Idiopathic Syringomyelia in a Military Helicopter Pilot

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BACKGROUND: A syrinx is a fluid-filled cavity within the spinal cord. They can lead to a variety of symptoms, including limb weakness and back pain. Incidental finding of syringomyelia provides a challenge for clinicians due to the wide variety of possible symptoms. In military aviation, neurological findings in pilots can result in extensive investigation that can lead to potentially invasive management. Conversely, the potential for chronic progression of a spinal syrinx and subsequent neurological deterioration makes early identification critical. Ultimately, the discovery of a lesion may have implications for flying status and operational capability.

CASE REPORT: A 25-yr-old man working as a navy Seahawk helicopter pilot presented with episodes of right arm paraesthesia and pain between the scapulae. On at least one occasion, these symptoms woke him at night. Upon magnetic resonance imaging, dilatation of the central canal in a syrinx-like pattern in the lower cervical region was noted. Neurology review suggested the finding was persistent and unlikely to be responsible for his symptoms. No surgical input was recommended. His symptoms were attributed to mild cervical spondylosis, which resolved with ongoing physiotherapy, and he was returned to flying status.

DISCUSSION: This case highlights several issues involved with the incidental finding of a syringomyelia. Surgical intervention has been known to worsen symptoms. Conversely, studies have identified minimal radiological progression in cases of idiopathic syringomyelia, with fewer individuals displaying neurological deterioration. For aircrew, potentially unnecessary neurosurgical intervention poses risks to a flying career and overall operational capability.

KEYWORDS: syrinx, paraesthesia, musculoskeletal.

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Syringomyelia is a term that refers to a dilatation of the central canal of the spinal cord, or the formation of abnormal cavities within its substance.[3] Most commonly, this cavity is the result of one of several associated disease processes or anatomical abnormalities, including Chiari malformation Type I (CM-I), spinal cord tumor, trauma, or infection.[16] Less commonly however, a syrinx is detected where no plausible causative factor or agent has been identified, with a wide variety of presentations noted in the current literature.[1,3,16] Such findings of idiopathic syringomyelia (IS) are becoming more frequent, due in part to the advent and increasing use of magnetic resonance imaging (MRI). Additionally, one must consider the terminology in use when assessing this phenomenon, as the terms hydromyelia and idiopathic syringomyelia, while interchangeable to some,[16] have also been considered as two distinct entities,[15] where the term syringomyelia is referring to a condition in those patients with an underlying pathology.

From an aerospace medicine perspective, the finding of such spinal cord entities presents a challenge for flight surgeons with regards to monitoring and ongoing management. The invasive nature of neurosurgical intervention can have long-term sequelae, particularly when decompressive surgery may not be necessary. The purpose of this case report is to add to the literature that identifies findings of IS in active aircrew and assist in an approach to management.

CASE REPORT

A 25-yr-old man working as a navy S-70B-2 Seahawk helicopter pilot presented with several weeks of sharp pains in his upper back upon waking, with radiation through both scapulae. He also described intermittent tingling paraesthesia

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through the right arm, extending to and including the little finger. He could not recall the circumstances surrounding symptom onset, though he suggested it was not abrupt. His symptoms were not worsening or abating in intensity or frequency. They woke him from sleep on at least one occasion. There was no gait disturbance suggested, nor was there a preceding respiratory or gastrointestinal illness that the patient could recall, or noted in his records. Items of note in his history included a head impact after diving into a sandbar while surfing the year prior to the current presentation. His c-spine was cleared in the local emergency department (a process that included a CT scan) and there was no suggestion of fracture, ongoing injury or disability following the impact. He also suffers from intermittent left lower back pain following a kickboxing injury at age 21, which responds well to physiotherapy on an as-needed basis. These symptoms, however, were identified by the patient as clearly distinct from the newer symptoms prompting the review in question.

