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Massive Gastric Polyposis: A Rare Cause of Upper GI Bleed

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Abstract: ***This is a case report of a patient who presented multiple times to a district general hospital with symptoms caused by gastric polyposis. Eventually, the patient underwent a total gastrectomy due to persistent symptoms and the high number of polyps in her stomach. We investigate the possible causes of the patient's polyposis and review the current guidelines on management and surveillance.***

Keywords: gastric polyposis, total gastrectomy, patient case report, management guidelines, surveillance of polyps

1. Background

"Gastric polyps" are defined as luminal lesions projecting above the plane of the mucosal surface. It is a term that is used in patients where multiple gastric polyps are found. The term "diffuse polyposis" is used when all or a large proportion of the gastric mucosa is covered with gastric polyps. [1]

In the United States, gastric polyps are found in approximately 6% of oesophago - gastro - duodenoscopies (OGDs). In areas where *Helicobacter pylori* infections are prevalent, the most common polyps found are gastric hyperplastic polyps (GHPs). In areas where proton pump inhibitor (PPI) use is prevalent fundic gland polyps are more common secondary to the trophic effect on parietal cells. [2] Other rarer types of gastric polyps are gastric adenomas, neuroendocrine tumours, and inflammatory fibroid polyps.

Inherited gastrointestinal (GI) polyposis should be considered in patients with diffuse polyposis. The diagnosis requires correlation with clinical, endoscopic, and molecular findings. The histopathologic findings are useful in guiding or supporting diagnostic workups. If symptoms of this disease are observed, performing appropriate clinical surveillance and management is significantly important.

We report a case of a patient who was admitted numerous times with symptoms of upper GI bleeding and anaemia, requiring multiple blood transfusions and OGDs. Since multiple polyps were observed in her stomach, she eventually required a total gastrectomy.

2. Case Presentation

A female patient, aged 48, was admitted to the emergency department due to experiencing mild epigastric pain and coffee - ground vomiting. Her medical background included a diagnosis of asthma, chronic anemia, diverticular disease, irritable bowel syndrome, and a previous right - sided salpingo - oophorectomy for cysts. The patient had not previously sought consultation with gastroenterology or undergone endoscopy.

3. Investigations

During the initial evaluation, the patient exhibited a systolic blood pressure of 107 mmHg and a pulse rate of 98 bpm. The Glasgow - Blatchford bleeding score yielded a value of 7, with specific measurements including a haemoglobin level of 94 g/l, a mean corpuscular volume of 102 µm3, and a urea level of 4.8A 48 - year - old woman presented to the emergency department with mild epigastric pain and coffee - ground vomiting. She had a medical history of asthma, chronic anaemia, diverticular disease, irritable bowel syndrome, and right - sided salpingo - oophorectomy for cysts. She had no previous correspondence with gastroenterology or endoscopies. She had a family history of colon cancer. She did not undergo any surveillance endoscopy screening.

Investigations

Table 1 - Key observations and bloods on presentation
Haemoglobin - 94 g/l
MCV - 102 µm³102 µm^{3}102 µm^{3}102 µm^3
Systolic Blood Pressure - 107mmg
Pulse Rate - 98
Urea - 4.8mmol/L
Glasgow Blatchford Score - 7

On initial assessment, the patient's systolic blood pressure and pulse rate were 107 mmHg and 98 bpm, respectively. We obtained a Glasgow - Blatchford bleeding score of 7 (haemoglobin [Hb] 94 g/l; mean corpuscular volume, 102 µm^3; and urea, 4.8 mmol/L). Whilst performing an OGD on the day of admission, several gastric polyps and a small amount of fresh blood around the scattered polyp site were observed

(Figures 1 and 2)

4. Treatment

The patient received supportive intravenous fluid and a proton pump inhibitor (PPI). Additionally, two units of red blood cells (RBCs) were transfused the next day due to a decrease

[1] Powar D, Kanbarkar D. Diffuse gastric polyposis: a rare case report. *IOSR J Dent Med Sci* 2017;16:117–20.
[2] Miyamoto, S. *et al.* (2017) 'Gastric hyperplastic polyps associated with proton pump inhibitor use in a case without a history of *helicobacter pylori* infection', *Internal Medicine*, 56(14), pp. 1825–1829. doi:10.2169/internalmedicine.56.8040.

in hemoglobin level (76 g/l). Coffee - ground vomiting was observed. An esophagogastroduodenoscopy (OGD) was performed, which revealed the presence of multiple polyps. (image - 1)

