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Case Report

SPINAL ARTERIOVENOUS METAMERIC SYNDROME: ROLE OF PHYSIOTHERAPY: A CASE REPORT

Binita Lama *[1], Pallavi Wajapey [2], Ishwor Pyatha [3].

[1] Student, RV College of Physiotherapy, Bangalore, Karnataka, India.

[2] Assistant Professor, RV College of Physiotherapy, Bangalore, Karnataka, India.

[3] Student, RV College of Physiotherapy, Bangalore, Karnataka, India.

ABSTRACT

Background: Spinal arteriovenous metameric syndrome (SAMS), also known as Cobb syndrome is a rare metameric developmental disorder presenting as an extradural-intradural vascular malformation that involves bone, muscle, skin, spinal cord, and nerve roots.

Case Report: An 11-year-old girl presented with history of progressive paraparesis, lower extremity sensory loss, along with bowel and bladder incontinence who was diagnosed with Cobb syndrome. She underwent endovascular embolization of one arterial feeder and two metameric components. Post embolization patient experienced further decline in muscle strength. Physiotherapy regimen was implemented for 4-5 days per week for period of two months. The patient was discharged with a home exercise programme.

Result: At the time of discharge, muscle power was improved. Patient was able to walk with the help of walker.

Conclusion: Spinal metameric arteriovenous syndrome is a complex nonhereditary genetic vascular disorder associated with variety of neurologic deficits. Physiotherapy management will play a major role in minimizing disability hence improve clinical outcome.

KEY WORDS: Spinal Arteriovenous Metameric Syndrome, Progressive Paraparesis, Embolization, Physiotherapy.

Address for correspondence: Binita Lama, RV College of Physiotherapy, Bangalore, Karnataka, India. **E-Mail:** bntaso7@gmail.com

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INTRODUCTION

Spinal arteriovenous metameric syndrome (SAMS), also known as Cobb syndrome, is a rare embryonic metameric syndrome, in which cutaneous, muscular, and/or bony vascular lesions as well as paraspinal and/or spinal vascular lesions are found in the same metameres [1]. Cobb syndrome is typically diagnosed following the onset of neurological symptoms. Although this order is most commonly seen during late childhood, it may occur at any age. Onset of signs usually manifests over weeks to years, but a sudden onset of weakness with rapid progression has also been reported [2]. Neurological presentation may vary from monoparesis to sudden-onset paraplegia or quadriplegia. Bladder and bowel involvement is common and occurs as the disease progresses [3].

Cobb syndrome probably has an underreported prevalence, and the number of documented cases of Cobb syndrome in the literature is less than 50 [4]. Owing to the rarity of this syndrome, definitive management recommendations remain unclear. We report a case of spinal metameric AVM following embolization surgery.

CASE REPORT

An 11-year-old female was referred to the Neuro-Rehabilitation department of our college with the chief complains of inability to stand and walk along with left knee pain. She belonged to a medium socio-economic status and was doing her schooling from normal school. She was a second child of her family with normal full term delivery. Her mother had experienced ges tational hypertension which was managed medically. She had history of fits at age of one year which was managed with medications. When the patient was 10 years of age, she developed mid back pain radiating to the right knee and progressive weakness of both lower limbs leading to restriction in daily activities. Over the course of six months, she developed constipation and bladder incontinence.

The patient underwent an evaluation including MRI of the dorsolumbar spine and diagnostic spinal angiogram. MRI findings are consistent with extramedullary Dural arteriovenous malformations. Digital spinal angiogram revealed a perimedullary fistula feeding from left D8 and D10 intersegmental artery. Also a note is made of multiple paraspinal and epidural AVM component with feeders from the radicular branches of bilateral D8 to L2 intersegmental arteries with venous drainage through the dilated epidural and paravertebral veins into the Azygous and hemiazygous system and subsequently into the inferior venacava. Perimedullary fistula with nidus with feeder from left radiculomedullary artery of D11 was catheterized and fistulous component was embolized. Paraspinal metameric component at left D11 and D12 was embolized on 29/09/2016. No postoperative complication was present. The patient was admitted again 1.5 month after the procedure with complaints of inability to stand and walk.

Patient was advised for tab neurobion, tab baclofen and physiotherapy.

Examination

On observation, patient had a raised patch on D8-L1 dermatome that was warm on palpation. The patient had a lateral curvature with convexity on the right side. The attitude of the limbs revealed hips and knees in flexion. Higher mental functions and cranial nerve functions were intact. On further examination, neurological examination of both the upper limbs was normal. She had sensory deficit to touch, pin prick, tactile localization and tactile discrimination on lower limbs and trunk from dermatome level of D6 to S1. The patient had grade 2-/5, 2/5 strength on the hip, knee and ankle musculature of left and right lower limb respectively. Trunk strength was found to be reduced with grades 1/5, 1/5, 2/5 and 2+/5 for flexors, extensors, rotators and lateral flexors respectively. The muscle tone on bilateral upper and lower limbs was found to be grade 0/4 on modified Ashworth Scale.

