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SMA SYNDROME:- ABDOMINAL ANGINA IN EMERGENCY ROOM

General Surgery

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ABSTRACT

22 year female, Presented with complaints of epigastric abdominal pain & persistent vomiting for 4-5 days. Multiple hospital admissions over the past 2-3 years for the same with H/O weight loss of 6-8 kgs. All the laboratory and radiological investigations turned out to be normal until a high degree of suspicion pointed to a rare surgical condition. She was managed surgically and discharged. She recovered with complete resolution of symptoms over time.

KEYWORDS

Clinical Presentation:-
A 22 year female presented to the Emergency department with complaints of epigastric, abdominal pain & persistent vomiting for 4-5 days. Past history of similar complaints for 2-3 years .She had recurrent episodes of abdominal pain, associated with vomiting for which she had multiple hospital admissions in the past.She has weight loss of 6-8 kgs over last 6 months.

Past Medical History
She was diagnosed as having chronic gastritis and started on PPI for the same since last 6 months.

No relevant past medical or surgical history.

Medications
The patient was taking Tablet Pantoprazole(40mg) once daily before breakfast.

Allergies
The patient has no known drug allergies.

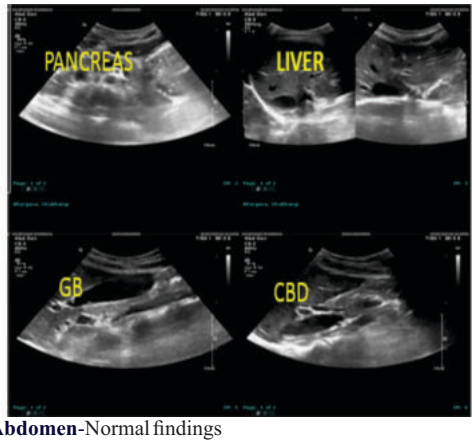
Physical exam and ancillary studies:-
Vital signs:
Temperature-97.4˚F.
BP-104/76 mmHg
Pulse-114 bpm,
Respiratory rate-20/min
Oxygen saturation-98% on room air.

General: The patient is conscious, oriented and nauseous looking.

Head,Eye,Ear,Nose,Throat Examination: No abnormal findings noted

Lungs: Bilateral Air entry is equal. There was no added sounds.

Cardiovascular: The patient's heart beat is regular.



Ultrasound Abdomen-Normal findings

Abdomen: The patient's abdomen is soft, Tenderness is present over epigastric and left hypochondriac regions.

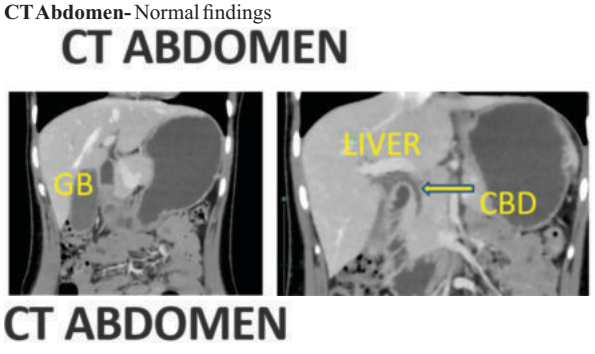
Extremity: No abnormality noted.

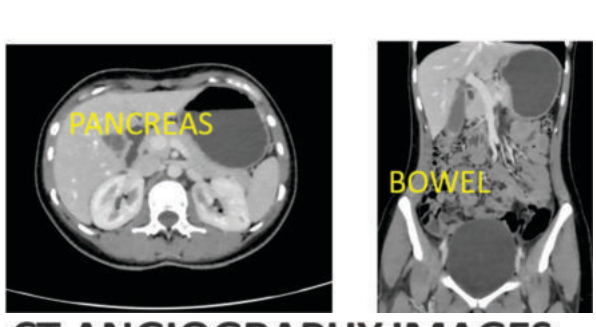
Neurologic: The patient has a normal neurologic exam.

