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Syringomyelia and bilateral vocal fold palsy

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Abstract
This is a case of a 35-year-old who had complained of noisy breathing for 15 years, and had been on treatment for 'chronic asthma'. She presented to the Accident and Emergency department with an acute episode of difficulty with breathing, and on admission was found to have stridor and bilateral abductor vocal fold palsy. Further workup revealed a syringomyelia with an associated Chiari type 1 malformation. It is important to consider the above diagnosis in the differential of young adults with breathing difficulties. A discussion of syringomyelia, Chiai malformations and bilateral vocal fold palsy follows.

Key words: Syringomyelia; Arnold-Chiari Malformation; Larynx; Vocal Fold Paralysis; Apnoea, Sleep

Introduction

Olivier first used the term syringomyelia in 1824, and described an abnormal longitudinal cavity (syrinx) within the spinal cord.[1] This cavity is irregular and asymmetrical and often occupies the central part of the cord. Although filled with CSF, it may, or may not, have an obvious connection with the central canal. It can extend throughout the entire length of the cord, but has a predilection for the cervical segment. An extension rostrally into the medullary area is described as syringobulbia.

The association of syringomyelia and bilateral vocal fold palsy is uncommon but well recognized. This usually occurs with bulbar extension of the syrinx cavity or in the presence of a Chiari malformation. The latter is a relatively common cause of vocal fold palsy in children. In our review of the literature, we could find only five adult cases of bilateral vocal fold palsy in association with either the Chiari malformation and/or syringomyelia. However, vocal fold palsy is rarely the presenting complication of an adult with syringomyelia or a Chiari malformation.[2] We present a case where diagnosis of syringomyelia was made only after the patient's presentation with bilateral vocal fold palsy.

Case report

A 35-year-old Afro-Caribbean lady presented to the Accident and Emergency department with what was initially thought to be an acute exacerbation of asthma. She had apparently suffered chronic asthma for 15 years and used bronchodilators and steroids. On admission under the physicians, she was found to have severe oxygen desaturation during sleep, chronic carbon dioxide retention, and severe noisy breathing while sleeping. An urgent otolaryngological referral was made and the patient was noted to have inspiratory stridor, with bilateral vocal fold paralysis on nasendoscopy. Further otolaryngological examination was unremarkable.

The airway was secured by means of an emergency tracheotomy under general anaesthesia. Recovery from anaesthesia was prolonged, and her breathing erratic. This fuelled the initial suspicion that her problems might be central. A magnetic resonance image (MRI) of the brain and brainstem was requested, as well as a neurological opinion.

The change in her voice had been investigated some four years ago in a different hospital, and right vocal fold palsy noted. No apparent cause was found for this. Her voice subsequently improved. At the same time, complaints of severe snoring problems, violent early morning headaches, and severe daytime somnolence had led to a diagnosis of obstructive sleep apnoea. Further enquiry revealed that she had suffered with intermittent difficulties with swallowing, but this complaint had been so vague that it had not been investigated. For years, she had noted a tendency to frequently burn herself on the left side, often after she had fallen asleep beside the fireplace.

A neurological assessment revealed a thin lady with mild scoliosis, concave to the left. There was paresis of the soft palate, that was more marked on the right. She had nasal speech, that was evident even with the speaking valve of the tracheotomy tube. There was some intrinsic muscle wasting of the fingers, and a notable hyporeflexia of the left upper and lower limbs. Pinprick and temperature sensations were also diminished in the left upper limb and shoulder, but there was preservation of the vibratory and position sense.

The MRI showed a dilatation of the central canal, most marked opposite the C3-C7 vertebrae, and measuring 6 mm and 14 mm in its anterior-posterior (A-P) and transverse diameters respectively. There was also a cerebellar tonsilar descent of 7–8 mm through the foramen magnum (Figures 1–4). A computed tomography (CT) scan did not add any additional information. Routine blood tests were normal, and autoantibodies, antiganglioside antibodies, syphilis serology and ACE screens were all negative.