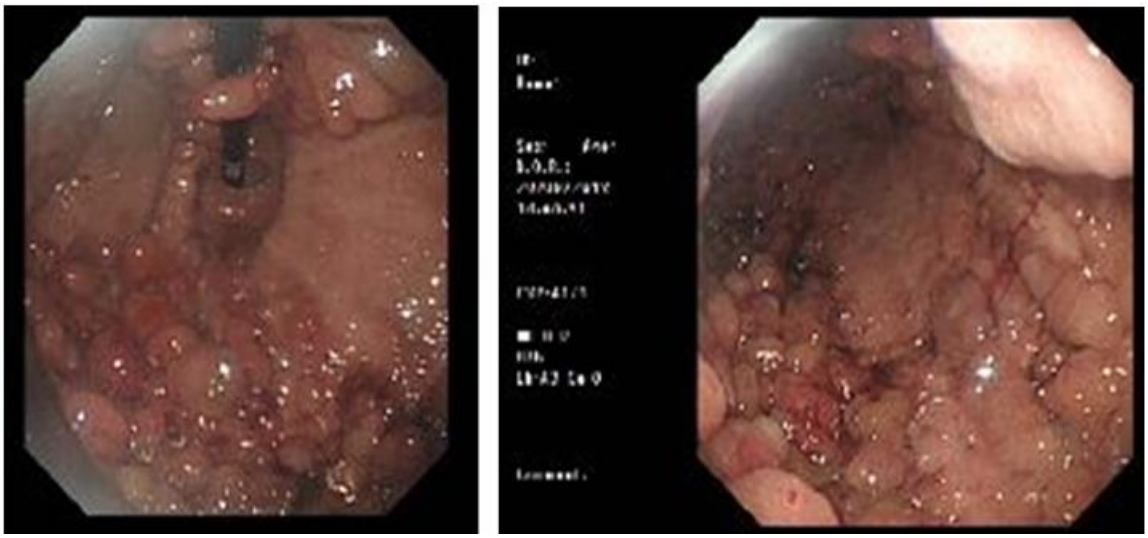


Image 1

Outcome and Follow - Up

The patient was readmitted to the emergency department four days after being discharged. She reported experiencing coffee - ground vomiting, epigastric pain, and overall malaise. Her hemoglobin (Hb) level remained stable at 92 g/l. Despite her systolic blood pressure measuring 104 mmHg, her pulse rate was 91 beats per minute (bpm).

The patient exhibited recurrent vomiting and intense abdominal pain, primarily localized to the left iliac fossa necessitating IV Morphine. The general surgical team was consulted and they recommended an abdominopelvic computed tomography (CT) scan. The CT scan (Image - 2) revealed indications of significant obstruction in the gastric outflow, with a distinct transition point observed.

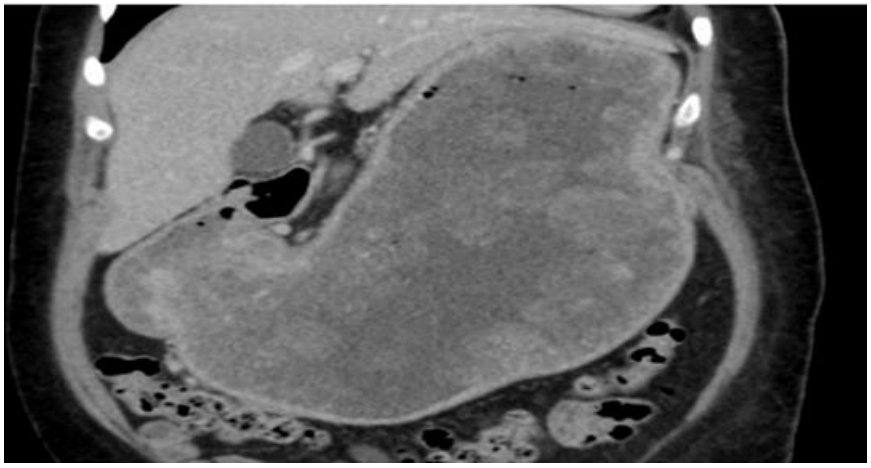


Image 2

Consequently, the patient was admitted on three separate occasions due to coffee - ground emesis and reduced hemoglobin (Hb) levels. A total of six upper gastrointestinal (GI) endoscopies were performed, along with a colonoscopy within a span of 2 months. Over the course of this period, more than 20 units of red blood cells (RBCs) were transfused.

