posterior enteric remnants and spinal malformation. The split notochord syndrome. *Arch Dis Child* 1960;35:76-86.