On examination, a review of cranial nerves and higher cerebral function noted no abnormalities. Similarly, limb strength and reflexes were normal, as was gait. There was a suggestion of reduction in light touch sensation on the right arm in a medial cord distribution; however, this was difficult to reproduce. The examination was otherwise unremarkable.

Given the nature of the symptoms and the imaging modalities available, MRI of the spine was arranged. A subtle lower cervical cord dilatation was noted over a length of 4–5 cm, most prominent between C_5 and C_7, with a maximum transverse diameter of 3 mm (**Fig. 1**). A further subtle prominence of the central canal was seen opposite T_3. No distinct tumor formation or abnormal enhancement was identified. Notably, there was no impingement within the cervical canal, nor were there any disc, facet joint, or neural exit abnormalities identified. Given the symptoms, the radiological findings, and the solo-pilot flight profile for an S-70B-2 captain, the member was made temporarily medically unfit for flying (TMUFF) pending specialist review. The neurologist noted no abnormal findings on examination, but sought input from a specialist complex case conference given the nonspecific nature of the presentation. The final specialist opinion suggested that the central cord dilatation was likely persistent and not responsible for his symptoms, which were attributed to cervical spondylosis. No follow-up was recommended. The patient was returned to unrestricted flying status as his symptoms gradually improved following regular physiotherapy sessions. These involved a targeted postural exercise program aimed at correcting strength and flexibility in the middle back, neck and shoulders.

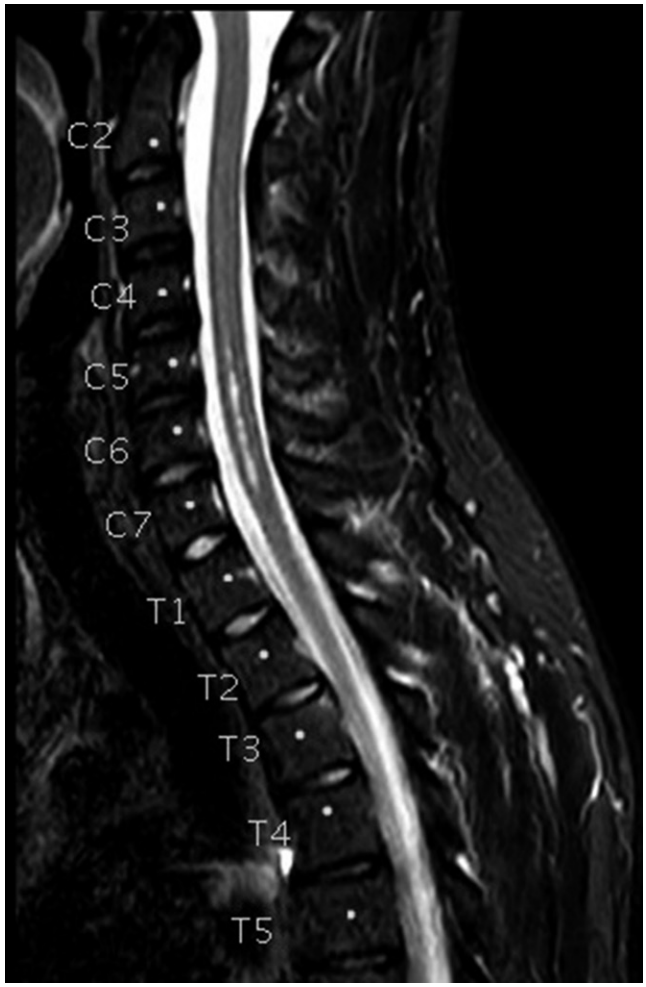

Fig. 1. MRI showing dilatation of the central canal.

