Anterior Spinal Artery Syndrome in a Girl With Down Syndrome: Case Report and Literature Review

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Received August 20, 2008; accepted November, 2008

Abstract

Background/Objective: Anterior spinal artery syndrome is an extremely rare cause of acute ischemic cord infarction in children. It is caused by hypoperfusion of the anterior spinal artery, leading to ischemia in the anterior two thirds of the spinal cord. The presentation is usually with an acute and painful myelopathy with impaired bladder and bowel control. Pain and temperature sensation below the lesion are lost, whereas vibration and position sense is intact because of the preservation of the posterior columns.

Methods: Case report.

Results: A 16-year-old girl with Down syndrome presented with urinary retention and acute complete flaccid paralysis of the legs with absent deep tendon and abdominal reflexes. Magnetic resonance imaging showed a signal abnormality in the anterior half of the thoracic cord from T5 to T12, consistent with anterior spinal artery infarction.

Conclusions: Pediatricians should consider anterior spinal artery syndrome in the child who presents with acute, painful myelopathy. We summarize the etiology, neurological findings and outcomes of 19 children found in the literature with anterior spinal artery syndrome.

J Spinal Cord Med. June 2009;32(3):349–354

Key Words: Anterior spinal artery syndrome; Myelopathy, acute; Spinal cord infarction; Down syndrome; Rehabilitation; Paralysis; Spasticity

INTRODUCTION

Anterior spinal artery syndrome is an extremely rare cause of acute ischemic cord infarction in children. It is caused by occlusion or hypoperfusion of the anterior spinal artery, which supplies the anterior two thirds of the spinal cord. The anterior spinal artery is vulnerable in the T4 to T8 region, because the radicular artery that supplies it (the artery of Adamkiewicz) has poor anastomoses and is effectively an end artery. Review of the literature on ischemic cord infarction from any cause in childhood identified 110 cases (1). Of these, only 13 had proven anterior spinal artery syndrome. In 6 other children, the diagnosis was unproven, but the clinical description and imaging findings were highly suggestive of anterior spinal

artery syndrome. We summarized these 19 cases and included the details of our patient (Table 1).

CASE REPORT

A previously well, obese, 16-year-old girl with Down syndrome presented with acute flaccid paralysis of the legs and retention of urine. Three days previously, she had sudden onset of severe lower back and groin pain with weakness of her legs while traveling on vacation. She was admitted to a local hospital, and a diagnosis of “hysterical paralysis” was made after normal computed tomography scan appearance of her lumbar spine. She returned to the United Kingdom and was admitted to our center.

Physical examination showed a complete flaccid paralysis of the legs with absent deep tendon and abdominal reflexes. Accurate sensory examination of the lower limbs was difficult because of her learning difficulties and level of distress, but sensation to light touch, vibration, and joint position in the lower limbs appeared to be intact. She had acute retention of urine and was in mild obstructive renal failure. Magnetic

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Table 1. Summary of clinical details of 20 children described in the medical literature with anterior spinal artery syndrome including etiology, neurological findings, and outcome

