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Anterior Spinal Artery Syndrome in a 14-Year-Old Boy

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Abstract

Acute flaccid paralysis caused by anterior spinal artery syndrome (ASAS) is rare in children. It typically manifests as bilateral loss of motor function, pain, and temperature sensation below the level of occlusion, with relatively little impairment in proprioception and vibration sense. We present such a case in a 14-year-old child who presented with a sudden onset of neck pain followed by the typical symptoms of ASAS with impaired breathing due to the height of the lesion, which was found in the magnetic resonance imaging examination at the level of C1–5. An initially suspected thrombogenic cause proved inapplicable. Ultimately, despite extensive diagnosis, as in most cases of ASAS in children, the cause remains unclear.

Keywords
- acute flaccid paralysis
- spinal cord infarction
- anterior spinal artery syndrome
- neck pain
- thrombolysis

Progressive acute paresis without trauma in a previously healthy child is an emergency that requires immediate diagnosis. Ischemia of the spinal cord is rare and most often due to anterior spinal artery syndrome (ASAS).[1,2]

Here, we present the case of a 14-year-old boy who had ASAS diagnosed clinically and with magnetic resonance imaging (MRI).

Case Presentation

A 14-year old Caucasian boy experienced sudden onset of pain in the neck and thoracic spine. A relative attempted to achieve relief by performing a spine maneuver, and there was a cracking sound. Approximately 1 hour after the onset of symptoms, progressive paresis of the arms and legs developed, so the boy was presented to the emergency room of a childreńs hospital.

The previously healthy boy had prior experience of recurrent neck pain and thus performed stretching exercises; however, history was otherwise uneventful, and no smoking was reported. Family history showed antiphospholipid antibody syndrome in his father, treated with rivaroxaban.

On examination, the alert boy showed bilateral paresis of the arms (1/5), right accentuated paresis of the legs (2/5), and mild weakness of the left leg. The biceps and triceps tendon reflexes were very weak, and the reflexes of the lower extremities were normal. There was less pain in the left leg. The patient also showed increased respiratory symptoms, with tachypnea and paradoxical breathing.

Blood gas analysis showed mild respiratory acidosis, and the initial emergency laboratory sampling was unremarkable (normal blood count and negative inflammatory parameters). Chest radiography findings were normal.

Spinal MRI, T2- weighted sequence, showed no fracture, no hemorrhage, no spinal cord compression, or signal change in vertebral bodies. In addition, no contrast uptake was observed. Cranial magnetic resonance imaging showed no cerebral involvement.

With continued progressive symptoms, the patient was transferred to a tertiary care hospital equipped with neurosurgery.

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Upon further investigation, CT angiography did not reveal any vascular malformations, dissection, or bleeding. Liquor examination showed negative cell count and no blood–brain barrier disruption. Infectious diagnostics of liquor, blood, stool, and urine, including multiplex PCR block analysis of the liquor for meningitis-causing patterns, did not detect any pathogens (syphilis, herpes simplex virus 1 and -2, varicella zoster virus, enterovirus, Lyme borreliosis, human immunodeficiency virus, cytomegalovirus, coxsackievirus A/B, adenovirus, and Epstein–Barr virus tested negative). Blood examinations showed no evidence of autoimmune (negative antinuclear and antineutrophil cytoplasmic antibodies indicative of vasculitis) or coagulation abnormalities (antithrombin III, protein C- and S activity, factor V Leiden, lupus anticoagulant, prothrombin variant, apolipoprotein A and B, anticardiolipin antibodies, β2- glycoprotein, and homocysteine, factor VIII).

The following day, the boy had sensory dysfunction from level C4, left-sided flaccid hemiparesis (plantar flexion M4-5, knee flexion M4, leg raising, and first closure M4), and right-sided hemiplegia. The Achilles and patellar tendon reflexes were triggerable bilaterally. Motor reflexes were absent in the upper extremities. Electroneuromyography was not part of the acute diagnostic procedure, because there was no evidence of demyelinating disease.

The boy was suspected to have arterial spinal anterior syndrome, and intravenous lysis therapy (alteplase 50 mg) was started. In addition, the patient received antiplatelet therapy (ASS) and low-molecular-weight heparin (enoxaparin).

In the subsequent course, the patient experienced respiratory failure and needed mechanical ventilation.

