

Case Study

**MASSIVE HEMATEMESIS FROM GASTRIC DIEULAFOY'S
LESION, surgery as a curative option when lack of access to
expertise.**

ABSTRACT:

DIEULAFOY'S LESION IS ACUTE GI (GASTROINTESTINAL) BLEEDING DISORDER, A RARE BUT LIFE THREATENING DUE TO RECURRENT BLEEDING. THE AFFECTED ARTERIOLE CAN PROTRUDE THROUGH A SMALL DEFECT IN MUCOSA, BECOME SUSCEPTIBLE TO EVEN MECHANICAL TRAUMA OF A MINOR LEVEL AND GRADUALLY ERODES INTO THE LUMEN TO CAUSE SEVERE ACUTE GI BLEEDING.

40 YEARS OLD MARRIED WOMAN WHO WAS ADMITTED WITH A HISTORY OF MASSIVE HEMATEMESIS FOR 17 DAYS. SERIES OF 3 UPPER GI ENDOSCOPIES WERE DONE AND PATIENT WAS DIAGNOSED WITH DIEULAFOY'S LESION. 2 TRIALS OF LOCAL EPINEPHRINE VIA EGD FAILED TO STOP HEMATEMESIS, HENCE SURGICAL CONSULT WAS REFERRED. EXPLORATORY LAPAROTOMY WAS DONE. A LESION WAS FOUND NEAR GREATER CURVATURE WHICH WAS LIGATED.

SURGICAL INTERVENTION WAS SELECTED ON BASIS OF LESION'S PRESENTATION AND CLINICAL ASPECTS OF PATIENT. SURGICAL INTERVENTION REMAINS A BETTER CHOICE IN CASE OF MASSIVE HEMATEMESIS IN A SETTING WHERE THERE IS LESS ACCESS TO AVAILABILITY OF EXPERTISE.

Keywords:

Dieulafoy's lesion, Vascular Malformation, Upper GI bleeding,

Introduction:

DIEULAFOY'S LESION IS A FAIRLY UNCOMMON DISEASE WHICH REPRESENTS AN IMPORTANT ETIOLOGY OF ACUTE GASTROINTESTINAL (GI) BLEEDING BECAUSE OF ITS TENDENCY TO CAUSE MASSIVE, LIFE-THREATENING, AND RECURRENT BLEEDING [1] . IT IS A SUB MUCOSAL ARTERIOLE, WITH THE ABILITY TO MAINTAIN AN ABNORMALLY LARGE CALIBER OF APPROXIMATELY 1-3 MM [2]. DIEULAFOY'S LESIONS ARE TYPICALLY PRESENTED WITH SEVERE, ACTIVE GASTROINTESTINAL BLEEDING. IT IS MOSTLY FOUND IN UPPER GI TRACT. ALMOST 75% OF LESIONS ARE FOUND IN THE STOMACH [3], THE AFFECTED ARTERY CAN PROTRUDE THROUGH A SMALL DEFECT IN MUCOSA, AND CAN BECOME SUSCEPTIBLE TO MECHANICAL TRAUMA OF EVEN A MINOR LEVEL (E.G., PASSAGE OF FOOD BOLUS IN STOMACH OR SOLID STOOL IN COLON), AND GRADUALLY ERODES INTO THE LUMEN TO CAUSE SEVERE ACUTE GI BLEEDING [4]. HOMEOSTATIC THERAPY IS PARAMOUNT DUE TO SEVERITY OF BLEEDING FROM DIEULAFOY'S LESION. THE BLEEDING SHOULD BE CONTROLLED IMMEDIATELY BY NONSURGICAL AND SURGICAL INTERVENTION, OTHERWISE MORBIDITY AND MORTALITY IS VERY HIGH [5].

DIEULAFOY LESION WAS FIRST REPORTED BY GALLARD [6] IN 1884; BUT IT WAS DESCRIBED ACCURATELY 40 YEARS LATER BY GEORGE DIEULAFOY A FRENCH SURGEON [7], WHO FOUND GI HEMORRHAGE OF FATAL KIND IN 3 YOUNG ASYMPTOMATIC MALE PATIENTS HE CALLED THEM AS "EXELCERATIO SIMPLEX".