DISCUSSION

The findings in this case immediately raise questions regarding discrepancies in terminology and classification. Some identify that the term syringomyelia has previously been classically referred to as a central medullary syndrome,[16] which is not the case here. Similarly, others consider syringomyelia to be a condition related to an underlying disease process, which again does not fit with the findings in our case patient.[15] Others still identify a dilatation of the spinal cord (hydromyelia) vs. a dissection of the ependymal lining of the central canal and a cerebrospinal fluid collection within the cord itself (syringomyelia).[11] From a clinical perspective, this distinction is largely irrelevant and depends on the size of the cyst and the potential compression on the surrounding nerve tissue. Consideration should also be given to the likelihood of a persistent canal dilatation, which is again considered by some to be a distinctly different entity to a 'true' syringomyelia.[6] Holly and Batzdorf[6] examined 32 patients with 'slit-like' syrinx cavities, with a mean central spinal cord cavity diameter of 2 mm. Of those patients, 50% had other diagnoses to explain their symptoms (such as the case patient), and 10 had a history of trauma. Our case patient, however, would likely fit with those cases with syringomyelias deemed idiopathic. Nakamura et al.[13] divided IS into a 'localized' type (possibly representing congenital central canal enlargement) and an 'extended' type (which causes progressive neurological dysfunction and would be more likely to require surgical intervention).

Clearly there are differences in opinion regarding the classification of syringomyelias and multiple factors come into play

when attempting to determine their true nature. As surmised by Roy et al.,[16] it is difficult to determine if slitlike syringes, hydromyelia, and IS are truly different entities or simply a continuum on a spectrum.

There are several theories regarding the pathogenesis of syringomyelia.[4,5,12] Most research into the area focuses on development of the condition in those patients so predisposed with CM-I, with a classification system raised by Koyanagi and Houkin[12] regarding existing evidence as to the cerebrospinal fluid source of origin: entrance from the fourth ventricle, the subarachnoid space, or extracellularly (the latter of which is also discussed by Klekamp[10]). A detailed analysis of these theories is beyond the scope of this report. A relevant factor in these existing theories, however, is the anatomical features that predispose individuals to the formation of a syrinx in those that do not have evidence of primary disease.[2] Bogdanov et al.[2] identified clinical features and posterior fossa measurements (PF) noted in 17 idiopathic syringomyelia patients, 17 patients with Chiari I-type malformation, and 32 controls. They surmised that 'idiopathic syringomyelia and Chiari I-type syringomyelia manifested central cervical myelopathy and a small PF with narrow CSF spaces, suggesting they develop by the same mechanism'. They found that the IS patients were similar in anatomical structure compared to the CM-I patients; compared to the controls, they had 'significantly shortened PF bones, reduced PF height [and thus generally] underdeveloped osseous PF structures and skull based flattening.'[2] Further, Heiss[5] looked again at patients with Chiari I malformations and notes the relevant soft tissue changes that occur following decompressive surgery, particularly the disappearance of the abnormal shape and position of the cerebellar tonsils. This would suggest that some of the aforementioned anatomical features may be acquired rather than genetic.

More pertinent to this report, however, are those patients identified in the literature with IS. Since the case patient did not have posterior fossa deficiencies noted on his imaging, nor any other anatomical or historical item predisposing to the development of such a condition, the 'idiopathic' identifier is likely suitable given the current evidence. Upon review, there are several studies and case reports in the literature looking at IS, the presenting characteristics, the progression of symptoms,[7,9] and management of the condition.[13,16]

Symptoms at presentation are varied. From the aforementioned cases identifying patients with IS, there are complaints of severe neck and back pain,[1] proximal upper limb weakness and gait disturbance,[3] radicular pain, paraesthesia, numbness, and muscle spasm.[6] Based on symptoms alone, it is very difficult for the clinician to establish a sound diagnosis without the added information obtained by a variety of imaging techniques that may be available. Any one of these symptoms may very well prompt the request for an MRI, depending on the overall clinical picture obtained from both a detailed history and neurological examination. In short, the symptoms at presentation do not necessarily inform the case management. As with most presentations in medicine, the relevance lies more in the patient response to management following relevant investigations (if any), and the progression, abatement, or persistence of their presenting symptoms. In this case patient, while the imaging findings lead to specialist input suggesting the syrinx was unlikely to be responsible for the presenting symptoms, it was the response to treatment that enabled a more confident plan involving ongoing monitoring, management, and a decision regarding flight status.

