CASE REPORT

Upper cervical cord enterogenous cyst mimicking transient ischaemic attacks

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Nikolaos Tzerakis, Nikolaos.Tzerakis@uhns.nhs.uk**SUMMARY**

A 76-year-old woman presenting with tetraparesis, left-sided hemisensory loss and occasional neck pain was urgently admitted to our department. A cervical spine MRI scan revealed a partially cystic lesion compressing the cord at the C₂₋₄ level. The lesion was surgically excised. The histopathological diagnosis was that of an enterogenous cyst. No postoperative complications were noted and the patient significantly recovered from the preoperative tetraparesis. Eleven months before surgery, a waxing and waning presentation of the myelopathic signs with normal neuroradiological findings on a non-contrast-enhanced head CT scan, had unfortunately led to the misdiagnosis of transient ischaemic attacks and ischaemic stroke which delayed the treatment of an essentially benign disease the total surgical excision of which not only is usually curative but also improves the preoperative signs and symptoms.

BACKGROUND

A sudden onset, transient or persistent haemiparesis or hemisensory loss is commonly observed in medical practice especially in older population of patients. However, in our case, a clinical scenario mimicking cerebrovascular disease was sustained by an upper cervical cord enterogenous cyst, a rare but benign disorder which, when recognised, can be surgically removed. A radical surgical approach is, when feasible, mandatory since partial removal of these cystic lesions is often associated with recurrence.

CASE PRESENTATION

A 76-year-old right-handed woman was admitted to our department with a 1 year history of slow onset and progressively worsening weakness of all four limbs (left side worse than the right), left-sided hemisensory loss and occasional episodes of cervical neck pain.

She used to live independently and walked with the aid of a stick.

Neurological examination revealed a left-sided hemiplegia (grade 0 MRC) and a right-sided hemiparesis (grade 4 MRC in the upper limb and grade 3 MRC in the lower limb), left-sided hemisensory loss below C₄, absent reflexes in the upper limbs and reduced in the lower limbs. Babinsky sign was bilaterally present. Cranial nerves, higher mental functions and sphincter functions were intact.

There was no history of head or neck traumatic injuries. Her medical history was that of diabetes mellitus type II, chronic obstructive pulmonary disease, peripheral neuropathy and iron deficiency

anaemia. Interestingly, 11 months before the patient required hospital care because of recurrence of sudden onset episodes of mild left-sided hemiparesis and hemisensory loss which had been diagnosed as transient ischaemic attacks and, when the neurological deficit became persistent, as ischaemic stroke. At that time, a head CT scan did not show any abnormal findings and treatment with antiplatelet agent was accordingly started. During the months following this diagnosis, however, despite the ongoing medical treatment, the patient continued in experiencing a step-wise worsening of the muscle weakness until she became hemiplegia on the left side and hemiparesis on the right side and therefore completely bedridden.

INVESTIGATIONS

A cervical spine MRI scan (figure 1) revealed an intradural, extramedullary and intramedullary lesion, non-contrast-enhancing T₁-isointense and T₂-hyperintense, partially cystic extending from C₂ to C₄ and displacing the cord to the right lateral aspect.

OUTCOME AND FOLLOW-UP

The patient underwent a cervical laminoplasty and total excision of the lesion and no complications related to the surgical procedure were observed. Postoperatively, an additional MRI scan (figure 2) confirmed the radical excision of the cyst following the surgical treatment. The patient then experienced a slow but constant improvement of muscle power in upper as well as lower limbs although her comorbidities, the prolonged period of immobilisation and

Figure 1 Preoperative T₁-weighted sagittal cervical spine MRI scan.

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Figure 2 Postoperative T₁-weighted sagittal cervical spine MRI scan.

hospital acquired pneumonia were slowing her recovery. An intense physiotherapy course and an adequate level of supportive care played also an important role in supporting the patient during the process of improving her mobilisation abilities.

DISCUSSION

Enterogenous cysts are rarely encountered lesions of the central nervous system, representing 0.7–1.3% of spinal cord tumours¹; they most commonly present in young adults population (second, third decades) with a predilection for the male sex (2:1 male-to-female ratio).^{2–4}

Topographically, about 90% of enterogenous cysts are located intradurally and extramedullary and only 10% have an extradural or intradural and intramedullary location.⁵

Enterogenous cysts were initially described (1928) as ‘teratomatous cysts’ by Kubie and Fulton⁶ or Intestinomas (1934) by Puusepp⁷; the terms neuroenteric (1954) and enterogenous (1958) were introduced by Holcomb⁸ and Harriman.⁹

Macroscopically enterogenous cysts are formed by an external membrane delimiting a collection whose overall appearance may significantly vary in terms of consistency and colour.

The histopathological features of the wall of the cyst as well as the presence or absence of gastrointestinal or respiratory structures/components (figures 3 and 4) are the hallmarks used in order to classify these lesions.¹⁰

Clinically, patients present with localised pain associated with fluctuating signs and symptoms of myelopathy or radiculopathy.

The volume of these cystic lesions in time may undergo slight variations in diameter and this may account (at least at an early stage of presentation) for the clinical instability of symptoms

Figure 3 Pseudostratified columnar epithelium.

Figure 4 Immunohistochemistry for cytokeratin (CK) 7 (staining for CK 7 and no staining for glial fibrillary acid protein, S100 and vimentin demonstrate that this is an enterogenous cyst rather than an ependymal cyst).

which may lead to misdiagnosis and delay in providing adequate treatment.¹¹

Usually surgical treatment is curative provided that the resection is total: radical excision of the cyst and its wall is of paramount importance since recurrence of enterogenous cysts is correlated with partial excision and no other known factors, such as age, sex or dimension of the cystic lesion, may predispose to future recurrence.^{12,13}

Learning points

- ▶ The clinical presentation of enterogenous cysts located in the spinal cord is usually associated with myelopathy or radiculopathy and the symptoms at an early clinical stage may be vague and therefore likely leading to misdiagnosis.
- ▶ Although enterogenous cysts are rare, they are essentially benign lesions and their total surgical resection is curative and often reverses the preoperative clinical signs and symptoms. Hence, early diagnosis is paramount to avoid development of established and irreversible neurological deficits.
- ▶ When the diagnosis of transient ischaemic attack is made on patients presenting with recurrent episodes of focal neurological deficit in addition to a plain head CT scan, further neuroradiological investigations should be performed in order to rule out a lesser common cause underlying the progression or recurrence of the neurological symptoms.

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Competing interests None.

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