DIEULAFOY'S LESION CAN BE RELATIVELY RARE BUT IS MOSTLY UNDER-RECOGNIZED ALTHOUGH IT ACCOUNTS FOR 1-2% OF ACUTE GI BLEEDING.

THE DISEASE HAS BEEN GIVEN MANY NAMES, SOME OF THE ALTERNATIVE NAMES FOR 'GASTRIC ARTERIOSCLEROSIS','CALIBRE PERSISTENT ARTERY','CIRSOID ANEURYSYM' AND 'SUB MUCOSAL ARTERIAL MALFORMATION' [8]. ONE OF THE SPECIAL FEATURES OF THE DIEULAFOY'S LESION IS THAT THEY MAINTAIN AN ABNORMALLY LARGE CALIBER ARTERIOLE OF APPROXIMATELY 1 TO 3 MM (10 TIMES LARGER THAN NORMAL CALIBER OF SUCH SUB MUCOSAL VESSELS), DESPITE THEIR PERIPHERAL, SUB MUCOSAL LOCATION. DIEULAFOY'S LESIONS ARE TYPICALLY PRESENTED WITH SEVERE, ACTIVE GASTROINTESTINAL BLEEDING; HENCE THE NAME 'CALIBER PERSISTENT ARTERY'.

DIEULAFOY'S LESION IS A VERY RARE DISEASE, REPRESENTS AN ARCHAEOLOGY OF ACUTE GASTROINTESTINAL BLEEDING BECAUSE OF ITS TENDENCY TO CAUSE MASSIVE, LIFE THREATENING AND RECURRENT BLEEDING; AND ITS ACQUIESCENCE TO ENDOSCOPIC THERAPY.

IT IS MOSTLY FOUND TO CAUSE UPPER GI BLEEDING BUT CAN ALSO CAUSE MIDDLE GI BLEEDING AND RARELY CAUSE LOWER GI BLEEDING.

ALMOST 75% OF LESIONS ARE FOUND IN THE STOMACH USUALLY CLOSE TO THE GASTRO ESOPHAGEAL JUNCTION, PROXIMAL STOMACH (6CM FROM GASTRO ESOPHAGEAL JUNCTION) OR ALONG THE LESSER GASTRIC CURVE; BUT THE LESIONS CAN ALSO BE FOUND IN DUODENUM OR ESOPHAGUS.

DIEULAFOY'S LESION IS THE CAUSE OF ALMOST 1.5% OF ACUTE UPPER GI BLEEDING [9,10], AND IS RESPONSIBLE FOR ALMOST 3.5% OF JEJUNOILEAL GI BLEEDING [11].

THE AFFECTED ARTERIOLE CAN PROTRUDE THROUGH A SMALL DEFECT IN MUCOSA, BECOME SUSCEPTIBLE TO EVEN MECHANICAL TRAUMA OF A MINOR LEVEL (E.G., PASSAGE OF FOOD BOLUS IN STOMACH A SOLID STOOL IN COLON), AND GRADUALLY ERODES INTO THE LUMEN TO CAUSE SEVERAL ACUTE GI BLEEDINGS. EVERY ARTERIAL PULSATION TRANSMITS THE MECHANICAL PRESSURE THAT CAN CAUSE TRAUMA TO THE FRAGILE, THIN LAYER OF MUCOSA PRESENT OVER THE VESSEL. ON THE OTHER HAND, FLOW THROUGH THE ARTERY THAT HAS BEEN ENLARGED ALREADY, MAY CAUSE HYPO PERFUSION, ISCHEMIA AND EROSION OF OVERLYING MUCOSA FROM SHUNTING AND REDISTRIBUTION OF BLOOD PERFUSION. THIS "VASCULAR STEAL" PHENOMENON (HYPOTHESIS) HAS A RESEMBLANCE TO THAT WHICH SOMETIMES PRODUCES PALE MUCOSAL HALO THAT SOMETIMES SURROUNDS ANGIODYSPLASIA.