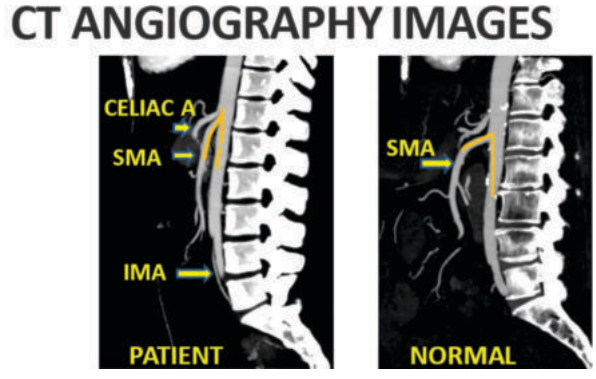
Pertinent labs: All of the patient's lab values are within normal limits.

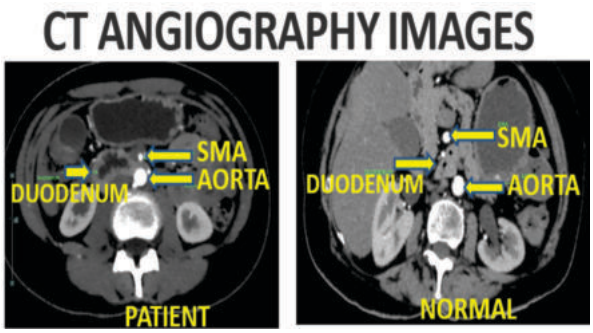
Radiographs: The patient's chest X-ray, Abdominal sonography, CT Abdomen came out to be normal.

CT Abdomen- Normal findings









Clinical course in the ED:-

The patient was managed in the ED conservatively.Pantoprazole infusion was started at 8 mg/hr.

She was planned for Upper GI endoscopy which had normal findings.

She was admitted to surgical ward and was planned for CT angiography of abdomen with oral and IV contrast after getting the serum creatinine report.

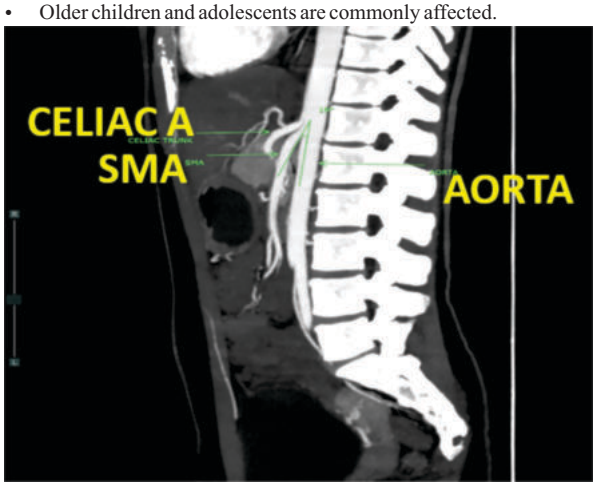
DIAGNOSIS:-

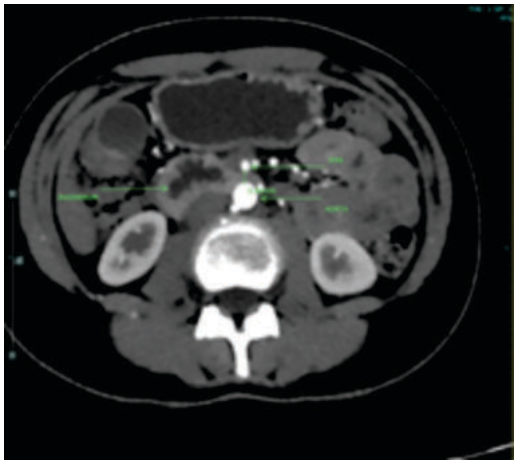
The patient was diagnosed as having a rare congenital condition, **Superior mesenteric artery syndrome** after complete workup.

DISCUSSION:-

Superior Mesenteric Artery syndrome

- Also known as Wilkie syndrome, cast syndrome or aorto-mesenteric duodenal compression syndrome.
- Rare acquired vascular compression disorder.
- Acute angulation of the superior mesenteric artery results in compression of third part of duodenum, leading to obstruction.
- Females > Males.
- Older children and adolescents are commonly affected.





- Fat and lymphatic tissues around the SMA provide protection to the duodenum against compression
- Under conditions of severe weight loss, this cushion around SMA is diminished, causing angulation and reduction in distance between aorta and SMA.

