Postgraduate Medical Journal (July 1982) **58**, 424–426

CLINICAL REPORTS

Childhood cervical enterogenous cyst presenting with hemiparesis

P. Y. C. WOO
F.R.C.S

M. M. SHARR
F.R.C.S., M.R.C.P.*

Mersey Regional Department of Medical and Surgical Neurology, Walton Hospital, Rice Lane, Liverpool

Summary

The clinical features of a cervical enterogenous cyst in a one-year-old boy mimicked those of an intracranial mass. Following removal of the cyst the neurological signs rapidly disappeared. The literature is reviewed from histological and clinical aspects and a classification of these cysts is proposed.

Introduction

There have been many reports of intraspinal enterogenous cysts since the first description by Kubie and Fulton (1928) and they have been variously termed neurenteric or gastrogenic cysts. The purpose of this paper is to present an unusual example of such a lesion in a child which produced clinical features more suggestive of an intracranial mass.

Case report

The patient, a one-year-old boy, was initially admitted under a paediatrician with an influenza-like illness for 2 weeks, followed by a 4-day history of apathy, and a 2-day history of severe neck retraction. In addition the parents also noticed a relative lack of movement of both his upper limbs for one day. The child had been born prematurely with a birth weight of 4 pounds but had had normal developmental milestones; the family history was unremarkable.

On examination the child was drowsy, responding only to painful stimuli; there was marked neck stiffness with severe opisthotonus, and a flaccid grade 1 (motor power) left hemiparesis. Lower limb reflexes were brisk but equal with bilateral extensor plantar responses. Because of the clinical features a computerized tomographic scan was done but this was normal; an emergency metrizamide lumbar myelogram was then performed and this showed a complete block at the level of C2 due to a presumed intradural extramedullary mass anterior to the spinal cord (Fig. 1). Subsequent cerebrospinal fluid (CSF) biochemistry was normal.

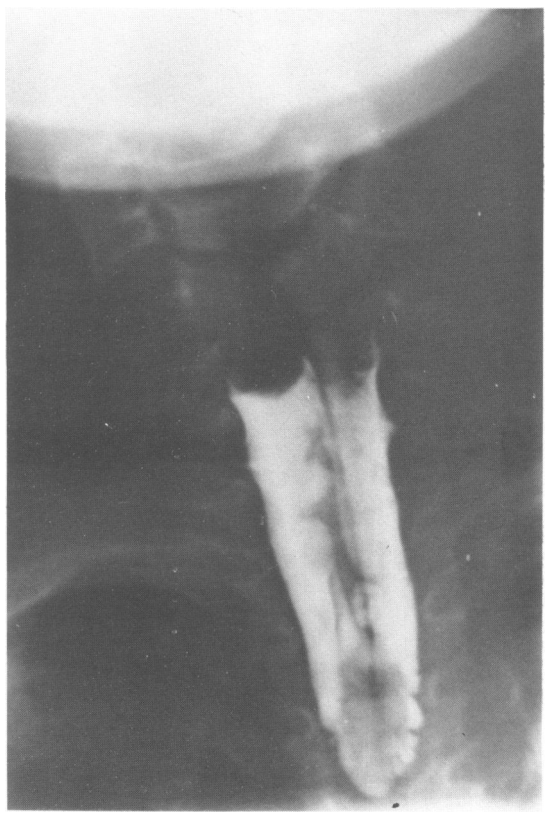

FIG. 1. Metrizamide lumbar myelogram showing complete block at C2.

*Present address: South East Regional Neurosurgical Unit, Brook General Hospital, Shooters Hill Road, Woolwich, London SE18 4LW.

0032-5473/82/0700-0424 $02.00

An upper cervical laminectomy and a small suboccipital craniectomy were carried out and a cyst containing clear colourless fluid was found adherent to the anterolateral aspect of the spinal cord but unattached to any nerve root; the cyst wall was removed. Postoperatively the child made a rapid recovery and, on discharge 2 weeks later, he was able to walk with some aid, since there was still very mild left lower limb weakness present. Subsequent histology revealed a thin-walled cyst composed of mucus-secreting columnar epithelium compatible with an enterogenous cyst (Fig. 2).

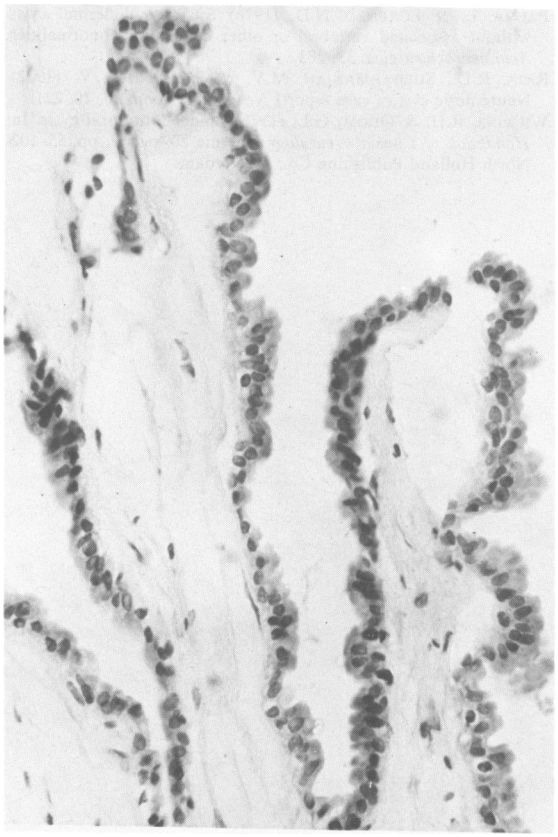

FIG. 2. Histology of cyst wall.