Several endoscopic examinations were conducted, which confirmed the presence of polyps throughout the entire stomach. The polyps varied in size, with the largest measuring 25 mm and the smallest measuring 10 mm. Additionally, three benign sessile polyps were observed, with the largest measuring 35 mm. Notably, the larger polyps exhibited signs of active bleeding in the region extending from the fundus. The colonoscopy examination revealed a limited presence of diverticula, second - degree haemorrhoids, and skin tags. No discernible origin of bleeding or polyps could be identified.

The original plan entailed conducting outpatient polypectomies in multiple phases. However, due to the patient's unstable condition, persistent need for blood transfusions, and challenges associated with resecting numerous stomach polyps, the decision was made to transfer the patient to a tertiary center for a complete gastrectomy (refer to Image - 3).

The histological examination revealed the presence of numerous polyps, likely exceeding 100 in number, on the gastric mucosa. These polyps exhibited a diverse range of morphologies, including bulbous and pedunculated shapes

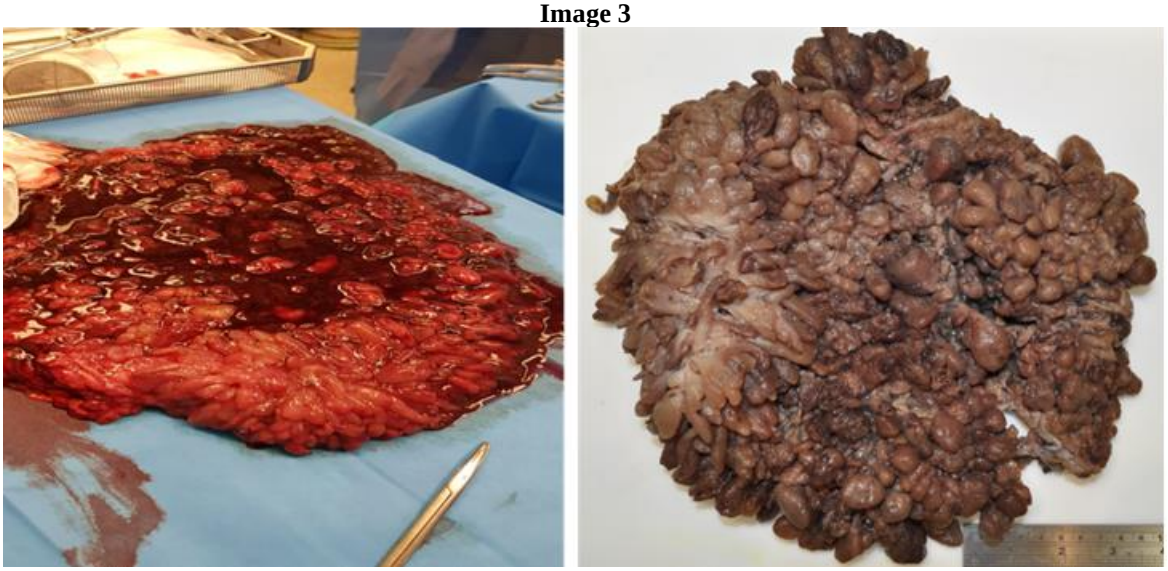
measuring up to 40 x 22 x 17 mm, as well as more slender and finger - like structures measuring up to 44 x 12 x 6 mm. A significant number of polyps displayed signs of erosion, ulceration, and congestion. The body and distal region of the stomach were particularly affected by a dense concentration of polyps. An ulcer was observed within the distal stomach, however, no prominently alarming characteristics were observed upon examining the sliced ulcerated area.

Under microscopic examination, the observed polyps exhibited a spherical shape and lacked villi - like structures, indicating a lack of specialized mucosa. The presence of pronounced stromal edema and sporadic strands of smooth muscle were also observed. Additionally, several glands displayed dilation. The presence of distinctly localized acute and chronic inflammation suggested the possibility of very focal intestinal metaplasia. The presence of distal gastric ulceration accompanied by a multitude of hyperplastic - type polyps, exceeding a count of 100, was observed to affect the entirety of the stomach.

The histopathology report presented several potential diagnoses, including gastric hyperplastic polyposis, juvenile polyposis syndrome (JPS), and Cowden syndrome (CS). Additionally, the possibility of massive gastric juvenile - type polyposis (MGJP) was considered due to the similarity of the patient's symptoms to those typically observed in this particular form of polyposis.

