Idiopathic Syringomyelia

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12.1 Introduction

Although no precise definition of idiopathic syringomyelia exists, the general consensus is that the term applies to a syrinx not associated with Chiari I malformation, spinal cord tumour, post-traumatic or infectious adhesive arachnoiditis (Bogdanov et al. 2004; Mauer et al. 2008; Nakamura et al. 2009; Roser et al. 2010; Struck and Haughton 2009) or other obstruction to cerebrospinal flow (CSF) detected by imaging studies. Given the high prevalence of MRI in the routine evaluation of back and neck pain, incidental discovery of idiopathic syringomyelia is becoming more common (Roser et al. 2010; Nakamura et al. 2009). Although syringomyelia can be associated with a clinical centromedullary syndrome, with predominantly sensory symptoms such as pain and temperature insensitivity, in many cases it is an incidental finding. We present here a broad overview of the topic referencing much of the work from our recent review on the subject (Roy et al. 2011).

The underlying pathophysiology of idiopathic syringomyelia is unclear and probably multifactorial. Few studies directly address this topic, although it has been hypothesised that the underlying filling mechanisms for idiopathic syringomyelia may be similar to those which operate with Chiari malformation type 1 (Bogdanov et al. 2004; Heiss et al. 2012). A study comparing patients with idiopathic syringomyelia, people with Chiari malformation type 1 and controls found similar morphometric abnormalities in

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both the idiopathic syringomyelia and the Chiari type one patients (Bogdanov et al. 2004). This included similar shortening of the posterior fossa and a reduction in ventral CSF space, although the dorsal CSF space was significantly smaller in Chiari type 1 patients. Syrinx diameter was also found to be significantly larger in Chiari malformation type 1 patients (5.5 ± 4.7 mm versus 2.7 ± 1.9 mm). This study posits that a posterior fossa with decreased compliance promotes the development of pulsatile CSF subarachnoid pressure waves aiding in the development of syringomyelia. Abnormal CSF flow velocities have been reported in idiopathic syringomyelia patients, raising the possibility of similarities in flow patterns at the foramen magnum in both Chiari type 1 and idiopathic syringomyelia patients (Struck and Haughton 2009). The idea of similar morphometric abnormalities in idiopathic syringomyelia and Chiari malformation type 1 seems to follow the reasoning by Klekamp (2002), who questions if a true idiopathic syrinx actually exists. A report on two patients with idiopathic syringomyelia postulated that subtle micro trauma may have been contributory (Porensky et al. 2007). In summary, the available evidence strongly suggests that a thorough anatomical investigation is required before labelling a patient as truly idiopathic.

12.2 Imaging and Classification

By the very meaning of the term, idiopathic syringomyelia does not have an underlying anatomical abnormality readily seen on imaging. With the benefit of modern imaging, however, we can, more often than not, find a structural abnormality. Increased peak CSF flow velocities have been reported in idiopathic syringomyelia (Struck and Haughton 2009). A small posterior fossa and narrow CSF spaces have been reported in idiopathic syringomyelia (Bogdanov et al. 2004). A series of four patients with idiopathic syringomyelia were all documented as having a 'tight cisterna magna', referring to the cisterna magna being impacted by the tonsils (Kyoshima et al. 2002). In a study looking at 10 patients, with apparently idiopathic syringomyelia, subsequent CT myelography revealed obstruction to CSF flow in nine of them (Mallucci et al. 1997). Those patients who subsequently underwent laminectomy and release of arachnoid adhesions or webs fared much better than those who were treated with direct shunting of the syrinx. Another reported patient had a T2–C5 syrinx, together with a spinal intradural arachnoid cyst extending between T6 and T3, which was revealed by aqueous myelography. After laminectomy and collapse of the cyst, the patient improved (Clifton et al. 1987). A study looking at 125 patients with idiopathic syringomyelia, using cardiac-gated, phase-contrast CSF flow studies, found blockage of flow in 33 patients (Mauer et al. 2008). The most common level of blockage was T6. In 8 of these 33 patients, the evidence of CSF flow blockage was unequivocal, and yet CT myelography revealed a blockage in only 2 of these cases. The authors concluded that conventional myelography is not a useful tool in the diagnosis of idiopathic syringomyelia and that cardiac-gated CSF flow studies should suffice.