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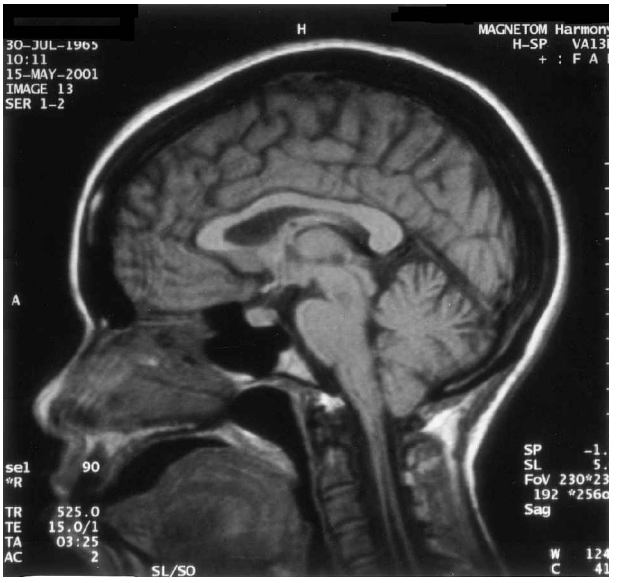

FIG. 1
MRI saggital scan showing cerebellar tonsilar descent.

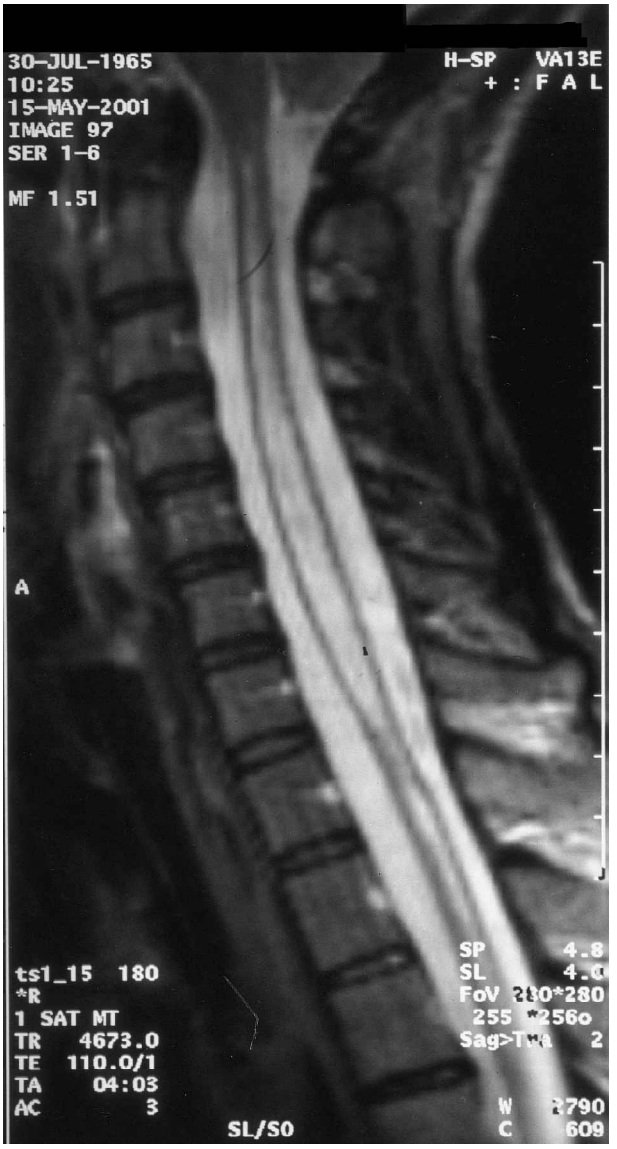

FIG. 2
MRI saggital scan of spinal cord showing syrinx cavity.

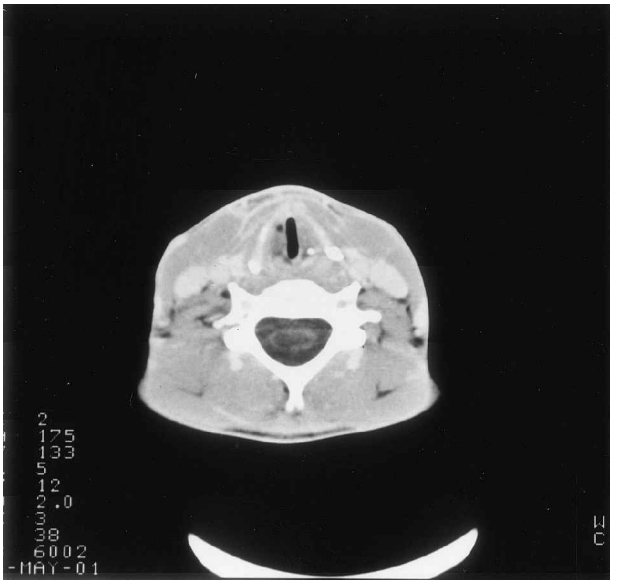

FIG. 3
CT scan showing both syrinx cavity within the spinal cord, and bilateral vocal fold palsy.