The patient underwent a consultation with a clinical geneticist in an outpatient setting, revealing the presence of two unidentified variants of SMAD4 and BMPR1A genes. Molecular testing did not definitively establish her diagnosis, however, the patient was recommended to undergo regular colonoscopy screenings to monitor her heightened susceptibility to developing malignancies.

Image 3



5. Discussion

This case is unusual as it highlights a polyposis syndrome that is exclusive to the upper GI tract and no evidence of lower GI polyps or extra - intestinal manifestations.

It is a possible that this patient may have Juvenile Polyposis Syndrome (JPS) with purely upper GI involvement as this patient was found to have two unclassified variants of SMAD4 and BMPR1A although we were unable to confirm her diagnosis at a molecular level. There is also a phenomenon called Massive Gastric Juvenile type polyposis (MGJP), a rare associated overlapping syndrome JPS. Massive gastric juvenile - type polyposis appears to occur later in life unlike JPS which is more common in the paediatric population. We believe that this is an important differential for clinicians to be aware of as it has implications in terms of management and surveillance.

The main difference between MGJP and JPS is the location and extent of polyps. In MGJP, the polyps are limited to the stomach. However in JPS they can occur at multiple locations in the GI tract

MGJP is often referred in the literature using various terms ranging from ‘massive gastric polyposis’, ‘juvenile polyposis of the stomach’, ‘gastric juvenile polyposis’, to ‘hyperplastic gastric polyposis’. This phenomenon is not entirely surprising as it is often difficult, or impossible, to confidently differentiate between MGJP and JPS based on their histology [2].

JPS is a hamartomatous polyposis syndrome that has an autosomal dominant mode of inheritance with variable penetrance. It is estimated to affect approximately 1 in 100, 000 individuals [3] with up to 20–50% of cases having a family history of JPS [4]. The diagnosis of JPS is established when any of the following clinical criteria are met: more than five juvenile polyps of the colon or rectum, juvenile polyps in other parts of the GI tract, any number of juvenile polyps, and a positive family history [5].

Histologically, the polyps in JPS are harmatomas comprising normal epithelium with a dense stroma, inflammatory infiltrate, and smooth surface with dilated, mucus - filled cystic glands in the lamina propria [6].

Mutations in SMAD4 and BMPR1A genes involved in the transforming growth factor β pathway are associated with JPS

development. It is estimated that there is a 20% probability of mutation in each of these genes in patients with this syndrome [7]. In a study conducted by Sayed et al. [8], who investigated 54 familial and sporadic cases of JPS, it was found that 41% of patients were positive for either SMAD4 or BMPR1A mutations. In this study, SMAD4 - positive groups had a higher age of diagnosis, frequency of positive family history of GI cancer, frequency of GI polyps, and frequency of upper GI polyps compared to mutation - negative patients. Based on Friedl et al. 's study [9] highlighting 29 JPS patients, massive gastric polyposis was a feature only observed in SMAD4 - positive patients, and not in BMPR1A - positive and mutation - negative patients. It has been documented that SMAD4 +ve patients with JPS are more likely to have upper GI involvement.

Massive gastric juvenile type polyposis is a rare entity often associated with Juvenile polyposis syndrome. SMAD4 germline mutation (but not other JPS - associated mutations) increase the risk of gastric polyps and gastric carcinoma in syndromic patients; massive gastric juvenile - type polyposis may indicate the presence of such mutations.

Patients with MGJP typically present with complaints of iron deficiency anaemia, upper GI bleeding, abdominal pain, haematemsis, vomiting, early satiety, weight loss, malaena, hypoproteinaemia and hypergastrinemia.

Reference 22 Patient Paper

Table 2: Genetic mutations in other similar conditions [10]

Familial adenomatous polyposis and associated polyps	Adenomatous polyposis coli (APC) gene located on chromosome 5q21 - q22
MutYH - associated polyps (MAP)	MutYH Gene - Y179C and G396D (most common gene mutations found in caucasion population)
Peutz - Jeghjers syndrome and Peutz - Jeghers polyps	80% in STK11/LKB1 gene, 25% sporadic
Juvenile Polyposis syndrome and Juvenile Polyps	15% SMAD4, 25% in BMPR1A
PTEN syndromes and associated hamartomatous polyps	Tumour suppressor gene phosphate and tensin homologoue (PTEN)

Management of MGJP – Gastrectomy

A 2017 clinicopathological analysis of 22 patients presenting with MGJP suggested that a large proportion of these patients required major surgival intervention as part of their management. For example, 16 of the patients in this particular analysis underwent total gastrectomy, whereas others underwent partial gastrectomy and serial polypectomies [11].