Associated with:-

- Anorexia nervosa
- Malabsorption
- Hypercatabolic states

(burns, major surgery and malignancy)

Signs and Symptoms:-

Signs and symptoms include early satiety, nausea, vomiting, extreme "stabbing" postprandial abdominal pain (due to both the duodenal compression and the compensatory reversed peristalsis), abdominal distention/distortion, burping, external hypersensitivity or tenderness of the abdominal area, reflux, and heartburn.

- In infants, feeding difficulties and poor weight gain are also frequent symptoms.In some cases of SMA syndrome, severe malnutrition accompanying spontaneous wasting may occur.
- This, in turn, increases the duodenal compression, which worsens the underlying cause, creating a cycle of worsening symptoms.
- Fear of eating is commonly seen among those with the chronic form of SMA syndrome.
- For many, symptoms are partially relieved when in the left lateral decubitus or knee-to-chest position, or in the prone (face down) position.
- A Hayes maneuver, which corresponds to applying pressure below the umbilicus in cephalad and dorsal direction, elevates the root of the SMA, also slightly easing the constriction.
- Symptoms can be aggravated when leaning to the right or taking a face up position.

Causes:-

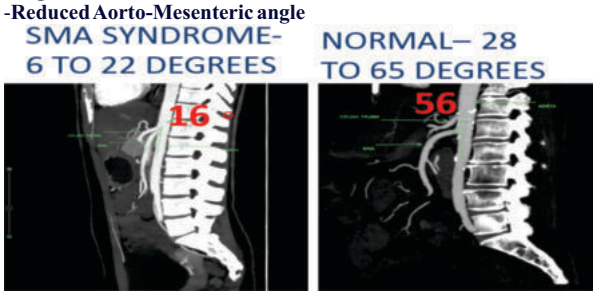
- Retroperitoneal fat and lymphatic tissue normally serve as a cushion for the duodenum, protecting it from compression by the SMA. SMA syndrome is thus triggered by any condition involving an insubstantial cushion and narrow mesenteric angle.
- SMA syndrome can present in two forms: chronic/congenital or acute/induced.
- Patients with the chronic, congenital form of SMA syndrome predominantly have a lengthy or even lifelong history of abdominal complaints with intermittent exacerbations depending on the degree of duodenal compression.

Risk factors include anatomic characteristics such as:

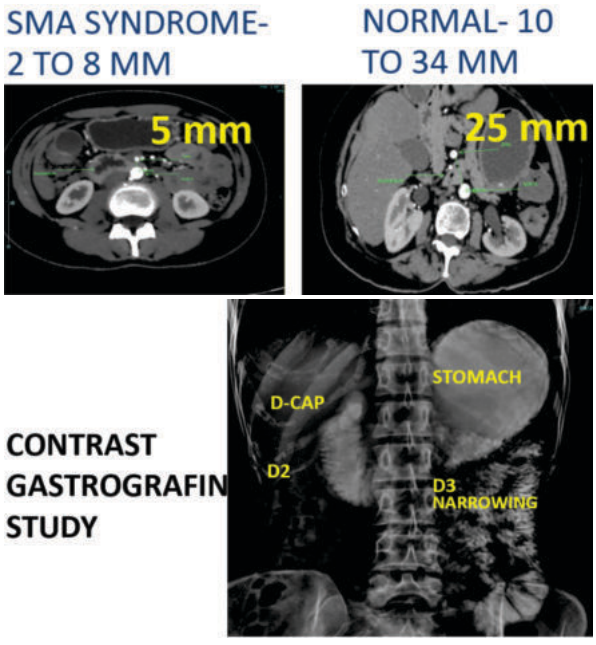
- Asthenic (very thin or "lanky") body build, an unusually high insertion of the duodenum at the ligament of Treitz, a particularly low origin of the SMA, or intestinal malrotation around an axis formed by the SMA.
- Predisposition is easily aggravated by any of the following: poor motility of the digestive tract, retroperitional tumors, loss of appetite, malabsorption, cachexia, exaggerated lumbar lordosis, visceroptosis, abdominal wall laxity, peritoneal adhesions, abdominal trauma, rapid linear adolescent growth spurt, weight loss, starvation, catabolic states (as with cancer and burns), and history of neurological injury.
- The acute form of SMA syndrome develops rapidly after traumatic incidents that forcibly hyper-extend the SMA across the duodenum, inducing the obstruction, or sudden weight loss for any reason. Causes include prolonged supine bed rest, scoliosis surgery, left nephrectomy, ileo-anal pouch surgery.
- SMA syndrome is usually difficult to differentiate with anorexia nervosa. Patients with SMA syndrome can be hindered restoring weight in those with anorexia nervosa

Diagnostic criteria:-

-Reduced Aorto-Mesenteric angle



Reduced Aorto-Mesenteric Distance-



Showing Narrowing Of Third Part Of Duodenum.