After the tracheotomy, she slept well with the complete resolution of her early morning headaches and daytime somnolence. Her swallowing was assessed on the ward by the speech and language therapists, and a mild degree of aspiration noted. A fine bore nasogastric tube was passed to help with her feeding. Her asthma medications were stopped as no asthmatic problems were noted on the ward.

She was transferred to the care of the neurosurgeons, who performed craniovertebral decompression after further assessment. Post-operative recovery was relatively trouble-free, but for some neurogenic pain in the lower spinal dermatomes, which improved with gabapentin. Although her feeding gastrostomy and a tracheotomy are still in place, she is being kept under review to see if her

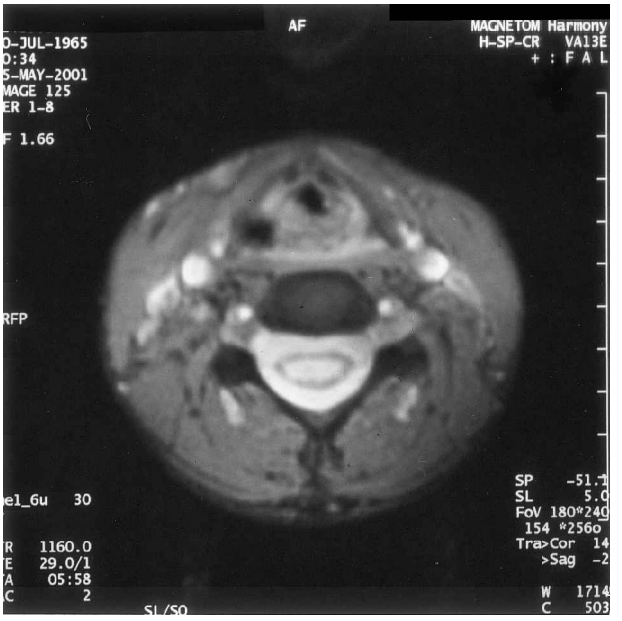

FIG. 4
MRI (transverse) showing syrinx cavity.

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swallowing and vocal fold palsies will improve enough to enable both decannulation and removal of the gastrostomy. So far, nasendoscopy performed two months after surgery has failed to show any changes in laryngeal function.

Discussion

Syringomyelia is a rare condition and in the UK, a prevalence of 8.4 cases per 100 000 population has been quoted.[3] The aetiology of syringomyelia is not very clear. Frequent assocation with spinal dysraphism had led to early suggestions that it might be caused by imperfect closure of the neural tube with persistent of embryonic cell rests and subsequently frequent liquefaction and cavity formation,[4] but this is not the current hypothesis. The frequent association of this condition with congenital anomalies and lesions around the foramen magnum has led to a hydrodynamic theory of causation, that is more widely accepted. There are a number of congenital malformations associated with this condition,[5] with the Chiari malformations as the commonest.[6] These are now thought to be responsible for the development of a syringomyelia, and other recognized causes include spinal tumours in the vicinty of the foramen magnum, trauma (spinal injury), and spinal arachnoiditis (often caused by subarachnoid haemorrhage or meningitis). Barnett found that a quarter of his syringomyelia patients previously had had a basal arachnoiditis.[6] These all tend to support the view that the aetiology of this condition might be related to mechanical obstruction of CSF flow at the level of the foramen magnum.

In 1883, Cleland reported two infant autopsies with brain stem and cerebellar herniation through an occipital cranial defect,[7] but it was Chiari (in 1896) that classified the hindbrain deformities into four categories[8] as shown in Table I. In 1894, Arnold[9] reported a case of cerebellar herniation with an associated spina bifida. This was the same as the Chiari type 2 malformations, hence the term Arnold-Chiari malformation. This term is often applied loosely, but should be used only to describe the type 2 malformations. In 1958, Gardner noted that there might be a causal relationship between these malformations and syringomyelia.[10] The Chiari type 1 malformation is the commonest congenital anomaly associated with syringomyelia,[6] and it is now generally accepted that abnormalities within the vicinity of the foramen magnum are responsible for the causation of the syringomyelia. These are thought to cause mechanical obstruction, thereby preventing egress of CSF from the fourth ventricle into the subarachnoid space, and thus leading to pressure waves of fluids being forced down into the central canal.[11] If elevated fluid pressure persists, CSF may be forced through the ependymal lining into the substance of the spinal cord.