THE LESION IS FOUND TO BE MORE COMMON IN MALES THAN FEMALES WITH A SEX RATIO OF 2:1. THE SPECIFIC AGE AT WHICH THIS DISEASE CAN OCCUR IS NOT PRECISE BUT MOST OF THE CASES REPORTED ARE OF PEOPLE IN THE 6TH OR 7TH DECADES OF THEIR LIVES [9]

PATIENTS SUFFERING FROM THIS DISEASE ARE OFTEN FOUND TO BE HAVING MULTIPLE COMORBIDITIES LIKE CARDIOVASCULAR DISEASES, HYPERTENSION, CHRONIC RENAL INSUFFICIENCY AND DIABETES. THESE PATIENTS ARE NOT ADMINISTERED NONSTEROIDAL INFLAMMATORY DRUGS BECAUSE THESE DRUGS PROMOTE BLEEDING FROM DIEULAFOY'S LESIONS UNDERNEATH, WHICH CAN RESULT IN CLINICAL DETECTION.

THE PATIENTS PRESENTED WITH THIS DISEASE USUALLY SHOW NO SYMPTOMS, BEFORE PRESENTED WITH ACUTE EXCESSIVE GI BLEEDING, WHICH CAN BE DISPLAYED AS HEMATEMESIS, MELENA OR HEMATOCHYZIA [9,12]. ABOUT HALF OF THE PATIENTS ARE PRESENTED WITH BOTH HEMATEMESIS AND MELENA [8]. PATIENTS FOUND WITH COLONIC DIEULAFOY'S LESION ARE USUALLY PRESENTED WITH PROFUSE BRIGHT RED BLOOD PER RECTUM. THE BLEEDING IS TYPICALLY SEVERE OWING TO THE ARTERIAL NATURE OF THE BLEEDING AND THE TYPICAL ENLARGED ARTERIAL VESSEL.

THESE LESIONS CAN ALSO BE A CAUSE OF OBSCURE GI BLEEDING BUT INTERMITTENT BLEEDING IT MAY TYPICALLY INVOLVES UNUSUAL BLEEDING SITES IN GI THAT MAY BE RELATIVELY INACCESSIBLE TO BASIC ENDOSCOPY SUCH AS JEJUNUM, ILEUM; THE LESIONS ARE MOSTLY SMALL AND INCONSPICUOUS DESPITE REPETITIVE USE OF STANDARD DIAGNOSTIC TECHNIQUES. THE SYMPTOMS MAY MIMIC OTHER DISEASES IN THE SETTING OF ACUTE GI BLEEDING. FOR EXAMPLE, TWO RECENT REPORTS FROM JAPAN SHOW PATIENTS WHOSE INITIAL CLINICAL PRESENTATION AND ENDOSCOPIC FINDINGS SUGGESTED THAT THEY WERE HAVING GASTRIC DIEULAFOY'S LESIONS BUT WHO WERE SUBSEQUENTLY DIAGNOSED WITH GI STROMAL TUMORS SO DIEULAFOY'S LESIONS ARE NOT ASSOCIATED WITH OTHER VASCULAR LESIONS OF GI LIKE ANGIODYSPLASIA OR HEMANGIOMAS.

THE FIRST DIAGNOSTIC TEST PERFORMED FOR A UPPER GI BLEEDING LIKE DIEULAFOY'S LESION IS ESOPHAGOGASTRODUODENOSCOPY (EGD). IF UPPER GI ENDOSCOPY IS UNABLE TO LOCALIZE THE LESION, ANGIOGRAPHY IS DONE AND SEEING TORTUOUS ARTERY WILL SIGNIFY THE PRESENCE OF LESION. TECHNETIUM-99MM LABELLED RED BLOOD CELL SCANS IS ALSO AN OPTION IN CASES WHERE ENDOSCOPY IS UNABLE TO FIND THE LESION.

THERAPEUTIC INTERVENTION:

ACCORDING TO GUIDELINES, THE THERAPEUTIC OPTIONS FOR DIEULAFOY'S LESION ARE:

- 1- ENDOSCOPIC ELECTROCOAGULATION, SCLEROTHERAPY OR BAND LIGATION
- 2- ARTERIAL EMBOLIZATION VIA ANGIOGRAPHY
- 3- SURGICAL INTERVENTION
- 4- LAPAROSCOPIC INTERVENTION.

THERE IS NO CLEAR OPTION FOR TREATING THE DIEULAFOY'S LESION [13] THE OPTION FOR TREATMENT IS CHOSEN ON THE BASIS OF PRESENTATION, LOCATION OF THE LESION AND THE ACCESS TO EXPERTISE.

