CASE REPORT

COBB Syndrome Treated by Staged Intravascular Embolisation and Surgery

S Sayuthi, MBBCh.BAO*, J Moret, MD*, A Pany, MMed***, A Sobri, MMed***, M Shafie, MMed**, J Abdullah, Phd***

*Department of Neurosciences, **Department of Radiology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia, ***Service de Neuroradiologic Interventionnelle, Hospital de la Foundation Rothschild, 25-29Rue Manin, 75940 Paris Cedex 19, France

Summary

A 28-year old Malay man with evidence of an upper motor neuron cord lesion was diagnosed to have a C7 to T2 spinal arterio-venous malformation and associated cutaneous vascular lesion. He finally agreed for treatment after 5 years of progressive spastic right lower limb weakness leading to inability to mobilize. A two staged intravascular procedure was done followed by surgery with recovery of ASIA impairment scale grade B.

Key Words: Cobb syndrome,Spinal cord arteriovenous, Embolisation, Surgery

Introduction

Cobb syndrome better known as cutaneomeningospinal angiomatosis is defined by the presence of a cutaneous vascular lesion with corresponding spinal cord vascular malformation within a segment or more of the involved dermatome. Various treatments of the cutaneous lesions have been done ranging from Nd: Yag laser to corticosteroid therapy [1-2]. All cases of Cobb syndrome have so far been reported as rare in the literature and managed mainly by interventional methods [3]. Previous imaging such as Computed Tomography of the spine has been replaced with Magnetic Resonance Imaging to rule out other differential diagnoses and involvement of the brain. We report the first case of Cobb syndrome in the Asian literature which was managed by an interventional radiological procedure and surgery.

Case report

A 28- year old man presented with over a five year history of progressive right lower limb weakness and numbess over the whole of the right trunk and right lower limb. He was admitted for radiological evaluation of the same limb which became more spastic over the preceding 12 months that caused him to lose his work as a rubber-tapper. On examination the limb was spastic with power of 3/5 proximally and 1/5 distally. Reflexes were brisk with demonstratable clonus and upgoing plantar. This resulted in a circumduction gait. The pin prick and light touch sensation was lost over the right T1 dermatome and distally. Patient's bowel and bladder functions were intact. There was a cutaneous port wine discolouration over the back mainly over the right scapula reagion (Fig 1). This corresponded to the dermatomal distribution to his intraspinal lesion.

His spinal MRI showed serpinginous tubular signal void structures within the spinal canal on both T1WI and T2WI images. It extended from the foramen of magnum until the T2/T3 intervertebral disc anterior to the spinal cord suggestive of prominent draining veins to a similar signal void appearance within the spinal cord forming a mass from C7 until T2 level. This was a nidus of

This article was accepted: 27 October 2005
Corresponding Author: Sani Sayuthi, Department of Neurosciences, School of Medical Sciences, Universiti Sains Malaysia, Health Campus, 16150 Kubang Kerian, Kelantan

extensive spinal cord arteriovenous malformation 4cm in length (Fig 1). A spinal angiography revealed a type II spinal Arterio Venous Malformation (AVM) supplied by the right T6 and T8 intercostal artery.

He then underwent endovascular embolization of the AVM. The T6 and T8 intercostal artery was cannulated and the feeders were embolized by histoacryl glue mixed with lipiodol (Fig.2). Follow-up intravascular embolisation was done two months later followed by an emergency removal of the remainding nidus via a laminectomy of C6-Thoracic 1. A follow-up imaging showed no new vessel formation supplying the AVM.