TABLE I
TYPES OF CHIARI MALFORMATIONS

Type 1	Downward protrusion of cerebellar tonsils through the foramen magnum
Type 2	Protrusion of inferior cerebellar vermis, thin and elongated pons + associated spinal meningocele or meningomyelocele
Type 3	Descent of the entire cerebellum into the cervical spinal area + large bony occipital defect +/− hydrocephalus
Type 4	Cerebellar hypoplasia

As the spinothalamic fibres decussate just anterior to the central canal before relaying in the tract, it is quite predictable that these fibres would often be the first affected by an expanding syrinx cavity. This gives rise to the dissociated sensory loss often seen over the shoulder and upper limbs. Further extension laterally leads to the involvement of the anterior horn cells, with loss of reflexes, and wasting of muscles. These constitute the commonest findings in syringomyelia.

The extension of syrinx into the medulla may affect the nucleus ambiguus, that is motor to the pharynx and larynx. Pressure at this point is responsible for the vocal fold palsy and dysphagia, sometimes seen in this disorder. Medullary involvement may also result in nystagmus and diplopia, as well as palatal paralysis. There may be a centrally mediated respiratory disorder leading to a central sleep apnoea, and this could further complicate an obstructive picture, possibly from vocal fold palsy.[12–14] The central apnoea is explained by pressure on the respiratory centre, or insensitivity of the peripheral chemoreceptors due to glossopharyngeal nerve dysfunction.[15]

An asssociated Chiari malformation may also cause pressure on the medulla, giving rise to similar findings. Dysfunction of the 10th nerve may occur as a result of direct mechanical injury because of the elongated reach between the brain stem nerve origin and the jugular foramen.[2] The more severe the Chiari malformation, the more likely these are to be found. However, it is very seldom that the Chiari type 1 malformation causes vocal fold palsy, as these are seen most commonly with the type 2 and 3.[14]

The more severe malformations are often diagnosed at birth because of the associated spinal dysraphism. However, with the Chiari type 1 malformation, the presenting symptoms can remain vague for so many years that doctors and the patients could easily dismiss them. The average age of diagnosis is the early 40s with a slight female preponderance.[16] A high index of suspicion is required in these patients, and some of the neurological signs can easily be missed without a detailed examination. The exacerbation of symptoms and possible extension of a syrinx cavity can often be related to different variations of the Valsalva manoeuvre that lead to an increase in intracranial/intraspinal pressure, such as coughing, sneezing, exertion, anaesthesia, etc.[17]

The incidence of vocal fold paresis in adults with the Chiari type 1 malformation has been put at 15 per cent.[19] In a series of 24 patients, all cases of bilateral vocal fold paralysis were secondary to type 2 anomalies and none to the type 1 anomaly.[14] In infants, the Chiari malformations are a relatively common cause of vocal fold palsy, but even these are usually in association with a meningomyelocele (type 2 anomaly). The vocal fold palsy in these cases often follows an attempt to repair the associated myelomeningocele.[19] In our patient, the supposed wheeze that led to the initial diagnosis of asthma is perhaps an indication of how subtle the presentation of this disorder can be. It illustrates how difficult it can be sometimes to tell upper airway obstructive noises from that caused by the lower airway. The ENT doctor must be especially wary of patients who have been labelled as asthmatic.

The MRI is the imaging method of choice in identifying a brainstem cause for syringomyelia.[20] It delineates the anatomy of the region very clearly, demonstrates any syrinx cavity as well as any associated anomalies. The high CT bony artefact in areas such as the cranio-cervical junction and posterior fossa, make it less desirable for imaging this area.

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Treatment is directed towards relieving the immediate life threat, such as an emergency tracheotomy for a compromised airway. In the absence of brainstem and cranial nerve dysfunction, a policy of observing the syrinx cavity might suffice, assuming a state of hydrodynamic equilibrium. However, early surgical decompression is the mainstay of treatment, and aims to stem the relentless progress of this disease. Surgical techniques include posterior fossa bony decompression, cervical laminectomies and duraplasty.[21] In addition, shunting of the ventricles has been advocated and used with some success, especially in the presence of hydrocephalus.[22]

This condition is progressive if untreated, although progress is frequently slow, and a prolonged arrest lasting for many years may occur.[23]

The prognosis depends upon the extent of neurological damage already sustained prior to intervention. Vocal fold improvement may not occur despite decompressive surgery. Loss of motor cells will suggest a permanent lesion, while axonal damage of a nerve may retain the capacity for regeneration. Levy[16] reported that 50 per cent of his patients stabilized after intervention, 30 per cent showed neurological improvement, while 20 per cent kept on deteriorating. Apart from the life-saving nature of a tracheotomy, it was found that our patient's symptoms of violent early morning headaches and daytime somnolence improved markedly after the procedure. This may suggest the contributory role of hypercarbia to the patient's symptoms of headache and hypersomnolence, or the increase in intracranial pressure associated with an obstructed upper airway. The optimal management of these patients remains best under a multidisciplinary team.