Gastric cancer risk is also increased in patient with SMAD4 pathogenic variants. The lack of significant pathology being found in childhood, it is recommended that those with SMAD4 mutation undergo upper GI tract surveillance 1 - 3 yearly from the age of 18 years, and those with BMPR1A mutation from 25 years of age (12)

6. Conclusion

Gastric polyps are often found in routine endoscopies. Although patients are often asymptomatic and polyps are incidentally found, occasionally, they can cause symptoms such as anaemia, melaena, and gastric outlet obstruction. One should consider a polyposis syndrome in patient presenting with high burden of GI polyps as this can have direct implications on management and surveillance.

Learning Points/Take Home Messages

- To consider polyposis syndrome in patients presenting with high burden of GI polyps
- Early consideration of surgical intervention
- Importance of liaising with histopathologists to support diagnosis

References

[1] Powar D, Kanbarkar D. Diffuse gastric polyposis: a rare case report. *IOSR J Dent Med Sci* 2017; 16: 117–20.
[2] Gonzalez R, Adsay V, Graham R, et al. Massive gastric juvenile - type polyposis: a clinicopathological analysis of 22 cases. *Histopathology*, 2017; 70: 918–28.
[3] Burt RW, Bishop DT, Lynch HT, et al. Risk and surveillance of individuals with heritable factors for colorectal cancer. *Bull World Health Organ* 1993; 68: 655–64.
[4] Howe JR, Ringold JC, Hughes JH, et al. Direct genetic testing for Smad4 mutations in patients at risk for juvenile polyposis. *Surgery* 1999; 126: 162–70.
[5] Jass JR, Williams CB, Bussey HJ, et al. Juvenile polyposis—a precancerous condition. *Histopathology* 1988; 13: 619–30.
[6] Kelly S, Dwerryhouse S, Safranek P, et al. Juvenile polyposis syndrome affecting the stomach: a case report. *J Med Case Rep* 2008; 2: 314.
[7] Howe JR, Sayed MG, Ahmed AF, et al. The prevalence of MADH4 and BMPR1A mutations in juvenile polyposis and absence of BMPR2, BMPR1B, and ACVR1 mutations. *J Med Genet* 2004; 41: 484–91.
[8] Sayed MG, Ahmed AF, Ringold JR, et al. Germline SMAD4 or BMPR1A mutations and phenotype of juvenile polyposis. *Ann Surg Oncol* 2002; 9: 901–6.
[9] Friedl W, Uhlhaas S, Schulmann K, et al. Juvenile polyposis: massive gastric polyposis is more common in MADH4 mutation carriers than in BMPR1A mutation carriers. *Hum Genet* 2002; 111: 108–11.
[10] Lam - Himlin et al. Gastric polyps and polyposis syndromes, Diagnostic Histopathology 2014; 1: 1 - 11.
[11] Ngeow J, Eng C. PTEN hamartoma tumor syndrome: clinical risk assessment and management protocol. *Methods* 2015; 77: 11–9.
[12] Monahan KJ, et al. Guidelines for the management of hereditary colorectal cancer from the British Society of Gastroenterology (BSG) /Association of Coloproctology of Great Britain and Ireland (ACPGBI) / United Kingdom Cancer Genetics Group (UKCGG) Gut 2019; 0: 1–34. .

Patient's Perspective

I was shocked when I was diagnosed with polyps in my stomach. I had been prepared for the possible presence of polyps in my bowel as my father had them and eventually died

of bowel cancer at the age of 29 years. There is no other history of stomach polyps or stomach cancer in my family.

Initially, I thought that the doctors would be able to resect the polyps, and I would still have my stomach. I was extremely weak due to blood loss as a result of polyp bleeding, tired from 7 weeks of staying in the hospital, and was immobile (I usually lead a busy life). Upon hearing the news that my stomach would have to be completely removed, I was shocked since I had never heard of people having their stomach removed. Moreover, I was afraid in case I had cancer or was going to die. I did not want to leave my husband to raise our children, one of whom is autistic.

I did not think that I had the strength to recover from the operation and to go on to enjoy a good quality of life. It has taken me 2 years to reach the stage where I can now eat three meals a day, although it has been a long and tough journey. I experience some discomfort, and bile buildup suggests that I am sick every once in a while. However, my quality of life has improved. I know that I am extremely lucky to have received the medical care and attention that I have and that the outcome could have been very different. I hope that my experience can help in the treatment process of other patients who find themselves in the same situation. I wish my stomach had been removed a little earlier, when it became clear that there was no other solution.