Two days after admission, another MRI scan showed increasing demarcation of a prolonged spinalis anterior infarction in the gray matter of the rostral myelon at the level of C1–5 (►**Figs. 1** and **2**). A thrombophilia workup showed a strongly elevated factor VIII.

After 10 days, the patient was transferred to a rehabilitation center and tracheotomized. Three days after transfer to the rehabilitation center, the patient was able to breathe on his own and was no longer tracheotomized. Motor development was also very encouraging owing to intensive rehabilitative therapy. Three months after the onset of symptoms, the patient could walk without aid and perform activities of daily living independently. Deficits were still evident on the right side of the body, whereas the left side was not impaired. Follow-up MRI examinations still showed isolated postischemic small-volume myelon lesions with completely regressed perifocal edema compared with the initial findings (►**Fig. 3**).

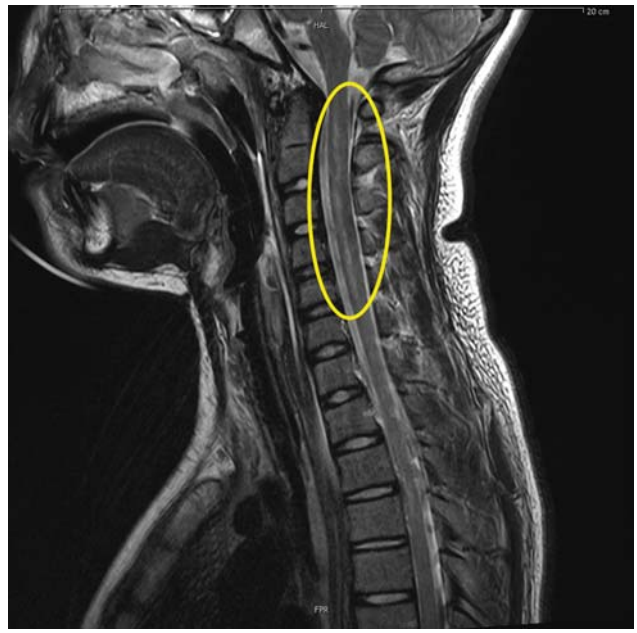

Fig. 1 Sagital T2-weighted MRI: Demarcation of a spinal infarct in the gray matter of the rostral myelon of C1-5 with diffusion disturbance and edematous volume increase of the cervical myelon.

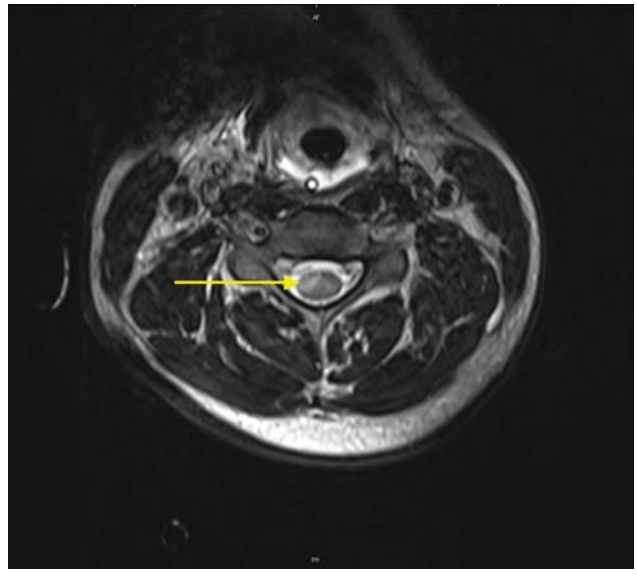

Fig. 2 Axial T2-weighted MRI sequence at the level of C3 shows hyperintensity with restricted diffusion.

Discussion

Acute flaccid paralysis is a clinical syndrome characterized by rapid onset of weakness, possibly including respiratory muscle weakness.[3] The most common differential diagnoses include transverse myelitis, poliomyelitis, Guillain–Barré syndrome, and all possible forms of spinal trauma or infection.[4]

The anterior two-thirds of the spinal cord contains important pathways for the functioning of the central nervous system and is supplied by the anterior spinal artery (ASA).[2,5–7]

Infarction of the ASA typically manifests as bilateral loss of motor function, pain, and temperature sensation below the level of occlusion, with relatively little impairment in proprioception and vibration sense.[1,2,8] Moreover, the sympathetic nervous system may be involved in autonomic dysfunction (hypotension, sexual dysfunction, and bowel/bladder dysfunction).[2,5] If the lesion is located in the rostral nuchal medulla, as observed in our patient, the phrenic nerve and respiration may be affected.[1,2]