ENDOSCOPIC PROCEDURE IS PREFERRED IN EASY ACCESS TO LESIONS. THE SUCCESS RATE IS NEAR TO 90%. ENDOSCOPIC METHODS ARE FURTHER CATEGORIZED AS (I) THERMAL – ELECTROCOAGULATION, HEAT PROBE COAGULATION AND ARGON PLASMA COAGULATION; (II) REGIONAL INJECTION – LOCAL EPINEPHRINE INJECTION AND SCLEROTHERAPY; AND (III) MECHANICAL – BANDING AND HAEMOCLIP.

ANGIOGRAPHY IS ANOTHER MODALITY OF TREATMENT, USED TO EMBOLIZE ACTIVELY BLEEDING LESIONS. IT IS HELPFUL WHEN THE ENDOSCOPIC METHODS FAIL TO ACHIEVE HEMOSTASIS. THE LIMITATION OF THIS MODALITY IS ISCHEMIA TO THE SITES SUPPLIED BY THE SAME ARTERY

SURGICAL INTERVENTION IS A CLASSICAL TREATMENT OPTION, MOSTLY DONE BY UNDER-RUNNING OF THE LESION OR A WEDGE RESECTION OF THE AFFECTED SECTION OF GUT [14] SURGICAL RESECTION DEPENDS UPON THE LOCATION OF LESION AND CLINICAL ASPECTS PATIENTS.

LAPAROSCOPIC INTERVENTION IS ALSO RECOMMENDED AS IT IS LEAST INVASIVE OPTION. BUT IT DEPENDS ON THE WELL LOCALIZATION OF BLEEDING VESSEL.

THE ADVANCEMENT IN THE MODALITY OF DIAGNOSIS HAS BETTER DECREASED THE MORTALITY FROM 80%. TO 8.6%. ACCORDING TO LUIS *ET AL* [12] THE RESULTS OF DIEULAFOY'S LESION PRESENT IN GI ARE BETTER AS COMPARED TO BLEEDING FROM GASTRIC OR DUODENAL ULCERS.

SURGICAL INTERVENTION REMAINS A BETTER CHOICE IN CASE OF MASSIVE HEMATEMESIS IN SETTINGS WHERE THERE IS LESS AVAILABILITY TO EXPERTISE. SURGICAL INTERVENTION IS SELECTED ON BASIS OF LESION'S PRESENTATION AND PHYSICAL CONDITION OF PATIENT.

Patient information:

A 40-year-old married woman was presented to gastro clinic with history of Hematemesis for 17 days. Hematemesis was episodic, with frequency of 2-3 times/ day, aggravated with meal intake approximately 2 cups of fresh and clotted blood per episode. Patient also had melena for the same duration. Patient had no history of such events in past but her medication history included the use of NSAIDS (Non-Steroidal anti-inflammatory Drugs) ,which she was self-medicating Diclofenac sodium (50mg) as needed for knee pain.

On general physical examination, patient was pale and anemic. Pulse= 110/ minute, Blood Pressure= 100/60mmHg, Temperature= 98 F, Respiratory Rate = 16/ minute. Patient had mild splenomegaly. The systemic examination was unremarkable.

Patient was admitted in gastric ward and diagnostic procedures were done. The esophageal, gastric and duodenal pathologies were among the differentials for this patient to focus. To have better idea of the cause and source of bleeding the following diagnostic assessments were done:

CBC (complete blood picture) : showed hemoglobin of 6.5 g/dl on admission.

Serology: Hepatitis B, Hepatitis C, Dengue, Malaria screening and Liver profile (**Table 1**) turned out to be normal.

Ultrasound: showed Splenomegaly.

