Case Report

Spine pathology in a girl with upper limb pain: A co-incidence or a causal relationship?

Maria Gogou (1), Martha Spilioti (2), Despoina Tramma (1), Efimia Papadopoulou-Alataki (1), Athanasios Evangeliou (1)

(1) Department of Pediatrics, School of Medicine, Aristotle University of Thessaloniki, General Hospital Papageorgiou, Thessaloniki, Greece
(2) Department of Neurology, Aristotle University of Thessaloniki, University General Hospital AHEPA, Thessaloniki, Greece

ABSTRACT

A 10-year-old girl was admitted to the Emergency Department due to a history of intermittent pain located in the left radiocarpal joint for a month, as well as in the interphalangeal joints of the left hand without any additional symptoms. Clinical examination revealed mild sensory deficits and diminished muscle strength of the left upper limb without any other pathologic findings. A Magnetic Resonance Imaging scan of the brain and spinal cord was performed, which confirmed a diagnosis of thoracic syringomyelia. We briefly discuss specific traits and diagnostic challenges of this entity in childhood. Our case highlights the difficulty in efficiently correlating a pathologic imaging finding with clinical neurologic symptoms and signs, as well as the value of a thorough clinical neurological evaluation. Furthermore, a clear discrimination of a causal relationship against an incidental co-existence of a radiological finding and a specific symptom is not always possible.

Keywords:

Syringomyelia; Children; Intermittent Pain; Neurologic Examination; Spine Cord Pathology

INTRODUCTION

Pediatric spine pathology poses a diagnostic challenge for clinicians, as it often yields nonspecific signs and symptoms, especially in the younger age groups, thus leading to diagnostic delays [1]. We present a case here that demonstartes the difficulty in efficiently correlating a pathologic imaging finding with clinical neurological symptoms and signs in some cases.

CASE REPORT

A 10-year-old girl presented to the emergency pediatric department due to a month history of pain located in the left radiocarpal joint, as well as in the interphalangeal joints of the left hand. The pain was intermittent, had no specific traits, did not awake her from sleep and responded to analgesics. No signs of inflammation were present and there was full passive range of motion of the left hand. On the other hand, discomfort was revealed when the above joints were separately tested. Although no apparent neurological deficits were present, a more thorough neurological evaluation revealed hyperesthesia to light touch on the extensor aspect of the left forearm, as well as in dermatomes T4-T5. Muscle strength

Correspondence to:
Maria Gogou
Department of Pediatrics, School of Medicine, Aristotle University of Thessaloniki, General Hospital Papageorgiou, Thessaloniki Dimitriou Nika 44, 60 134, Katerini, Greece
Tel.: 0030 6978905873
E-mail: mariaangogou@gmail.com

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of the left hand was slightly diminished grade (4). The remainder of clinical examination had normal findings. Anthropometric data were within normal ranges for gender and age.

The girl was born to two phenotypically healthy parents and her medical history was unremarkable, apart from intermittent lower limbs scissoring during infancy and nocturnal enuresis until early childhood, which resolved spontaneously.

Common laboratory tests (complete blood count, biochemical tests, coagulation tests) and an X-ray of the left hand had no pathologic findings. Electromyography demonstrated no signs of root impingement or entrapment of the median or ulnar nerve. A Magnetic Resonance Imaging study of the brain and spinal cord was requested, which revealed thoracic syringomyelia (dilatation and visualization of central canal) from the C8 to T9 vertebral level. Contrast injection was negative Figure 1, 2. The girl was neurosurgically evaluated and after a mild improvement of the symptoms was discharged home with recommendations for a regular evaluation in the outpatient clinic of Pediatric Neurology.

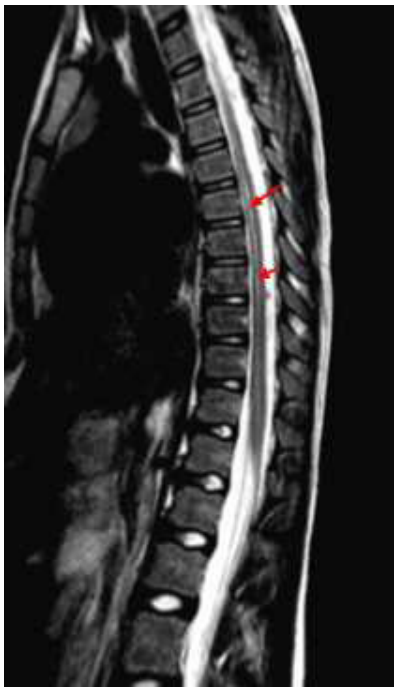

Figure 1 - Sagittal MRI scan showing visualization of central canal in the thoracic part of spinal cord (arrows), a finding compatible with syringomyelia.