Idiopathic syringomyelia may be subdivided into localised versus extended cavities, with localised being described as being under three vertebrae in length and extended as stretching behind four or more vertebrae. The localised variant, which may simply represent an enlargement of the central canal, usually has milder symptoms and can be treated conservatively (Nakamura et al. 2009). Numerous other classification systems delineate terms such as hydromyelia, simple hydromyelia, syringomyelia and syringohydromyelia (see Chap. 20) (Kyoshima et al. 2002), but it is unclear whether these terms aid in the diagnostic or management process.

Variants of syringomyelia referred to as dilated or persisting central canals, or hydromyelia, are based on the concept that the central canal normally involutes with aging and is often not easily seen on imaging. Holly and Batzdorf (2002) looked at 32 patients with slitlike syrinx cavities, which they referred to as asymptomatic persistent central canal. Their study found symmetrically enlarged, central-placed cavities within the spinal cord, with a mean diameter of 2 mm (range 1–5 mm) and with no enhancement seen after intravenous gadolinium. Interestingly,

however, 10 of these 32 patients did give a history of trauma, even though the study did not classify them as being post-traumatic. Half of the patients in this study also had alternate diagnostic explanations for their symptoms. The authors therefore argued that that these slitlike syrinx cavities do not represent true syringomyelia and are possibly even different from a presyrinx-like state. Yet, in the literature, there is no uniformity regarding this opinion. For example, Roser et al. (2010) differentiated hydromyelia, which they refer to as simply a dilated central canal, from idiopathic syringomyelia which, they point out, is accompanied by different clinical and radiological signs. In particular, they portend that patients with hydromyelia (dilated central canal) have no neurological deficits and mainly present with pain, which could be radicular, burning or musculoskeletal; a constellation of symptoms is similar to those described by Holly and Batzdorf. They further suggest that hydromyelia (dilated central canal) is a congenital condition which, in the setting of trauma, could develop into syringomyelia. It is difficult to determine, based on these studies, if slitlike syringes, hydromyelia (dilated central canal) and idiopathic syringomyelia are truly different entities or simply a continuum on a spectrum.

A wide variety of syringes is also seen in association with spinal cord pathology, including various inflammatory conditions or lesions compressing the subarachnoid space (Heiss et al. 2012). The specific pathologies are varied in nature and include delayed reactions to meningitis, extramedullary tumours, osteophytes and herniated intervertebral discs.

12.3 Presentation

With noncommunicating syringomyelia, symptoms can include spastic weakness of the lower extremities, paraesthesias, dysaesthesias and segmental sensory loss (Milhorat et al. 1995a, b). The pace of neurological deterioration in syringomyelia may be rapid initially but slows down after neurological signs become well established (Bogdanov and Mendelevich 2002). In Chiari 0 patients with syringomyelia, presenting symptoms include scoliosis, headaches and neck, back or leg pain (Chern et al. 2011). With idiopathic syringomyelia, the most common presenting symptom is pain, followed by paraesthesias, numbness and unnoticed hand injuries, but it is also not uncommon for patients to present with long tract signs (Mallucci et al. 1997). Although syringomyelia symptoms are classically described as pain and temperature insensitivity in a cape-like distribution, few of our own patients and indeed few of the studies reviewed actually demonstrated this (Roy et al. 2011). Although we did find an overall increased syrinx diameter in the two operated cases, the relevance is unclear given the few cases we have. In a population of 48 children with idiopathic syringomyelia, symptoms that led to the diagnosis of this condition fell into five groups: scoliosis, cutaneous marker/developmental anomaly,[1] pain, neurological findings and screening/incidental findings (Magge et al. 2011).

With idiopathic syringomyelia, it is difficult to correlate the presenting symptoms with the location of the syrinx (Roy et al. 2011). Specifically, regarding the issue of pain, there does not appear to be any correlation between syrinx size or location and the pain (Magge et al. 2011). Even when we can attempt to localise the symptoms, patients may actually have comorbid symptoms, from degenerative changes of the spine and musculoskeletal complaints, which require separate medical evaluation. Thus, it is important to stress that idiopathic syringomyelia is commonly an incidental, asymptomatic finding. Non-specific symptoms, such as pain, may result from other causes and be coincidental to the finding of the syrinx.