EGD was planned to have a better look into esophagus, stomach and duodenum to know the cause of Bleeding. The first EGD reported the finding was “Pan Gastritis”. Patient was kept NPO (nil per os), IV (Intravenous) fluid and IV PPI (proton pump inhibitors) was started. After 24 hours when oral diet was tried, she developed Hematemesis again. Since the first EGD remained inconclusive, patient went through another Endoscopy, but no significant findings were found other than clotted Blood. She was kept NPO for 3 days but conservative management failed again. Whenever she was allowed to eat, the blood-stained vomiting occurred. 3rd time upper GI endoscopy revealed a visible vessel at the margin of body and fundus near lesser curvature of stomach and **Dieulafoy’s lesion** was suspected. (**Figure 1**)

When EGD revealed the Dieulafoy’s lesion, 2 attempts of regional injection – local epinephrine injection were done but failed to stop hemetamesis. Due to non-availability of further management options and expertise, massive hematemesis and location of lesion, further management was halted and surgery department was consulted for further plans. Exploratory Laparotomy was done. Per operative findings, “A lesion was found near greater curvature about 8- 10 cm from gastro esophageal junction with a pulsatile manner. It was

sutured with 2/0 Vicryl by under run suturing. Two small non-bleeding ulcers were also found near pyloric region that were secured as well. Stomach was closed in two layers. N/G (Nasogastric) tube was placed. Patient was kept NPO for 5 days.

Post operatively, patient had a smooth recovery without any episode of bleeding in N/G (Nasogastric) tube. She developed only surgical site infection (SSI)

Patient was discharged from hospital on her 7th day of surgery. Patient was scheduled for follow up after 2 weeks. On the day of follow up, Patient was assessed about symptoms and brief examination was also done. Patient had no symptoms and no further episode of Hematemesis. The surgical wound infection was resolved.

Discussion:

Dieulafoy's Lesion is acute GI (gastrointestinal) bleeding disorder, a rare but life threatening due to recurrent bleeding. The affected artery can protrude through a small defect in mucosa, become susceptible to even mechanical trauma of a minor level and gradually erodes into the lumen to cause severe acute GI bleeding.

40 years old married woman who was admitted with a history of massive Hematemesis for 17 days. Series of 3 upper GI endoscopies were done and patient was diagnosed with Dieulafoy's lesion. Due to failure of 2 attempts of local epinephrine injection via EGD and lack of further expertise, surgical department was consulted. Exploratory Laparotomy was done. Per operatively "A lesion was found near greater curvature about 8- 10 cm from gastro esophageal junction that showed a pulsatile manner. It was sutured with 2/0 Vicryl, by under run suturing. Patient had a smooth recovery without any episode of bleeding. The only post-operative complication was surgical site infection (SSI).

Surgical intervention is selected on basis of lesion's presentation and clinical aspects of patient. Surgical intervention remains a better choice in case of massive hematemesis in setting where there is less access to availability of expertise.

Conclusion :

Dieulafoy's lesion is a rare cause of Upper GI bleeding. In this case patient presented with massive hematemesis. series of 3 EGD were done to diagnose the disease. 2 attempts of local epinephrine injection via EGD failed to stop hemetamesis. Due to lack of access to expertise , massive hemetamesis and site of lesion, surgical intervention was done . Surgical outcomes were satisfactory.

DECLARATIONS

Ethics approval and consent to participate

Written consent was obtained from the patients for publication of study.

Availability of data and material

All data and material are shared in Table (Lab Values) and Figures

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Table: 1 Labs Value**CBC :**

Date	Hb g/dl	Platelets $\times 10^9/l$	RBC $\times 10^{12}/l$	WBC $\times 10^9/l$
When admitted in Gastro ward (before surgical ward admission)				
1 st day	6.5	107	3.34	4.7
7 th day	10.6	128	3.53	4.5
13 th day	9.0	211	2.80	9.0
17 th day	7.3	180	2.26	4.2
When admitted in surgical ward				
Day of admission	8.2	169	2.62	7.09
2 weeks after surgery	12.0	220	4.42	7.90

ELECTROLYTES :

On day 17th, When Patient was referred to Surgical department, following work up was done before :

Sodium	133.4 mmol/L	Urea	55 mg/dl
Potassium	3.35 mmol/L	Creatinine	0.7 mg/dl
Chloride	99.8 mmol/L	Prothrombin Time	15 sec
Alk Phosphatase	187 u/L	APTT	34 sec
ALT	24 u/L	INR	1.28
Gamma-GT	14u/L	Bilirubin Total	0.7 mg/dl
HbsAg	Negative	Anti HIV	Negative
HCV antibodies	Negative	Anti H.Pylori	Negative
Dengue Antibodies IgM ICT	Negative	Dengue antibodies IgG ICT	Negative



Figure 1 showing a visible vessel at the margin of body and fundus near lesser curvature of stomach