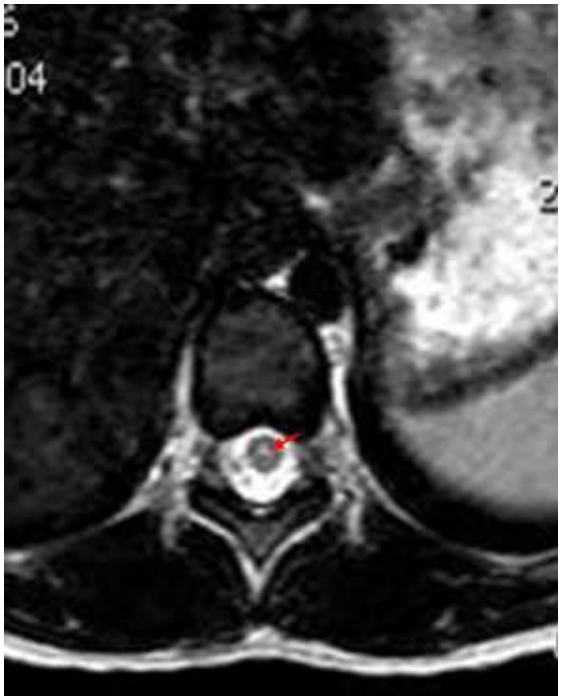

Figure 2 - Subtle visualization of central canal (arrow) in axial MRI scan of spinal cord.

DISCUSSION

Syringomyelia is a fluid-filled cystic cavitation of the spinal cord parenchyma and its estimated prevalence is 8 cases per 100.000 population. The underlying mechanism includes disturbance of normal cerebrospinal fluid flow dynamics, which either arises from an abnormal dilatation of the central canal of the spinal cord (congenital form) or results from a block of normal cerebrospinal fluid circulation (acquired form) [2]. Although it can be diagnosed in isolation, syringomyelia in children usually occurs in the setting of congenital anomalies, most commonly the Arnold-Chiari-I malformation and tethered cord, but it can also develop after meningitis and spinal trauma or with intramedullary and extra medullary tumors [3]. Polymorphisms of the

CollA1 gene and the vitamin D receptor gene, as well as 16p11.2 rearrangements have been correlated with this condition in specific populations [4,5].

Symptoms at presentation are usually mild and include limb numbness, sensory and motor deficits and headaches, although the contribution of the syrinx to the above symptoms has been in many cases controversial [6]. Waseem, et al. (2012) reported a case of syringomyelia in an adolescent female with paroxysmal neuropathetic pain, while cases of cervical syringes in children with isolated Horner syndrome have also been described [7,8]. Moreover, syringomyelia of the thoracic cord has been revealed in children with urinary tract dysfunction without any other abnormality of Central Nervous System and it is also interesting that syringomyelia often co-exists with scoliosis in childhood [9]. According to Innoue, et al. (2005), 18% of patients classified as having idiopathic scoliosis were found to have neural axis abnormalities and mainly syringomyelia when evaluated with Magnetic Resonance Imaging, while Haniech, et al. (2000) reported that 20% of children with idiopathic scoliosis were found to have syringomyelia when referred for imaging. A significant indication for selecting children with scoliosis likely to have syringomyelia is an abnormal neurologic examination (asymmetric reflexes, abnormal superficial abdominal reflexes) [10,11].

With regards to natural history, in most cases the syrinx either remains unchanged or diminishes in size. Cases of spontaneous resolution have also been reported and attributed to spontaneous recanalization of cerebrospinal fluid pathways [12]. In the minority of cases there is an increase in syrinx size, not necessarily combined with worsening of the course [13]. Due to this benign clinical course, syringomyelia can be managed expectantly and an etiology-driven approach including periodic imaging in the follow-up is recommended, while increase in syrinx size along with continual progression of symptoms consist indications for neurosurgery intervention [6]. However, deficits, once established, may show only little improvement after surgery. In this way, an individualized approach is recommended in most cases.