First Author (Reference #)	Age (y)/Sex	Cause	Length of F/U (D/W/M/Y)	Initial Neurological Signs	Final Neurological Signs	Outcome
Non-latrogenic						
Sohal ^a	16/F	Unknown	12 M	Paraplegia. Urinary retention	Power 4 lower limbs. Bladder function impaired	Mobile with rollator device
Wilmshurst (21)	14/F	Unknown	6 M	Paraplegia. Sensory level at T10. Urinary retention	Poor motor activity. Bladder function impaired	Wheel chair dependent
Wilmshurst (21)	15/F	Unknown	6 M	Paraplegia. Sensory level below T9	Walk 5 steps with support. Power-R leg > L leg. Persistent sensory abnormalities. Bladder function impaired	Wheel chair dependent
Wilmshurst (21)	14/M	Unknown	6 M	Paraplegia. Sensory level below L1	Little motor improvement. Power-L leg > R leg.	Immobile ^b
Wilmshurst (21)	16/M	Unknown	8 M	Paraparesis, L leg > R leg. Sensations intact.	Power R leg > L leg	Mobile with support
Wilmshurst (21)	15/F	Unknown	Not specified	Paraplegia. Sensory level at T11. Spared dorsal column modalities. Urinary retention	Minimal motor recovery. Bladder/Bowel function impaired	Immobile ^b
Nance (1)	14/F	Unknown	18 M	Tetraplegia. Facial weakness, absent cough, gag reflex	Minimal movement of right hand. Able to turn head and talk. Bladder/Bowel function impaired	Wheel chair/ventilator dependent
Vandertop (22)	17/M	Unknown	18 M	Left hemiplegia/Right hemiparesis. Intact posterior column modalities. Sensory level below T3–T4. Urinary retention	Power 3–4/5 in upper limbs, 3/5 distally in right leg, 4/5 left leg. Pain and temperature sensation diminished below T2–T3. No urinary/bowel incontinence	Mobile independently
Tosi (23)	16/F	Probable fibrocartilaginous embolism	5 M	Paraplegia. Sensory level below T12. Urinary retention	Not well described	“Poor” (no details)
Toro (24)	16/F	Fibrocartilaginous embolism	6 W	Paraplegia. Sensory level below L1	Slight motor improvement in lower limbs. (R > L). Vibration sensation reappeared below knee. Hypesthesia below L2.	Death secondary bronchial aspiration
Davis (25)	6/F	Fibrocartilaginous embolism.	2 M	Paraplegia. Sensory level below T11	Little improvement in motor activity	Immobile ^b
Yousef (26)	14/F	Fibrocartilaginous embolism	7 D	Paraplegia. Decreased pain, touch below T6	Unspecified. Died after 7 days of first symptom	Death

Table 1. Continued

First Author (Reference #)	Age (y)/Sex	Cause	Length of F/U (D/W/M/Y)	Initial Neurological Signs	Final Neurological Signs	Outcome
Nance (1)	17/M	Heterozygote for C677T, A1298C MTHFR mutations	8 M	Paraparesis. Absent sensations in feet up to shin. Bladder and bowel function retained	Good motor recovery. Decreased proprioception in toes, decreased vibration sensation to knee	Mobile with cane
Ramelli (27)	15.8/M	Protein S deficiency	3 Y	Paraplegia. Sensory level below T10. Loss of bladder and bowel function	Good motor recovery over 12 months. Bladder function impaired	Mobile with clutches
Hakimi(17)	15/M	Prothrombin variant	8 M	Paraplegia. Sensory level below T5	Power 2–4/5 in lower limbs. Able to void spontaneously	Mobile with support
Hakimi(17)	12/F	Protein S deficiency	7 M	Paraplegia. Sensory level below T9. Impaired proprioception	Power-R leg > L leg. Bladder function impaired	Mobile with walking frame
Isatrogenic Boglino (28)	1.5/F	Post removal mediastinal neuroblastoma	2 Y	Paraparesis. Sensory level below T9	Little improvement in motor activity. Anal sphincter preserved. Bladder function impaired	Immobile ^b
Boglino (28)	3/M	Post laminotomy thoracic neuroblastoma	5 Y	Paraplegia. Sensory level below T2. Urinary and faecal incontinence	Reflex motor activity, moderate muscle strength. Reasonable urine and faeces control.	Immobile ^b
Servais (29)	3/M	Post aortic surgery	3 M	Paraplegia. Normal tactile sensation/proprioception. Urinary/faecal incontinence	Able to move toes/feet against gravity, no proximal lower limb movements.	Immobile ^b
Lewis (30)	0.3/M	Hypotension post surgery for neuroblastoma	Not specified	Not specified	Paraplegic and unable to move limbs against gravity	Immobile ^b

F/U (D/W/M/Y), Follow up (Days/Weeks/Months/Years); R, Right; L, Left.

^a Authors' case.^b Not clear from literature whether patient was able to use a wheelchair.

Figure 1. Sagittal T2-weighted Turbo Spine Echo images show abnormal high signal intensity involving the anterior half of the thoracic spinal cord extending from T5 to T12 consistent with infarction.

Figure 2. Axial T2-weighted Turbo Spine Echo images show abnormal high signal intensity involving the anterior half of the thoracic spinal cord extending from T5 to T12 consistent with infarction.

resonance imaging (MRI) of her spine showed a signal abnormality in the anterior half of the thoracic cord, extending from T5 to T12, consistent with anterior spinal artery infarction (Figures 1 and 2).