12.4 Management

Table 12.1 provides a review of the key literature regarding idiopathic syringomyelia. Only some of the details for each study are included, and some of the patients, in some of the series, were eventually found to have other structural

[1] The original paper does not clarify further the meaning of these terms.

Table 12.1 Summary of papers reporting cases of idiopathic syringomyelia

Authors and year	Type	Number of patients (sex)	Age (mean)	Location	Symptoms	Treatment	Outcomes	Follow-upperiod
Ataizi et al. (2007)	Case report	1 (F)	28	C5–T1	Neck and back pain	Conservative (patient refused surgery)	Resolution of pain Spontaneous collapse of syrinx	16 months
Bogdanov et al. (2004) (*Note 1*)	Cross-sectional	17 (2 F, 15 M)	49	Cervical	Segmental sensory loss Pyramidal signs Muscle atrophy	Not applicable	Not applicable	Not applicable
Chen et al. (2004)	Case report	1 (F)	19	C2–C6	Proximal upper limb weakness Diminished pain and temperature sensation	Suboccipital craniectomy + C1 and C3–C5 laminectomies	Improved strength Sensory deficit unchanged Syrinx reduced	12 months
Chern et al. (2011)	Retrospective case series	15 (6 F,9 M)	11	Multiple	Scoliosis Headache Neck pain	Suboccipital craniectomy + C1 laminectomy	Resolved – 4 Improved – 6 Stable – 3 Worse – 1	12–75 months
Holly and Batzdorf (2002) (*Note 2*)	Prospective study	32 (14 F, 18 M)	40	Cervical (16 cases) Thoracic (12 cases) Cervical-thoracic (4 cases)	Mechanical spinal pain Radicular pain Numbness	Anterior fusion C6–C7 (1 patient) Conservative (31patients)	Improved – 6 Unchanged – 19 Worse – 7	6–110 months (mean 38)
Jinkins and Sener (1999)	Case series	3 (2 F, 1 M)	27	Lumbar Cervical Thoracic	Low back pain Headache	Conservative	Stable with resolution of pain in 2 patients	2–4 years (mean 3)
Kastrup et al. (2001)	Case report	1 (F)	61	C1–conus	Burning pain	Carbamazepine	Subsequent collapse of syrinx Symptoms unchanged	8 years
Kyoshima et al. (2002) (*Note 3*)	Retrospective case series	4 (3 F, 1 M)	38	Whole or near whole cord	Impaired touch and pain sensation Weakness Hypoalgesia	Craniocervical decompression	Improved symptoms in all Syrinx decreased in all but 1 case	2.5–11years (mean 8)
Lin et al. (2006)	Case report	1 (M)	35	T2–T9	Leg weakness Reduced touch and pinprick	T6–T8 laminectomy + shunt (syringosubarachnoid)	JOA score [a] improved from 10 to 14 at day 30 post-op	30 days