A diagnosis of ASAS should be made clinically using imaging (MRI).[2,5,8] MR examinations should include axial and sagittal diffusion-weighted imaging.[5] The characteristic

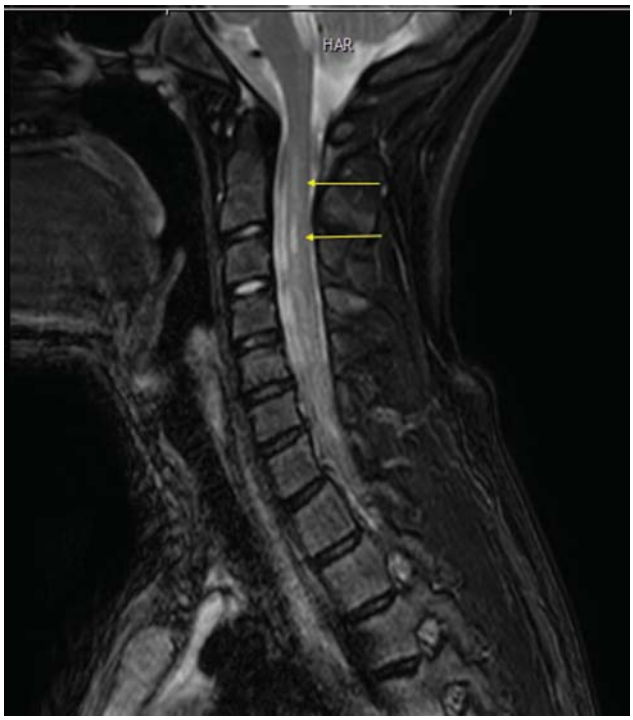

Fig. 3 Sagital T2-weighted MRI (13 month after onset of symptoms and ASAS diagnosis): postischemic small-volume myelon lesions at level C2-5.

MR features of ASAS include diffusion restriction in the ASA territory on diffusion-weighted images and a pencil-like hyperintense signal on sagittal T2-weighted images[1,5,9] that extends vertically and spans multiple spinal levels.[2] Corresponding axial images may show hyperintensity with restricted diffusion in the anterior spinal cord, the so-called owl's eye sign.[2,8,10] It is important to note that the results may be negative in the early phase during the first 24 hours.[2,9,11] Other imaging modalities may aid in the diagnostic classification. Spinal angiography can help assess the etiology of spinal cord infarcts,[12] especially when initial MRI imaging does not reveal any landmark findings, as in our patient.

ASAS in children is much less common in children than in adults. In adults, aortic surgery or signs of aging such as atherosclerosis are possible triggers; nevertheless, even in adults, the etiology is often not identified.[1,13] Trauma and hematoma can directly bruise or compress the spinal cord.[5] Indirectly, systemic hypotension or vascular occlusion (traumatic, iatrogenic, thrombotic, or embolic cause) may result in reduced blood flow in the spinal arteries or their supplying arteries.[7,14] In contrast, inflammatory or infectious myelitis should be excluded.[2,8,14,15] Determination of aquaporin-immunoglobulin G (IgG) and myelin oligodendrocyte glycoprotein-IgG in serum, which are associated with pediatric acquired demyelinating syndromes (like transverse myelitis, acute disseminated encephalomyelitis and multiple sclerosis), and oligoclonal bands in cerebrospinal fluid, as inflammatory marker,[16] was not performed in our patient. In patients with unclear flaccid paresis, the determination of these parameters should be considered.

Thrombophilia, which may predispose to thrombotic occlusion, has been detected in some children with ASAS. Protein S deficiency,[17] prothrombin variants,[17,18] antiphospholipid antibodies,[19] and factor V Leiden[20] have been reported in several case reports. In our case, factor VIII was elevated by 277% as a prothrombotic finding. However, patients with spinal cord infarction often have reactively increased factor VIII activity.[21] In our patient, during follow-up, the levels of factor VIII returned to normal. A genetic predisposition of antiphospholipid antibody syndrome was present due to the disease of the father, but antiphospholipid antibodies and lupus anticoagulant could not be detected. In addition, there were no mutations in factor V Leiden or a specific prothrombin variant.