However, the patient was under medication during the time of examination. Knee and ankle jerks were absent and plantar reflex was nonresponsive. There was no muscle wasting and fasciculation seen. She had no breathing difficulty. Muscle tightness of hamstring, hip flexors and gastro-soleus was significant bilaterally but more in left lower limb. Trunk control on sitting was fair, equilibrium reactions were restricted because of trunk and lower limb weakness. Functional independence measure score was 64/126, with motor and cognitive subtotal of 30/91 and 34/35 respectively. She had bowel and bladder incontinence.

Physiotherapy management: The initial physiotherapy goals were set to be patient and parent counselling, reduction of pain, reduction of tightness and progressive increase in muscle strength. Added to this, the physiotherapy long term goals were set to achieve weight bearing, standing with support and walking independently with help of walker.

The patient was put on a pain management programme, muscle re-education programme, stretching and exercises to improve trunk strength. This was carried out once a day, 5-6 days per week. The exercises performed were as follows:

Active and active assisted movement in gravity minimized plane for hip, knee and ankle muscles along with tapping on the muscle, isometric exercises for quadriceps, hamstring and gluteal muscle, moist hot bag with careful supervision followed by myofascial release of hamstring and calf, core muscle strengthening on Swiss ball, stretching of lateral spinal musculature of right

side. Rest period was provided in between the exercises to avoid fatigue.

After a period of two weeks following exercise were added along with the above exercises, sustained stretching of hamstring, calf and hip flexors, wall supported standing for 5 minutes with assistance of three therapists, one holding pelvis and other two pushing knees and shoulders back for postural reeducation. Walking with walker was done with the help of gaiters and in assistance of therapist for sitting to standing transition.

The family member was trained to provide sustained stretching, also advised to encourage the patient to use gaiters during night to prevent the knees from going into flexion. After a month of rehabilitation, patient was discharged on home exercise programme on their request. Home programme included sustained stretching of hamstring, calf and hip flexors, initiation of active movement against gravity for hip, knee and ankle, pelvic tilt, pelvic bridging, spinal rotation and walking with the help of support. Patient was asked to perform each exercise with 10 second hold and 10 repetitions twice a day. Family members was also advised to progress each exercise in terms of amount of assistance, number of repetitions and holding time. The family member was suggested for a follow up with neuro-surgeon. The family member was also suggested for a follow up with physiotherapy. Unfortunately, because the patient's location is in remote village out of Bangalore, we were not able to obtain a follow up.

Outcomes: At the time of discharge, muscle strength on the limbs was 3/5 and 3- for lower limbs and trunk respectively. Patient was able to walk with the help of walker with assistance required only during sitting to standing transition. Tightness of the hamstring and hip flexors was significantly reduced hence the attitude of limbs were improved.

DISCUSSION

SAMS was first described in 1915 by Stanly Cobb, who was a resident of Harvey Cushing at that time [5]. Thus, this syndrome has long been called Cobb syndrome.

In the process of vasculogenesis, endothelial cells derive from mesoderm, and the tunica media derives from the neural crest cells or mesodermal cells. These cells corporate to form vessels in the given metameres after migration to their own target regions. When embryological adverse events occur before the migration, more than two separate vascular malformations in the same metamere could be formed. This explains the pathogenesis of SAMS [6].

With consideration to neurological symptoms, cord ischemia due to steal syndrome and cord compression due to venous hypertension are the proposed mechanisms that would explain the myelopathy observed in unruptured spinal malformations in general, and in the large AVMs often described in Cobb syndrome [7].

Adult and young patients typically present with sudden onset of radicular pain in the lower extremities and associated numbness. Less commonly, sudden onset of weakness or rectal/and bladder dysfunction is the presenting symptom [8]. The clinical course may also progress over several years to paraplegia and sphincter dysfunction [7]. Our patient also presented with the sudden onset of back pain radiating to the right limb followed by progressive weakness of trunk and lower limbs.

With regard to the limited number of cases described in the literature, it is difficult to formulate a best possible therapeutic plan. The use of partial weight bearing treadmill training could have been implemented in the treatment setting. This was limited due to loss of follow up with the patient. Despite this, the embolization of the fistulous component and resection of residual AVM was still to be done.

CONCLUSION

Spinal metameric arteriovenous malformation, also known as Cobb syndrome or Cutaneomeningospinal angiomatosis, is a rare, non-inherited disorder characterized by a spinal AVM and a vascular skin lesion affecting the corresponding dermatome. Spinal involvement may include the vertebral bodies, dura mater, spinal cord, and nerve roots [9].

Our case report has documented the neurological examination of Cobb syndrome after partial embolization of the spinal metameric AVM, and the physiotherapy management with its out-

come. Reporting these rare conditions will help increase awareness about the condition and highlight the role of physiotherapy in minimizing disability and enabling these patients to be more independent in their functional activities thus improve their quality of life.

ABBREVIATIONS

SAMS - Spinal Arteriovenous Metameric Syndrome
AVM - Arteriovenous Malformations
MRI - Magnetic Resonance Imaging

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Conflicts of interest: None

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