Magge et al. (2011)	Retrospective case series	48 (30 F,18 M)	10	2–17 levels; mostly thoracic	Scoliosis, Cutaneous stigmata Leg or back pain (neurological symptoms were judged to be incidental)	Shunt (syringosubarachnoid) (1 patient) Fenestration of syrinx (1 patient) Conservative (remainder)	Operated cases: weakness and worsened gait (1st case) No change (2nd case)	Clinical: 3–56 months (mean 15.5) Radiographic: 2–64 months (mean 23.8)
Mallucci et al. (1997)	Retrospective case series	10 (2 F, 8 M)	48	Not described	Sensory disturbance weakness	Laminectomy and excision of web/cyst, Shunt (syringosubarachnoid) (2 patients)	Improved symptoms and syrinx reduced except for the 2 shunted cases	Not described
Mauer et al. (2008) (*Note 1*)	Prospective case series	125 (76 F,49 M)	36	1–18 levels not clearly defined	No surgery: Pain + sensory impairment With surgery: Bowel/ bladder dysfunction, gait problems Paralysis	Arachnoid scar or web resection (10 patients) Conservative (115 patients)	Surgery: 4 improved, rest stabilised Conservative: outcomes not described	Not described
Nakamura et al. (2009)	Retrospective case series	15 (4 F, 11 M)	45	Localised C3–T2 (12 cases) Extended C1–T8 (3 cases)	Upper limb numbness Neck pain, 3pts extended: also progressive upper limb weakness	Conservative (12 patients) Shunt (syringosubarachnoid) (3 patients)	Conservative: no changes Surgical cases: reduced syrinx and mean JOA [a] decreased	7–20 years (mean 10)
Porensky et al. (2007)	Case report	2 (M)	43y and 44y	T1–T2 and C5–T5	Ataxia Neck pain Progressive left leg paresis, pinprick deficit on right leg	Laminectomy, lysis of adhesions + duraplasty	Reduced syrinx and asymptomatic (1st case) Refilled syrinx and unchanged neurology (2nd case)	1 year and 7 months
Roser et al. (2010)	Prospective case series	40 (25 F, 15 M)	37	Cervical (23 %) Thoracic (51 %) Cervical-thoracic (25 %)	Pain or dysaesthesias in limbs	Conservative	No radiological changes Neurologically stable	6–93 months (mean 36.9)
Struck and Haughton (2009) (*Note 1*)	Retrospective case series	8 (4 F,4 M)	12	Lower cervical or thoracic	Scoliosis Headache Back pain Extremity numbness, family history Nausea and vomiting	Posterior fossa decompression (4 patients) Conservative (4 patients)	Surgical cases: decreased symptoms in all Reduction of syrinx in 1 case	Not described

Notes: (1) Imaging studies. (2) 10 patients had a history of trauma. (3) Technically not idiopathic since a tight cisterna magna is a definite lesion (Roy et al. 2011)
M male, *F* female
[a]*Japanese Orthopaedic Association spondylotic myelopathy score*

anomalies. A large number of treatment options are discussed, including posterior fossa and foramen magnum decompression, laminectomy, lysis of adhesions, syrinx fenestration and syringosubarachnoid, syringoperitoneal and syringopleural shunting. Recent studies have emphasised the importance of improving CSF flow dynamics, regardless of the treatment strategy employed (Bogdanov et al. 2004; Kyoshima et al. 2002; Lee et al. 2002; Nakamura et al. 2009).

Surgical treatment should be reserved for clearly symptomatic patients, with progression on serial clinical and radiological examination. The majority of the patients in our own series were conservatively managed. We caution against the automatic tying of symptoms to the syrinx and proceeding with surgical management, since the symptoms may be purely coincidental, as mentioned above. A nonoperative approach is perfectly justifiable when a patient is either asymptomatic or experiences relatively mild symptoms. Surgical management may not offer much utility, with cases either worsening or showing no changes clinically (Magge et al. 2011). The frequency of follow-up MRI may be dictated by symptomatology.

Historically, shunting strategies have been employed for idiopathic syringomyelia and have led to clinical and radiological improvement. However, some studies have reported a variety of complications, including shunt failure, syrinx relapse, catheter tip migration and comorbidities from mechanical damage to cord tissue (Batzdorf et al. 1998; Sgouros and Williams 1995). Shunts may not be an effective solution in preventing the progress of syringomyelia, given the subsequent gliosis that can follow within the cord (Mallucci et al. 1997). We believe that shunting should be used as a measure of last resort, when no aetiology is evident after repeated imaging studies and surgical exploration does not reveal any pathology around the site of the syrinx.

In the case where the aetiology is clearly evident, such as a tight cisterna magna or small posterior fossa, craniocervical decompression is the best option for restoring CSF flow dynamics. An overly wide suboccipital craniectomy can, however, lead to cerebellar ptosis (Holly and Batzdorf 2002). The key to successful management is a wider opening to the foramen magnum but not the posterior fossa (Kyoshima et al. 2002). If CSF flow obstruction originates from spinal subarachnoid pathology such as a cyst, web or scar, treatment should be targeted towards decompressing the spinal subarachnoid space and reconstituting flow. This is illustrated by the two patients in our series that underwent surgical treatment. There is no clear evidence on precisely what such decompression and reconstruction should include. Laminectomy followed by scar, web or cyst resection has been commonly employed in other studies (Clifton et al. 1987; Lee et al. 2002; Mallucci et al. 1997) and has been used with good results in our published series (Roy et al. 2011). In the event of no evidence of any anatomical abnormality, in the setting of progressive neurological dysfunction attributable to a syrinx, surgical exploration is a reasonable option with a shunt as the last resort.