Conclusions

This case highlights the fact that the otolaryngologist may well be the first clinician to encounter this disorder. Therefore a high index of suspicion as well as a fair understanding of the pathological process is required. The subtle nature of signs and symptoms can result in delayed diagnosis. We are also reminded of the old truth 'all that wheezes is not asthma'. In young adults presenting with vocal fold palsy, dysphagia, aspiration pneumonia, or sleep apnoea, syringomyelia and the Chiari malformations must be considered in the differential diagnosis. The MRI is an indispensable diagnostic tool in this situation.

References

1 Willis WH, Weaver DF. Syringomyelia with bilateral vocal cord paralysis. *Arch Otolaryngol* 1968;**87**:468–70
2 Belvins NH, Deschler DG, Kingdom TT, Lee KC. Chiari 1 malformation presenting as vocal cord paralysis in the adult. *Otolaryngol Head Neck Surg* 1997;**117**:S191–4
3 Brewis M, Poskanzer DC, Rolland C, Miller H. Neurological diseases in an English City. *Acta Neurol Scand* 1966;**42**(suppl 24):1–89
4 Sahu RN, Satapathy PC. Syringobulbia. *Indian Med J* 1962;**39**:146–7
5 Mcilroy WJ, Richardson JD. Syringomyelia: A clinical review of 75 cases. *J Canad Med Assoc* 1965;**93**:73–134
6 McLellan DL. Syringomyelia. In: Walton, J., ed. *Brain Diseases of the Nervous System*. Oxford: Oxford University Press, 1993;**14**:506
7 Cleland J. Contribution to the study of spina bifida, encephalocele and anencephalus. *J Anat Physiol* 1883;**17**:257–91
8 Chiari H. Ueber Veranderungen des Kleinhirns, des Pons and der Medulla oblongata infolge Von Congenitaler Hydrocephalie des Grossshirns. *Denschr Acad Wiss Wien* 1896;**63**:71–116
9 Arnold J. Myelocyste, transposition Von Gewebskeimen and Sumpodie. *Beitr Path Anat* 1894;**16**:1–28
10 Gardner WJ, Angel J. The cause of syringomyelia and its surgical treatment. *Clevel Clin Quart* 1958;**25**:4
11 Williams B. On the pathogenesis of syringomyelia: A review. *Proc R Soc Med* 1980;**73**:798
12 Zager EL, Ojemann RG, Poletti CE. Acute presentations of syringomyelia: report of three cases. *J Neurosurg* 1990;**72**:133–8
13 Kelly DB. Bilateral abductor laryngeal palsy in syringomyelia. *Br Med J* 1947;**2**:655–6
14 Choi SS, Tran LP, Zalzel GH. Airway abnormalities in patients with Arnold-Chiari malformation. *Otolaryngol Head Neck Surg* 1999;**121**:720–4
15 Bodinski GE, Hudson LD, Well JV. Impaired peripheral chemosensitivity and acute respiratory failure in Arnold Chiari malformation and syringomyelia. *N Engl J Med* 1973;**288**:947
16 Levy WJ, Mason L, Hahn JF. Chiari malformation presenting in adults: a surgical experience of 127 cases. *Neurosurgery* 1983;**12**:377–90
17 Ruff ME, Oakes WJ, Fisher SR, Spock A. Sleep apnoea and vocal cord paralysis secondary to type 1 Chiari malformation. *Pediatrics* 1987;**80**:231–4
18 Dobkin BH. The adult Chiari malformation. *Bull Los Angeles Neurol Soc* 1977;**42**:23
19 Emery PJ, Fearon B. Vocal cord palsy in paediatric practice: a review of 71 cases. *Int J Pediatr Otorhinolaryngol 1984;***8**:147–54
20 Dyste GN, Menzes AH. Presentation and management of paediatric Chiari malformation without myelodysplasia. *Neurosurgery* 1988;**23**:589–97
21 Paul KS, Lye RH, Strang FA, Dutton J. Arnold-Chiai malformation: review of 71 cases. *J Neurosurg* 1983;**58**:183–7
22 Dyste GN, Menezes AH, Van Gilder JC. Symptomatic Chiari malformations: an analysis of presentation, management and long-term outcome. *J Neurosurg* 1989;**71**:159–68
23 Logue V, Edwards MR. Syringomyelia and its surgical treatment: an analysis of 75 patients. *J Neurol Neurosurg Psychiatr* 1981;**44**:273

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