After a 6-month period of intensive rehabilitation, neurologic examination showed asymmetrical spastic paraparesis with Grade 4 muscle power in all groups. She was able to stand for transfer and walk during hydrotherapy. At 1-year follow-up, she was able to walk for short distances using a rollator device. Bladder control was still abnormal, and she needed daily intermittent urinary catheterization. No cause was identified for the anterior spinal artery infarction despite extensive investigations for cardiac causes (echocardiography and electrocardiography), autoimmune diseases (autoantibody testing for anti-nuclear antibodies, double-stranded DNA, extractable nuclear antigen screen including Ro, La, Sm, RNP, Scl-70, Jo-1, and complement studies), and coagulopathy (routine clotting screen and thrombophilia screen including antithrombin III activity, Factor V Leiden, protein S antigen, protein C antigen, activated protein C, activated protein C ratio, and prothrombin gene mutation). A follow-up MRI scan was not done because the patient declined this investigation.

DISCUSSION

The usual presentation of anterior spinal artery syndrome is an acute and painful myelopathy. Pain can be diffuse, radicular, or girdle-like and is usually severe. Bladder and

bowel control is impaired, and pain and temperature sensation below the lesion are lost, whereas vibration and position sense is intact because of the preservation of the posterior columns. Sometimes a partial syndrome is seen with the gray matter of the cord preferentially affected, and in this situation, sphincter control and sensory function are preserved. This entity has been increasingly recognized since MRI scanning of the spinal cord has become available. Before this, the diagnosis would have been made on clinical grounds or postmortem.

Anterior spinal artery syndrome is also rare in adults but is most commonly associated with aortic surgery, which accounts for the increased incidence (2). The causes of anterior spinal artery syndrome are usually grouped as either iatrogenic or noniatrogenic. In addition to the causes outlined in Table 1, anterior spinal artery syndrome has also been described in adults in association with atherosclerosis and diabetic arteriopathy (3), vasculitides, aortic disease (4), anemia, sickle cell disease, polycythemia, paradoxical embolism through a patent foramen ovale (5), atlanto-occipital dislocation (6), cervical spondylosis (7), cervical spinal trauma or sprain (8,9), recreational drugs such as cocaine (10), and some infections (tuberculosis, schistosomiasis, and *Neisseria meningitidis*) (11–13).

To our knowledge, anterior spinal artery syndrome has not been previously reported in a child or adult with Down syndrome. Patients with Down syndrome may have some risk factors for anterior spinal artery syndrome,

especially an embolic source if the patient has a congenital cardiac abnormality. In addition, Down syndrome is associated with other vasculopathies including Moyamoya disease and arterial dysplasia along with venous thrombosis (14,15). Despite extensive investigation, no risk factors were identified in our patient.

Management of anterior spinal artery syndrome in children is supportive, ideally with intensive multidisciplinary rehabilitation in a spinal unit (16). Given the rarity of the diagnosis, there is no consensus for any particular treatment approaches. One child with a genetic thrombotic disorder was treated with aspirin (81 mg/d), and the authors concluded that this may have improved the outcome (17). There is limited evidence for the acute use of steroids (methylprednisolone) in traumatic spinal cord injury (18); however, there is no evidence for their use in ischemic spinal cord infarction. As a preventative measure in adults requiring abdominal aortic aneurysm repair, an intercostal artery revascularization technique may have reduced the incidence of spinal cord ischemia (19).

The long-term prognosis is poorly described in children; from our review of the 20 reported cases, 2 children died acutely, and of the remainder, only 7 were mobile. Of these 7 children, all had neurologic sequelae and required assistance to walk. Similarly, the long-term outcome of anterior spinal artery syndrome in adults is poorly described, but 1 series of 44 patients with ischemic cord infarction from any cause has been reported. At follow-up (range: 1 day to 12 years, mean: 14.2 months), 8 (18%) had died, 9 (20%) showed no improvement in functional outcome, 10 (22%) were able to transfer, 12 (27%) were walking with support, and 5 (11%) were able to walk independently (20).

CONCLUSION

Despite its rarity, pediatricians should consider anterior spinal artery syndrome, especially in the child who presents with an acute, painful myelopathy. The clinical picture can be confusing because children may be uncooperative, and it may be difficult to conduct a reliable sensory examination. Because light touch sensation is preserved, the clinical diagnosis may not be suspected if only a cursory examination is undertaken. Diagnosis is confirmed with an MRI scan of the spinal cord, which clearly presents problems for hospitals without MRI facilities. Delay in diagnosis can lead to renal failure and other complications of acute myelopathy.

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