We have combined evidence on idiopathic syringomyelia and our experience into an algorithm (Fig. 12.1), to assist in the decision-making process. In most cases of incidental, asymptomatic findings, clinical review and imaging as indicated should be sufficient. For the more challenging symptomatic cases, the key focus is to resolve CSF flow problems since most of the evidence points to disrupted flow dynamics, leading to the development of syringomyelia.

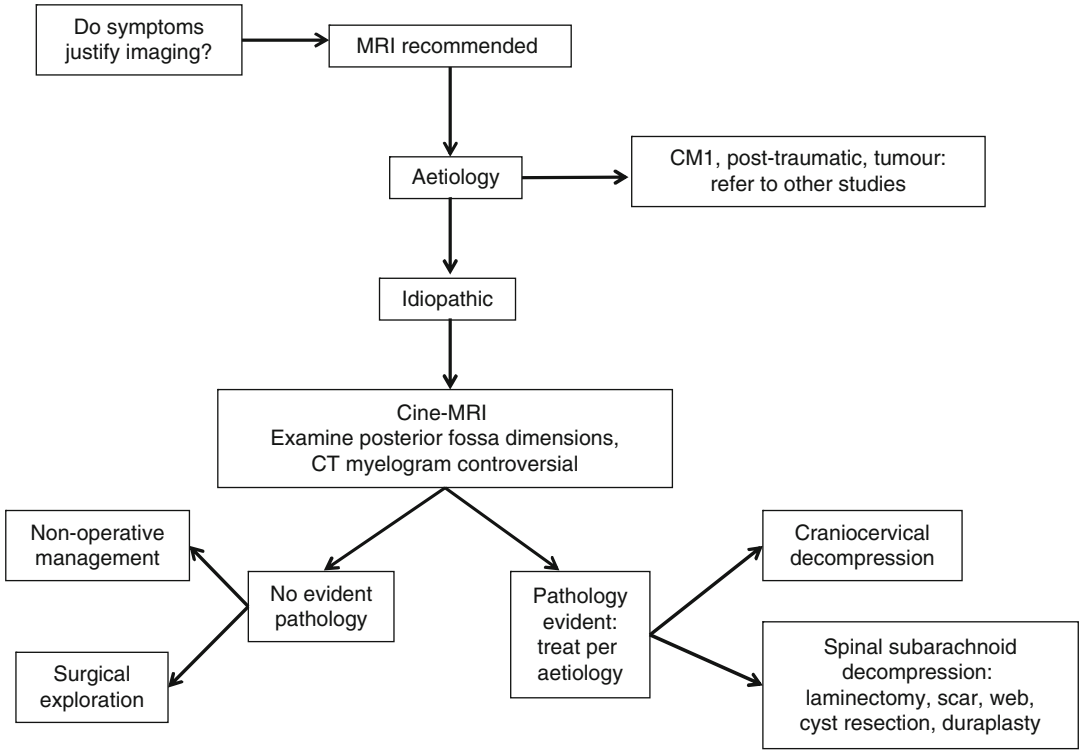

Fig. 12.1 Management algorithm for idiopathic syringomyelia. Indications for imaging include progressive deterioration in signs or symptoms (Roy et al. 2011)

Conclusion

Idiopathic syringomyelia is a pathological entity where no overt aetiology is evident for a syrinx. As imaging technology develops, increasing numbers of apparently idiopathic syringomyelia are being attributed to CSF flow abnormalities. Most incidental cases of idiopathic syringomyelia can be successfully managed using conservative approaches. With regard to surgical options, for continued progression of symptoms, syrinx shunting is generally a less-favoured approach as it does not resolve the underlying aetiology and is associated with high failure rates. A particular challenge to the neurosurgeon is surgical treatment of the patient with worsening symptoms but with no overt aetiology for their syrinx, even after a complete diagnostic workup, including flow studies. While we recommend surgical exploration in these cases, future studies will, hopefully, reveal a more systematic approach for these patients.

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