Online Case Report

Spinal neurenteric cyst presenting as recurrent midline sebaceous cysts

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A 59-year-old patient had been treated by the general surgery department at her local hospital for a recurrent sebaceous cyst on the back of her neck. Cervical MRI showed a cystic structure, posterior to fused vertebral bodies, connected to the skin by a sinus passing through the middle of a split cord malformation. The lesion was excised via an anterior approach. Histology confirmed a benign neurenteric cyst. This rare developmental anomaly, mimicking a common acquired surgical condition, highlights the need for caution in the diagnosis of midline pathology.

Key words: Neurenteric cyst – Split cord malformation – Vertebral malformation – Dermoid sinus

A 59-year-old, right-handed auxiliary nurse had presented to the general surgeons in her hospital complaining of recurrent abscesses at the base of her neck. She had undergone incision and drainage on a number of occasions, and had finally had a formal debridement.

Acute episodes had been heralded by pain in the neck, radiating to the left side of her face, which resolved on decompression of the abscess. She denied upper or lower limb neurological symptoms, nor any swallowing or speech problems. Systemic and neurological examinations were unremarkable other than for a transverse scar over C5 posteriorly, from her debridement.

MRI (Figs 1 and 3) revealed a track from the symptomatic area, through the subcutaneous tissues and muscles, into the spinal canal. Here it passed through a split cord malformation to join with a cystic lesion lying behind the fused bodies of C3 to C5. The track was seen to pass through the body of C3, possibly reaching and communicating with the oesophagus. Barium swallow, however, showed no abnormality in the pharynx or oesophagus, with no evidence of a fistulous tract;

hypopharyngoscopy and upper oesophagoscopy were also normal.

The patient initially declined intervention; however, at 6-month review, she reported increasing symptoms of left leg spasm with weakness of the forefoot, and requested surgery.

Excision was performed via an anterior right oblique cervical incision. A 14-mm hole was drilled through the fused segment at C3/4, with a diamond burr. There was a defect in the posterior longitudinal ligament and the remnant was reflected away, revealing a thin-walled grey cyst. Opening this exposed another cyst containing soft yellow keratinous material. This cyst wall had the appearance of arachnoid, and CSF was obtained. A further, thicker walled cyst was then revealed superiorly, which was divided from its attachments at the posterior surface of the split cord. The dural defect was closed and the bony defect filled with an iliac crest graft. In case a posterior decompression was required to excise the remaining dorsal tract as a further procedure, the affected spinal segment was then anteriorly plated.

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Figure 1 (Sagittal view) T2 weighted MRI cervical spine – pre-operative.

Figure 2 (Sagittal view) T2 weighted MRI cervical spine – 6 months post surgery.

Histological examination of the excised tissue showed pseudostratified ciliated columnar epithelium, similar to respiratory tissue, confirming the diagnosis of neurenteric cyst.

Postoperatively, she experienced mild left hemiparesis, which resolved over a few days, and a chest infection that was treated with antibiotics. She was discharged from hospital 11 days after surgery. At 6-month review (Fig. 2), she had only a mild weakness, unchanged from before her surgery.

Discussion

Puusepp¹ first reported an intraspinal endodermal cyst in 1934, calling it 'intestinoma'. Since then, nomenclature has varied widely, with the term 'neurenteric cyst' being coined by Holcomb and Matson² in 1954.

Wilkins and Odom³ classified neurenteric cysts into three types. Type A have a ciliated or non-ciliated cuboidal or columnar epithelium, lying on a basement membrane – similar to respiratory or GI epithelium. Type B contain in addition: mucous or serous glands, cartilage, bone, smooth muscle, elastic fibres or nerve ganglion. Type C have ependymal or glial tissue within them, as opposed to surrounding them.

Pang *et al.*⁴ postulated a theory for embryogenesis of split cord malformations (SCM), which explains the origin of neurenteric cysts. There is delayed separation of the

Figure 3 (Axial view) T2 weighted MRI at C5 level. Note SCM, anterior cyst and posterior track.

Figure 4 (Antero-posterior view) CT reconstruction showing corticated tract (arrowed) through the spinous process of C4.

ectoderm and endoderm in the third week of gestation, due to the formation of an accessory canal between the yolk sac and the amniotic cavity, through the embryonic disc. This persistent canal leads to division of the notochord, and thus development of a divided spinal cord – in our case, SCM Type II, with the hemicords contained in one dural sheath, separated by a thin fibrous sagittal septum.

The degree to which the accessory neurenteric canal differentiates or closes gives rise to the associated malformations. These range from the severe, where the GI tract passes through the vertebral body, the cord and vertebral arches and then via a sinus to the skin surface, to the mild and occult where the alimentary canal and spinal canal are closed with simply a cleft in the vertebral body. Our case lies somewhere between the two, with no open connection to the gut, but a tract from the fused vertebral bodies, via a cyst (representing a closed section of the tract) through the cord and on to the surface through the posterior spinous process (a Kovalesky canal; Fig. 4).

Neurenteric cysts usually occur in the cervicothoracic region^{5,6} or at the craniocervical junction.⁷ Their location determines the presenting symptoms. Paleologos *et al.*⁸ reviewed 80 cases from the literature, finding presenting

symptoms of quadri/paraparesis (83.8%), sensory dysfunction (74.2%), hyperreflexia (78.1%), pain (72.2%) and sphincter dysfunction (50.1%). Presentation as a sebaceous cyst, as in our case, has not previously been reported to our knowledge.

The majority of neurenteric cysts have been excised via a posterior approach, to protect the cord and to avoid compromising its blood supply.⁸ To reach the ventral location of our cyst from the back would have meant considerable manipulation of the cord; hence an anterior approach was used. This method has been advocated with complete excision of the cyst, to avoid recurrence.⁹

Conclusions

Neurenteric cyst is a rare congenital abnormality that develops in early gestation. It has a variety of presentations, depending on size and location, with an associated spectrum of symptom severity. We present a case that mimicked a common surgical condition, and would advocate that structural developmental abnormality be considered when confronted in clinic with any persistent or unusual midline pathology.

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