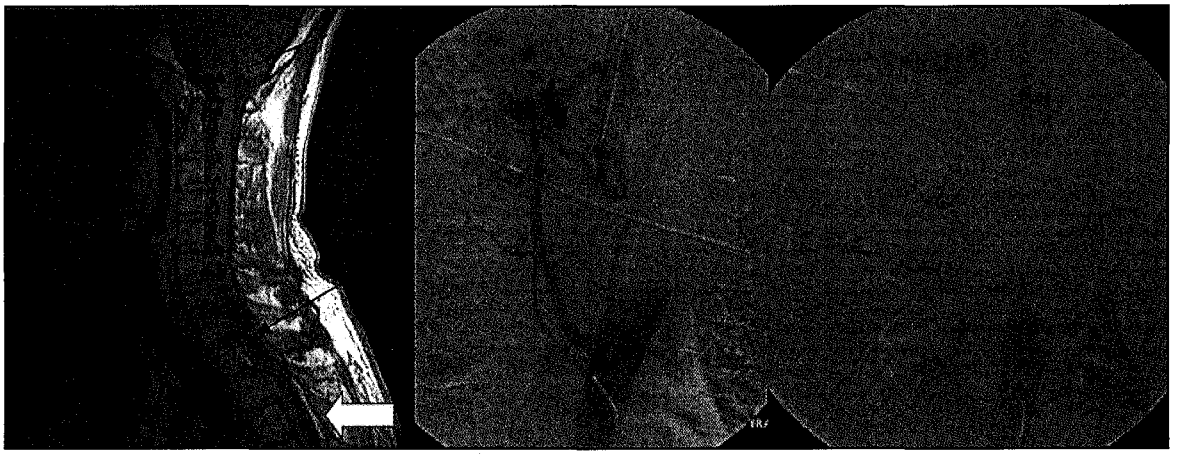

Fig 1: Serpinginous tubular signal void structures within the spinal canal on T1WI images. It is mostly anterior to the spinal cord until C6 level. Signal void appearance is seen within the spinal cord forming a mass from C7 until T2 (thick white arrow) suggestive of a nidus of an extensive spinal cord arteriovenous malformation. Angiogram demonstrating the blushing of the AVM (on the left) and repeat angiogram (on the right) after embolization.

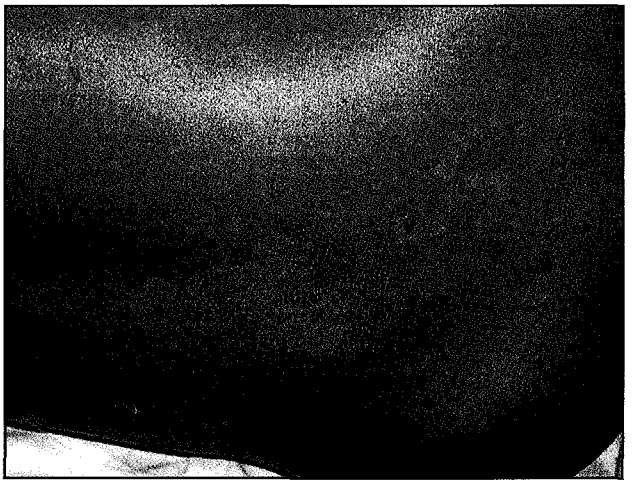

Fig 2: Cutanious lesion in the distribution of spinal AVM i.e. C7 until T6.

Discussion

Cobb syndrome is a rare, noninherited disorder that involves the association of spinal angiomas or arteriovenous malformations (AVM) with congenital, cutaneous vascular lesions in the same dermatome. Berenbruch first described the disorder in 1890, but it was not widely known until Cobb's report in 1915. The importance of this syndrome is the recognition that cutaneous vascular lesions may hint at an accompanying spinal cord angioma or AVM that may result in weakness or paralysis [1-3]. This high-flow capillary malformation on the posterior thorax area has associated with intra or perimedullary AVM with vertebral body involvement. The cutaneous lesion also known as nevus flammeus may give a tell tale sign when a patient comes with sudden onset of paraplegia or subarachnoid hemorrhage. The prevalence of Cobb syndrome is estimated to be approximately 12% in AVM patients and most of these cases end with progressive paraplegia associated with back pain if not treated when the patient reaches adolescence. Various treatments with pure decompression via a laminectomy have proved fatal. The differences in the management of this syndrome compared with other spinal AVM's is that Cobb syndrome is detected early because of its cutaneous manifestations which allow early diagnosis. More non-invasive treatment like radiotherapy has had no success. Our patient atypically presented with unilateral spasticity without bowel or bladder dysfunctions and his clinical findings were caused by a large nidus causing a myeolopathy at the junction of cervicothoracic area. In our patient straining during defeacation made the cutaneous lesion more prominent. At the current stage six months after follow up the patient has not shown any complications like subacute necrotic myelopathy better know as Foix-Alajouamine disease. The cutaneous lesions will be referred further for dermatological laser treatment. The patient is undergoing therapy for both his intrinsic hand movements as well as his right lower limb.

Acknowledgements

Prof. Jacques Moret from the Service de Neuroradiologie Interventionnelle, Hospital de la Fondation Ophtalmologique Adolphe de Rothschild, 75940 Paris Cedex 19, France was a visiting professor to the School of Medical Sciences and was directly involved in the initial embolisation procedure of this patient together with Dr Adam Pany and Dr Abdullah Sobri as well as Dr Shafiee Abdullah who is the interventional radiologist of the Department of Radiology, Universiti Sains Malaysia, Health Campus. The subsequent stages of embolisation were done both by Dr Shafie Abdullah and Dr Adam Pany. The final residual AVM was removed by the sixth co-author. The corresponding author thanks Professor Jacques Moret in the active involvement of the management of this patient.

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