

# SUPERIOR MESENTERIC ARTERY SYNDROME

Sana Noman,<sup>1</sup> Jawaid Iqbal,<sup>1</sup> Mukhtiar A. Memon<sup>2</sup>

<sup>1</sup> Department of Radiology, Darul Sehat Hospital, Karachi, Pakistan.

<sup>2</sup> Dow Institute of Radiology, Dow University of Health Sciences, Karachi, Pakistan.

PJR April - June 2016; 26(2): 114-116

## Introduction

Superior mesenteric artery syndrome is a rare disease occurring due to acute angulation of origin of superior mesenteric artery from aorta resulting in extrinsic compression on third part of duodenum. Its etiology is believed to be due to rapid weight loss resulting in decreased fatty tissue between aorta and proximal superior mesenteric artery. Patients present with abdominal pain and vomiting. We report a case of a 17 year old male with diagnosed Moya Moya disease, who has had left hemiparesis since 2 years and has been bed ridden for past 6 months due to repeated Neurological issues.

## Case Report

A 17 year old male presented to Emergency department with vomiting and abdominal discomfort for 3 days. He also complained of occasional post prandial abdominal discomfort for past 6 months. He was a known case of Moya Moya disease and has had left hemiparesis 2 years back. He has been bed ridden for past 6 months. He denied any significant weight loss.

On examination he was slightly tender in epigastric region. Bowel sounds were appreciated in all four quadrants. His laboratory tests were unremarkable. His endoscopy was performed the same day which failed as endoscope could not proceed beyond second part of duodenum. Esophagus, stomach and visualized duodenum were unremarkable.

His CT scan abdomen with contrast was performed the next day which showed marked distention of

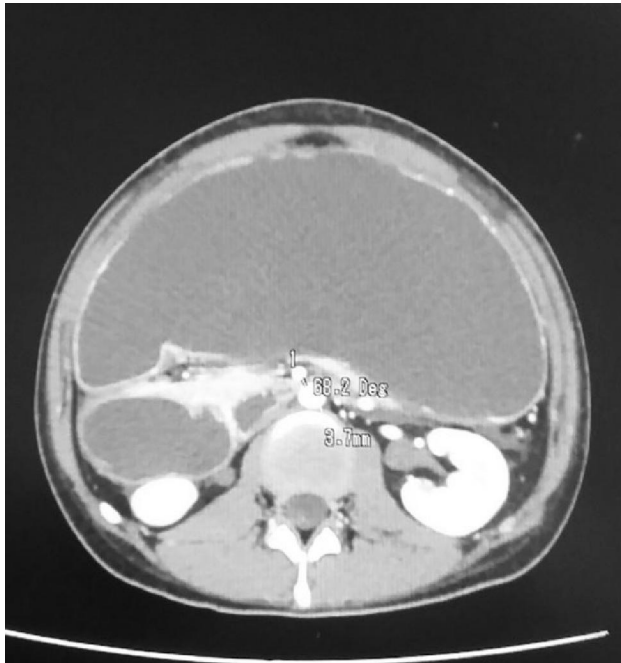
stomach and duodenum upto mid of third part. Sharp angulation at origin of superior mesenteric artery was seen with aortomesenteric angle of 12.7° (Fig. 1) and aortomesenteric distance of 3.7 mm (Fig 2). Normal aortomesenteric angle is 25-60° and aortomesenteric distance is reported to be 10-28 mm.<sup>1</sup> Bowel distal to the origin of superior mesenteric artery was collapsed.



Figure 1: Decreased Aortomesenteric angle, measuring 12.7 degree.

**Correspondence** : Dr. Sana Noman  
Department of Radiology,  
Darul Sehat Hospital,  
Karachi, Pakistan.  
Email: drsananoman@gmail.com

Submitted 8 September 2015, Accepted 8 October 2015



**Figure 2:** Decreased Aortomesenteric distance.

## Discussion

Superior Mesenteric Artery Syndrome also known as Wilkie's syndrome is a rare acquired vascular compression disorder first described in 1861 by Von Rokitanski<sup>2</sup> and later by Wilkie in a case series of 64 patients.<sup>3</sup> Patients usually present with signs and symptoms of upper GI obstruction with abdominal pain.

In this disease patient becomes symptomatic when third part of duodenum gets trapped between proximal superior mesenteric artery and aorta. This results in obstruction proximal to the mid of third part of duodenum. Normally, fat and lymphatic around SMA prevent its compression effect on duodenum in healthy individuals.<sup>4</sup> The abdominal pain in these patients is usually post prandial and it may relieve in prone and left lateral position.

The main etiological factor resulting in SMA syndrome is believed to be rapid weight loss, severe trauma, burns, anorexia nervosa or prolonged bed rest.<sup>4,5,6</sup> No definite direct relation between moya moya disease and superior artery mesenteric syndrome has been reported till.

As per our knowledge this is the first reported case

of patient having moya moya disease and SMA syndrome. We believe that our patient developed this pathology due to prolonged bed rest as a result of his neurological deficit.

Diagnostic modalities for this syndrome include upper GI contrast study, colour Doppler study and CT scan abdomen with contrast. Contrast enhanced CT scan has the advantage of providing assessment of entire abdominal cavity including accurate anatomical details.<sup>7</sup>

This disease is diagnosed on two parameters. First is the angle of superior mesenteric artery at its origin from aorta - the Aortomesenteric angle (AO angle) and the other is distance between Proximal Superior mesenteric artery and aorta - the aortomesenteric distance (AO distance). Normal AO angle is 25- 60° and AO distance is 10-28 mm. Both parameters should be decreased to diagnose this disease. Cut-off values of SMA syndrome are reported to be aortome-senteric angle of 22° (42.8% sensitivity and 100% specificity) and aortomesenteric distance of 8 mm (100% sensitivity and specificity).<sup>7</sup>

Management of SMA syndrome ranges from conservative to surgical approach. Initial treatment in acute settings is usually conservative which includes gastric decompression through Nasogastric tube and mobilization including change in posture to prone or lateral decubitus position.<sup>5</sup> Both enteral jejunal feeding and total parenteral nutrition have been found useful for increasing body weight and promoting restoration of the retroperitoneal fatty tissue with concomitant increase in the aortomesenteric angle and distance.<sup>8</sup> Surgical options, including open or laparoscopic duodenojejunostomy, are reserved for patients where conservative measures fail.<sup>9</sup>

## Conclusion

Superior mesenteric artery syndrome is a rare disease resulting in abdominal pain and vomiting. Our patient was a known case of moya moya disease since 2009 and had neurological deficit since then. He presented with abdominal pain and vomiting. To the best of our knowledge there is no known association of these two diseases and we believe that reason of SMA syndrome in our patient was prolonged bed rest.

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