ABSTRACT

The knowledge of anatomical variations in superior mesenteric artery is of importance to surgeons and radiologists while performing complicated procedures. Superior mesenteric artery syndrome is a rare life-threatening gastrointestinal disorder characterized by compression of the third part of Duodenum by Abdominal Aorta and the overlying Superior Mesenteric Artery. Superior mesenteric artery syndrome is triggered when there is narrowing of the mesenteric angle and shortening of the distance.

We report a case of SMA syndrome which sharply exemplifies its clinical and anatomical features which was identified during routine radiological studies of the Dept. of Radiology, KIMS, Narketpally. The aetiology, presentation, diagnosis and management of this unusual condition are discussed.

Key words: Superior mesenteric artery (SMA), Abdominal Aorta (AA), Duodenum (D), Computed Tomography (CT), Nasogastric tube (NG tube).

INTRODUCTION

Superior Mesenteric Artery syndrome was first described in 1861 by Carl Freiherr Von Rokitansky in victims at autopsy. Intestinal obstruction of the duodenum by entrapment between AA and SMA is an uncommon cause of megaduodenum.

Also known as Cast Syndrome, Wilkie’s Syndrome, Arteriomesenteric Duodenal Obstruction, Chronic Duodenal Ileus and Mesenteric root syndrome. Only 500 cases of SMA Syndrome in the history of English language medical literature have been reported.

Superior mesenteric artery arises at a 20-30 degree angle from AA opposite the middle of first lumbar vertebra. It supplies the larger part of the intestine from lower part of the Duodenum to the junction between the right 2/3rd and left 1/3rd of the transverse colon. The diameter of the artery is 8-10mm and it has an unifurcated course 3-6 cm before giving origin to inferior pancreaticoduodenal arterial. The third portion of duodenum passes between the AA and SMA at the level of L3. A typical angle created by these 2 vessels is 25-60 degrees. This angle is maintained by the mesenteric “fat pad” and the distance between SMA and AA is 10-28mm. The syndrome is typically caused by reduced angle and the distance b/w SMA & AA.

The incidence is 0.013-0.3% of upper gastrointestinal tract barium studies. SMA syndrome is estimated to have a mortality rate of 1 in 3. No racial differences have been identified. More females are affected with the ratio of 2:1 (F: M). The SMA syndrome usually occurs in older children and adolescents and patients being aged 10-30 years are commonly affected.

Types: SMA syndrome can be presented as two forms:

1. Chronic/congenital
2. Acute/induced
**Chronic / congenital form**
Predominantly have a life long history of abdominal complaints depending upon the duodenal compression. The individual is thin built. Clinically patient has unusually high insertion of the ligament of Trietz, low origin of SMA and Intestinal malrotation around SMA.

**Acute or Induced form**
Develops rapidly after traumatic incidents that forcibly hyper-extend the SMA. It may be due to spinal cord injury, following surgery for scoliosis & left nephrectomy etc.

**Case Presentation**
A 13 year old male admitted in Kamineni Hospital, Narketpally, Nalgonda (Dt.) in the month of August, 2011 Presented with the following complaints.
1. Vomittings since 2 months. 3-4 episodes of post prandial vomitings per day
2. Pain abdomen since 2 months, colicky in nature relieved on lying down on his left lateral position.

**Clinical Examination:**
Thin built and anaemic. CVS, RS & CNS - with in normal limits.
P/A- Soft, tenderness present at the epigastric and umbilical regions with mild guarding and rigidity present. Suspected as Gastric outlet obstruction or acute gastric dilation

**Investigations done:**
1. **Ultrasonography:**
Dilated proximal duodenum and stomach with normal vascularity of walls of duodenum and stomach and distal duodenum was collapsed.

2. **Upper GI Endoscopy:**
Bile stained and dilatation of the second part of duodenum indicating distal obstruction.

3. HIV – Negative, HBS Ag- Negative and Blood group – O positive
4. **Contrast CT scan abdomen done** (Fig I-VI).

**Diagnosis & Treatment:**
This case was diagnosed as chronic congenital form of SMA syndrome.

Duodenojejunostomy, bypassing the compression caused by AA and SMA was done.

**DISCUSSION**
Vascular compression of the duodenum can present at any age, but the majority of patients are young adults. Females are affected more often than males.

Etiological factors included structural and / or acquired factors. Incomplete rotation of the duodenum, abnormally high insertion and shortness of the ligament of Treitz, fast weight loss, anorexia nervosa, scoliosis and body casting were among these factors. In its acute or chronic forms, the syndrome becomes clinically manifest with predominantly postprandial epigastric pain, sense of repletion, and vomiting.

The third part of the duodenum is vulnerable to become pinched in between the SMA anteriorly and the AA and vertebral column posteriorly.

The duodenum is suspended by the ligament of Treitz in the vascular angle between SMA and AA. The site of the suspensory ligament, attachment and structure may differ from individual to individual.

It has been postulated that if a person loses weight rapidly, as a sequelae to debilitating injury or an eating disorder or gains height rapidly with out a concomitant increase in weight, the fatty cushion around SMA diminishes and the angle between the SMA and the AA decreases resulting in the occlusion of the third part of the duodenum. Other factors which contribute SMA Syndrome include lordosis, body cast in the treatment of scoliosis or vertebral fractures.

Conventional barium studies play an important role in the diagnosis of this syndrome. Lukes and associates have shown that hypotonic duodenography is more accurate in distinguishing between normal and abnormal states, especially when the barium meal studies have given negative results. Hypotonic Duodenography combined with simultaneous SMA arteriography has been observed as more accurate investigation. Dynamic thin section CT with sagittal reconstruction is an excellent imaging modality in diagnosing SMA syndrome.
The familial clustering of SMA syndrome was observed in a family raises a possibility of genetic predisposition to this syndrome was published in 1990 by Ortiz.

SMA Syndrome may be managed conservatively, though surgical treatment is sometimes required. Gastric dilatation occurs early in vascular compression of the duodenum and nasogastric suction accompanied by alterations in posture such as placing the patient into the prone position may abort full-blown duodenal obstruction.

Others favour aggressive nutritional support using nasojejunal feeding past the point of obstruction as the primary method of nutrition support.

For patients in whom conservative methods fail, several surgical procedures have been described that is Strong's procedure. When the Strong's operation was difficult, Staveley's duodenojejunostomy was preferred and majority of the results were excellent.

**The present study** is a male patient aged 13 years with chronic congenital form of SMA syndrome diagnosed radiologically (figure I- VI). In our study we observed a reduction in angle and distance between SMA and AA. The aortomesenteric angle is 25.86(Fig III) and aortomesenteric distance is 4.8mm (Fig V), whereas the normal values have been reported to be 25 to 60 and 10 to 28mm respectively. Measurements of the angle and distance between the SMA and AA have been taken using the images of CT scan Abdomen (fig- III). We could also be able to demonstrate the dilatation of the second part of Duodenum and abrupt cut off of the third part of duodenum in Barium meal follow through (fig-I) and Contrast enhanced CT Scan abdomen (fig-II,III,IV). Therefore we could correlate the radiological and clinical findings appropriately. Duodenojejunostomy has been done and the patient was found to be improving.
CONCLUSION

The present study conceptualizes a fact that the case report is chronic congenital SMA syndrome which is diagnosed radiologically.

We being Anatomists would like to highlight the unusual arterial variations like SMA syndrome which are diagnosed only by radiological studies and not seen in routine cadaveric dissections.

The authors declare that they have no conflict of interest.

REFERENCES