Discussion

Palma and Lorenzo (1976), in their review of enterogenous cyst without bony abnormality, were able to find only 23 cases including 4 of their own. The ages of the patients ranged from 10 weeks to 55 years a marked male predominance (20 patients) (only 4 patients presented under the age of 15 years, 2 male). In addition the majority of cysts (14) were in the cervical region mainly in the lower segments.

Holmes, Trader and Ignatiadis (1978) reported a 13-year-old boy with an intraspinal thoracic cyst; in addition they reviewed 26 paediatric cases in the English literature of whom 18 were male and 8 female (newborn to age 20 years). The cysts were mainly situated in the cervical and thoracic regions, and the main symptoms in 18 patients were paralysis of limbs and bladder disturbance; fourteen patients had spinal defects—spina bifida occulta, hemivertebrae, spondylolisthesis and Arnold Chiari malformation. The histological variants of enterogenous cysts have been classified by Wilkins and Odom (1976) (Table 1).

TABLE 1. A histological classification of enterogenous cyst (modified after Wilkins and Odom 1976)

Type 1	Single pseudostratified cuboidal or columnar epithelium with or without cilia, lying on a basement membrane
Type 2	As in Type 1 with the addition of mucous glands, serous glands, smooth muscle, fat, cartilage, bone, elastic fibres, lymphoid tissue, or nerve ganglion
Type 3	As in Type 2 with the addition of ependymal or glial tissue

The position and content of these cysts are of interest. In the 26 cases reviewed by Holmes *et al.*, (1978), 20 had histological confirmation and, of these, the cyst was posterior to the spinal cord in 14 cases (70%). The content of the cyst in 12 of these cases, irrespective of the spinal level, was of a mucoid viscous nature; two cases (Raja, Subrahmanian and Prabhaker, 1972) had clear colourless fluid.

Histologically, in almost all the cases quoted by Holmes *et al.* (1978) and Palma and Lorenzo (1976) (allowing for duplicated references), posteriorly positioned cysts commonly had additional mesodermal elements such as cartilage, lymphoid tissue, smooth muscle, and neuroectodermal derivations such as nerve cells and nerve fibres. It is precisely because of this mixture of tissues that some authors have considered these cysts to be teratomatous in origin. On the other hand those cervical or cervico-dorsal cysts lying anterior or antero-lateral to the spinal cord invariably had a single layer of low cuboidal to columnar epithelium, ciliated or non-ciliated, with clear colourless watery cystic content, and were more often unassociated with skeletal abnormalities. Thus it would seem that there are two types of cyst: those that are anteriorly placed and are probably of

developmental origin, and those that are posteriorly placed and are probably of teratomatous origin.

Of the clinical features Holmes *et al.* (1978) found that pain and paralysis of limbs were the major symptoms. A unilateral deficit, as occurred in the patient described in this paper, is however very uncommon in children. Kubie and Fulton (1928) described a 27-year-old female with recurrent hemi- or quadriparesis; Palma and Lorenzo (1976) reported a 43-year-old male who presented with cervical pain and a right Brown-Sequard syndrome. Herskowitz *et al.* (1978) described a 28-year-old male with cervical pain and left hemiparesis at the age of 9 years who became quadriparetic at the age of 22, 25, and 26 years, each episode being associated with a recurrence of his cyst which required exploration on each occasion. This pattern of events is most unusual; only one of the 26 cases reported by Holmes *et al* (1978) recurred.

Acknowledgment

We are grateful to Dr Peter Buxton for his neuropathological advice.

References

HERSKOWITZ, J., BIELAWSKI, A., VENNA, N. & SABIN, T.D. (1978) Anterior cervical arachnoid cyst simulating syringomyelia. *Archives of Neurology*, **35**, 57.

HOLMES, G.L., TRADER, S. & IGNATIADIS, P. (1978) Intraspinal enterogenous cyst. *American Journal of Diseases of Children*, **132**, 906.

KUBIE, L.S. & FULTON, J.F. (1928) A clinical and pathological study of two teratomatous cysts of the spinal cord, containing mucous and ciliated cells. *Surgery, Gynecology and Obstetrics*, **47**, 297.

PALMA, L. & LORENZO, N.D. (1976) Spinal endodermal cysts without associated vertebral or other congenital abnormalities. *Acta neurochirurgica*, **33**, 283.

RAJA, R.D., SUBRAHMANIAN, M.V. & PRABHAKER, V. (1972) Neurenteric cyst (a case report) *Neurology (Bombay)*, **20**, 221.

WILKINS, R.H. & ODOM, G.L. (1976) Spinal intradural cysts. In: *Handbook of Clinical Neurology* Volume 20, part 2, pp. 55–102. North Holland Publishing Co., Amsterdam.