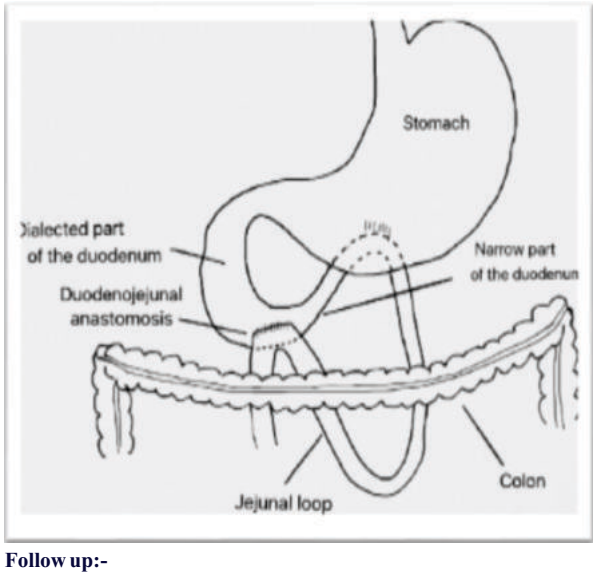
Management:-

Medical management-

- Decompression of stomach and duodenum with NG tube.
- Correction of nutritional and electrolyte deficiencies.
- Enteral feeding with nasoj-ejunal tube past the point of compression.
- Oral feeding if tolerated- small frequent meals to be given.
- Posture therapy- patient encouraged for lying in left lateral decubitus position.
- Aim is to improve nutrition & help build up the fat cushion between the SMA and Aorta.

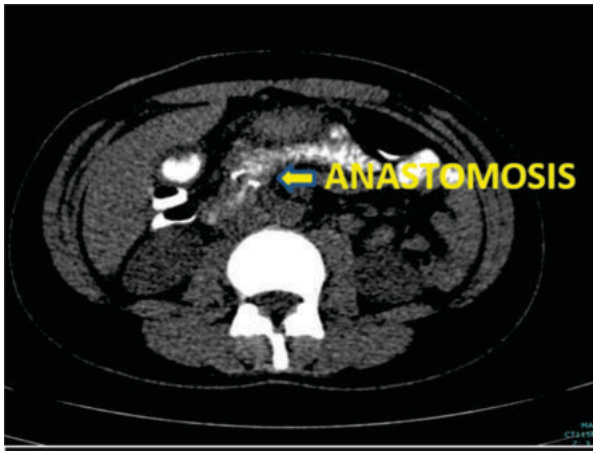
Surgical Management:-

Laparoscopic Duodenojejunostomy



Follow up:-

- Patient did not benefit much from conservative treatment.
- Required surgery –Duodenojejunostomy.
- In the immediate post-operative period , patient had an episode of abdominal pain, vomiting and loose motions which responded well to conservative management.
- Repeat CT was done to rule out any anastomotic leak/ stricture.
- Post this episode, at 6 months follow-up , patient is doing well and there is no recurrence of symptoms post-surgery.



Prognosis:-

Delay in the diagnosis of SMA syndrome can result –

- Fatal catabolysis (advanced malnutrition), dehydration, electrolyte abnormalities, hypokalemia, acute gastric rupture or intestinal perforation (from prolonged mesenteric ischemia), gastric distention, spontaneous upper gastrointestinal bleeding, hypovolemic shock, and aspiration pneumonia.
- A 1-in-3 mortality rate for Superior Mesenteric Artery syndrome has been quoted by a small number of sources.However, after extensive research, original data establishing this mortality rate has not been found, indicating that the number is likely to be unreliable.
- While research establishing an official mortality rate may not exist, two recent studies of SMA syndrome patients, one published in 2006 looking at 22 cases and one in 2012 looking at 80 cases,show mortality rates of 0% and 6.3%,respectively.
- According to the doctors in one of these studies, the expected outcome for SMA syndrome treatment is generally considered to be excellent.

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