Our girl presented for evaluation of chronic intermittent pain located in the left radiocarpal and interphalangeal joints. The pain had no specific traits and although no apparent neurological signs were present, a careful neurological evaluation revealed mild sensory disorders of the left forearm and slightly diminished muscle strength of the left hand. Sensory innervation of the distal parts of the upper limb is provided by the lower segments of the brachial plexus (C8-T1 nerves) and under this view pain and sensory disorders of these parts could be associated with thoracic syringomyelia. Moreover, the fact that hyperesthesia was also present in dermatomes T4 and T5 further strengthens a potential involvement of thoracic syringomyelia in the appearance of these symptoms.

The girl also presented symptoms in her infancy (lower limb scissoring) and early childhood (nocturnal enuresis), which have been reported in literature in cases of children with syringomyelia. Given the possibility of spontaneous resolution of syringes in childhood, it could be assumed that our girl presented in a younger age a more extended syringomyelia, including the lumbar and sacral parts of the spinal cord, which could have led to these symptoms, as motor innervation to the lower limbs is provided by lumbosacral plexus, whereas sympathetic and parasympathetic innervation to the urinary bladder is mediated through pelvic and sacral splanchnic nerves.

On the other side, it should be noted that in our case diagnosis of syringomyelia was mainly based on visualization of central canal in the thoracic part of the spinal cord. Nevertheless, this finding can often be within normal limits and in such cases there may also be an overlap between normal and pathologic findings. The subtle spine pathology could also account for normal findings in the electromyography testing in our patient. In this way, although the involvement of the thoracic syrinx to the girl's symptoms is reasonable, the exact degree of contribution is still under discussion.

CONCLUSION

In general, although the advancement of imaging techniques is anticipated to result in more incidental idiopathic syringes without evident symptoms, a detailed medical history and examination can reveal subtle disorders, which may be associated with the presence of the syrinx. Syringomyelia can lead to a variety of clinical features and may be the underlying mechanism or a contributing factor of common disorders of childhood, such as scoliosis. A thorough clinical neurologic evaluation seems to be the key for an early diagnosis of this condition in children presented with non-specific symptoms.

REFERENCES

1. Palasis S, Hayes LL. Acquired pathology of the pediatric spine and spinal cord. Pediatr Radiol 2015; 45:420-32.
2. Vandertop WP. Syringomyelia. Neuropediatrics 2014; 45:3-9.
3. Bixenmann BJ, Kline-Fath BM, Bierbrauer KS, Bansal D. Prenatal and postnatal evaluation for syringomyelia in patients with spinal dysraphism. J Neurosurg Pediatr 2014; 14:316-21.
4. Abulgatina AS, Khusainova RI, Khidiiatova IM, et al. The analysis of the SpI (1546G>T) polymorphism of the Col1A1 gene and the FokI (3663T>C) polymorphism of the vitamin D receptor gene in patients with syringomyelia. Zh Nevrol Psikhiatr Im S S Korsakova 2007; 107:60-6.
5. Schaaf CP, Goin-Kochel RP, Nowell KP, et al. Expanding the clinical spectrum of the 16p11.2 chromosomal rearrangements: three patients with syringomyelia. Eur J Hum Genet 2011; 19:152-6.
6. Singhal A, Bowen-Roberts T, Steinbok P, Cochrane D, Byrne AT, Kerr JM. Natural history of untreated syringomyelia in pediatric patients. Neurosurg Focus 2011; 31:E13.
7. Waseem M, Raja A, Jeun E, Prokhorov S, Dobrenkov K. Paroxysmal neuropathic pain in an adolescent female with syringomyelia: a review of literature. Pediatr Emerg Care 2012; 28:472-4.
8. Moreno TA, El-Dairi MA, Cabrera MT. Isolated Horner syndrome and syringomyelia in a child. J AAPOS 2012; 16:569-70.
9. Kothari MJ1, Kelly M, Darbey M, Bauer S, Scott RM. Neurophysiologic assessment of urinary dysfunction in children with thoracic syringomyelia. J Child Neurol 1995; 10:451-4.
10. Inoue M, Minami S, Nakata Y, et al. Preoperative MRI analysis of patients with idiopathic scoliosis: a prospective study. Spine 2005; 30:108-14.
11. Hanieh A, Sutherland A, Foster B, Cundy P. Syringomyelia in children with primary scoliosis. Childs Nerv Syst 2000; 16:200-2.
12. Kyoshima K, Bogdanov EI. Spontaneous resolution of syringomyelia: report of two cases and review of the literature. Neurosurgery 2003; 53:762-8.
13. Magge SN, Smyth MD, Governale LS, et al. Idiopathic syrinx in the pediatric population: a combined center experience. J Neurosurg Pediatr 2011; 7:30-6.