The theory of minitrauma is very frequently described, which may lead to ASAS, although the exact origin is not fully understood. Pang [22] proposed that in children, the relatively less elastic spinal cord is stressed within the more flexible spine during hyperflexion injuries, which may lead to reactive vasospasm of the spinal arteries causing ischemia. Traction on the spinal cord by hyperextension during surfing, combined with additional risk factors (slender habitus, little muscle), could lead to the severing of perforating vessels, vasospasm, or ischemia.[23]

In our patient, there was no known trauma beforehand, but he appeared to have chronic cervical and thoracic spine discomfort, for which he performed regular maneuvers himself or through others to provide relief. Whether and to what extent such maneuvering was related to the ASAS that ultimately developed remains unclear.

However, pre-existing chronic back pain has frequently been reported in patients with ASAS.[15]

An alternative cause of spinal cord infarction is embolism of the nucleus pulposus intervertebral disc, the so-called fibrocartilaginous embolism, which mostly involves the ASA[15] and occurs most commonly (two out of three of the cases) at the cervical level.[15,24]

Many of these cases are also associated with minor trauma or sudden jerky neck motion.[15] On MRI, spinal cord swelling associated with a collapsed disc space or a narrowed disc with Schmorl's nodes at the appropriate level is strong evidence of this genesis.[14,15,24] This could not be demonstrated in our patient by MRI. Fibrocartilaginous embolism of the spinal cord can be proven postmortem, but there are some cases in which the imaging diagnosis has been negative despite histopathologic evidence.[24,25] Thus, fibrocartilaginous embolism is a diagnosis of exclusion in surviving patients[13,15] and remains a possible cause of ASAS in our patient.

Other rare causes of ASAS include spinal AV malformation, iatrogenic causes such as surgery of the aorta, placement of an umbilical artery catheter in neonates, traction in scoliosis after orthopedic surgery, cerebellar herniation during lumbar puncture, and sclerotherapy for esophageal varices, which leads to the release of an embolus.[14]

Ultimately, in our patient, the cause remained unclear, as in most cases. However, it should be noted that there is an association with minitrauma, which can cause ischemia of the spinal arteries by different etiologic agents.

No standardized guidelines exist for acute treatment of spinal cord ischemia.[15] Once an underlying cause has been identified, it must be treated as soon as possible.[2] The current treatment is mainly supportive. As in our patient, it was important to secure the airway and maintain blood pressure during spinal shock.[5,8]

A few studies have shown a slight improvement in symptoms after the administration of high-dose glucocorticoids,[2,15] but most cases did not improve. Anticoagulation such as heparin and aspirin may be helpful if the cause is thrombotic.[14] Intravenous thrombolysis is an effective treatment for ischemic cerebral infarctions in children.[26] Thrombolysis is also thought to have a beneficial effect on spinal cord infarction but is not currently an established treatment regimen.[8,11,27]

Further treatment primarily involves improving the symptoms and preventing future complications.[5] This is done during early rehabilitation, which should start in the hospital and include intensive physical therapy and occupational therapy, which should be performed by medical professionals. In addition, psychosocial aspects, education of the patient and family regarding the new diagnosis,[6] and the prevention of secondary complications[2] were also included. The goal is to achieve greater independence and quality of life and, ultimately, a better prognosis.[2,5] This is often poor despite intensive therapy, and most patients recover with varying degrees of neurological deficits.[11] Positive predictors were young age and male sex.

Conclusion

Sudden neck or back pain in children, typically at the level of the lesion, motor dysfunction such as para- or tetraplegia, and dissociated sensory disturbances with hypalgesia and thermal hypoesthesia, but with preserved proprioception and vibratory sensation, should suggest spinalis anterior syndrome.

The imaging modality of the first choice is MRI, but this may be initially unremarkable. Often, the etiology remains unclear, and cerebrospinal fluid and blood studies can rule out numerous differential diagnoses.

There is no standardized therapeutic concept. If the etiology is identified, causal therapy should be administered if possible. In addition, supportive therapy and early rehabilitation in a multiprofessional team are in the foreground.

Authors' contribution

F.W. and P.G. concepted and drafted the manuscript, and reviewed and revised the manuscript. M.H., M.S., and S.M. had all significant parts of the conception of the manuscript, and they all critically reviewed it and revised it for important intellectual content.

All authors approved the final manuscript as submitted and agreed to be responsible for all aspects of the work.

Conflict of Interest

None declared.

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