Many syringomyelia cases will require surgical intervention to decompress the affected region with an aim to reduce the severity of symptoms and, perhaps more importantly, reduce symptomatic progression. Heiss[5] looked at 20 adult patients with symptomatic syringomyelia and known CM-I, and identified that 'clinical signs and symptoms improved or remained stable in all patients' following decompressive surgery. As mentioned, however, care must be given in determining the cause of the symptoms accurately, regardless of the abnormal findings on imaging that may confound the diagnosis. More relevant to this case are those previous reports identifying spontaneously resolving conditions[8] and reviews monitoring progression, such as that conducted by Kim et al.[9] in 2012. Over a mean of 39.9 mo, 12 patients with IS, none of whom had any predisposing pathology, were followed with radiological imaging. Of the 12 patients, 11 had no change on MRI and all patients included in the study experienced clinical symptomatic improvement without the need for surgical intervention. Jinkins et al.[7] followed three patients over 24 yr with similar results. As previously identified, Nakamura et al.[13] noted a distinction between localized and extended syringomyelia. Of 15 patients included in the study with IS, 12 were treated conservatively, and none had any changes in syrinx size or neurological symptoms throughout the follow-up (with a mean of 10 yr). Holly et al.[6] identified 32 case patients with slit-like syrinx cavities, which is possibly a suitable category into which the case patient in this report might fall. Of the 31 patients who did not undergo surgical intervention, 6 improved clinically, 7 got worse, and 19 underwent no change. More importantly, however, was that none of the syrinx cavities changed in size (after a mean follow-up period of 32 mo). Critically, 16 of these patients were later identified to have alternative diagnoses as the cause of their symptoms. The author concluded that these syrinx findings are often incidental and are unlikely to change in size.

Several factors need to be taken into account when determining whether or not this patient should have been returned to flying status. Considering a published suggestion for an aeromedical decision-making algorithm,[14] it is a relatively straightforward process to determine this patient's fitness to fly. The likelihood of a clinically significant event occurring in this case is quite low, given the lack of progression and recurrence of symptoms noted for IS patients identified in the current literature.[7,9,16] The symptoms were slow onset, not related to the syrinx, and not incapacitating. Given that the cavity is fluid filled, expansion secondary to ambient pressure changes is less of a concern when compared to an air cavity within a body space. Certainly consideration must be given to the type of flying involved, and solo-pilot military rotary wing operations would certainly prompt any flight surgeon to cautiously consider the options available. Given these findings and an

effectively treated condition with simple, nonsurgical intervention, there is no real reason to restrict this pilot in his capacity as a military rotary wing flyer. Considering the literature, as well as the input received from the specialists to whom we referred, there is no need for ongoing monitoring with imaging unless the symptoms recur and cannot be attributed to another cause. A follow-up MRI of the spine at 1–2 yr post review, however, to check for a change in the size of the syrinx would not be unreasonable.

Incidental findings of a syrinx present a challenge for clinicians. This becomes more difficult should no alternative diagnoses be apparent in a symptomatic patient. The literature would support a view to conservatively manage those patients with idiopathic syringomyelia unless symptoms are progressive, and decompressive surgery is considered to be beneficial at the time. From an aeromedical perspective, the symptom onset is rarely acute in a patient with a long-standing syrinx that has progressed to the point where symptoms are now apparent, which has relevance to the possibility of incapacitation in an aircraft. If the onset is acute, then alternative diagnoses need to be carefully considered before a management plan and potentially invasive intervention is decided upon. Regardless, should effective treatment be implemented and the syrinx is determined to be idiopathic and unlikely to be the cause of the patient's symptoms, a change in flight status is not warranted. A monitoring regimen can be implemented depending on access and availability of the relevant modality at the discretion of the treating team.

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