Brunei International Medical Journal

OFFICIAL PUBLICATION OF
THE MINISTRY OF HEALTH
AND
UNIVERSITI BRUNEI DARUSSALAM

Volume 17 27 November 2021 (22 Rabiulakhir 1443H)

RARE ENTITY OF TWO-POINT INTUSSUSCEPTIONS IN AN ADOLESCENT WITH PEUTZ-JEGHERS SYNDROME.

Jien Yen SOH, Iqtidaar OARIS , Siti Rahmah HASHIM ISA MERICAN, Zaidi ZAKARIA.

Department of Surgery, Hospital Universiti Sains Malaysia and School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.

ABSTRACT

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder characterized by hamartomatous polyposis and mucocutaneous pigmentation. We report a case of a 16-year-old adolescent with chronic abdominal pain and anemia who subsequently developed sudden severe abdominal pain. Clinical examination showed intestinal obstruction, and ultrasound suggested intussusception. He underwent immediate exploratory laparotomy. Intraoperatively, there were two points of jejunal intussusception with intraluminal polyps. Segmental bowel resection was performed with primary anastomosis. The histological examination came back as Peutz-Jegher polyps. We discuss difficulties in diagnosing PJS during the initial presentation, and the diagnosis was only made after the complication occurred. Clinical suspicion with an earlier endoscopic investigation can provide preventive intervention to avoid complication. In the case of intussusception with PJS, a complete assessment of the entire length of the bowel is necessary intraoperatively as there might be additional pathology or additional points of intussusception.

Keywords: Adolescent, Anemia, Intussusception, Peutz-Jegher Syndrome, Polyp, Surgery.

Brunei Int Med J. *2021;17:149-152*

ISSN 1560 5876 Print
ISSN 2079 3146 Online

Online version of the journal is available at www.bimjonline.com

Brunei International Medical Journal (BIMJ)
Official Publication of
The Ministry of Health
and
Universiti Brunei Darussalam

ISSN 1560-5876 Print
ISSN 2079-3146 Online

Aim and Scope of Brunei International Medical Journal

The Brunei International Medical Journal (BIMJ) is a six monthly peer reviewed official publication of the Ministry of Health under the auspices of the Clinical Research Unit, Ministry of Health, Brunei Darussalam.

The BIMJ publishes articles ranging from original research papers, review articles, medical practice papers, special reports, audits, case reports, images of interest, education and technical/innovation papers, editorials, commentaries and letters to the Editor. Topics of interest include all subjects that relate to clinical practice and research in all branches of medicine, basic and clinical including topics related to allied health care fields. The BIMJ welcomes manuscripts from contributors, but usually solicits reviews articles and special reports. Proposals for review papers can be sent to the Managing Editor directly. Please refer to the contact information of the Editorial Office.

Instruction to authors

Manuscript submissions

All manuscripts should be sent to the Managing Editor, BIMJ, Ministry of Health, Brunei Darussalam; e-mail: editor-in-chief@bimjonline.com. Subsequent correspondence between the BIMJ and authors will, as far as possible via should be conducted via email quoting the reference number.

Conditions

Submission of an article for consideration for publication implies the transfer of the copyright from the authors to the BIMJ upon acceptance. The final decision of acceptance rests with the Editor-in-Chief. All accepted papers become the permanent property of the BIMJ and may not be published elsewhere without written permission from the BIMJ.

Ethics

Ethical considerations will be taken into account in the assessment of papers that have experimental investigations of human or animal subjects. Authors should state clearly in the Materials and Methods section of the manuscript that institutional review board has approved the project. Those investigators without such review boards should ensure that the principles outlined in the Declaration of Helsinki have been followed.

Manuscript categories

Original articles

These include controlled trials, interventional studies, studies of screening and diagnostic tests, outcome studies, cost-effectiveness analyses, and large-scale epidemiological studies. Manuscript should include the following; introduction, materials and methods, results and conclusion. The objective should be stated clearly in the introduction. The text should not exceed 2500 words and references not more than 30.

Review articles

These are, in general, invited papers, but unsolicited reviews, if of good quality, may be considered. Reviews are systematic critical assessments of literature and data sources pertaining to clinical topics, emphasising factors such as cause, diagnosis, prognosis, therapy, or prevention. Reviews should be made relevant to our local setting and preferably supported by local data. The text should not exceed 3000 words and references not more than 40.

Special Reports

This section usually consist of invited reports that have significant impact on healthcare practice and usually cover disease outbreaks, management guidelines or policy statement paper.

Audits

Audits of relevant topics generally follow the same format as original article and the text should not exceed 1,500 words and references not more than 20.

Case reports

Case reports should highlight interesting rare cases or provide good learning points. The text should not exceed 1000 words; the number of tables, figures, or both should not be more than two, and references should not be more than 15.

Education section

This section includes papers (i.e. how to interpret ECG or chest radiography) with particular aim of broadening knowledge or serve as revision materials. Papers will usually be invited but well written paper on relevant topics may be accepted. The text should not exceed 1500 words and should include not more than 15 figures illustration and references should not be more than 15.

Images of interest

These are papers presenting unique clinical encounters that are illustrated by photographs, radiographs, or other figures. Image of interest should include a brief description of the case and discussion with educational aspects. Alternatively, a mini quiz can be presented and answers will be posted in a different section of the publication. A maximum of

three relevant references should be included. Only images of high quality (at least 300dpi) will be acceptable.

Technical innovations
This section include papers looking at novel or new techniques that have been developed or introduced to the local setting. The text should not exceed 1000 words and should include not more than 10 figures illustration and references should not be more than 10.

Letters to the Editor
Letters discussing a recent article published in the BIMJ are welcome and should be sent to the Editorial Office by e-mail. The text should not exceed 250 words; have no more than one figure or table, and five references.

Criteria for manuscripts
Manuscripts submitted to the BIMJ should meet the following criteria: the content is original; the writing is clear; the study methods are appropriate; the data are valid; the conclusions are reasonable and supported by the data; the information is important; and the topic has general medical interest. Manuscripts will be accepted only if both their contents and style meet the standards required by the BIMJ.

Authorship information
Designate one corresponding author and provide a complete address, telephone and fax numbers, and e-mail address. The number of authors of each paper should not be more than twelve; a greater number requires justification. Authors may add a publishable footnote explaining order of authorship.

Group authorship
If authorship is attributed to a group (either solely or in addition to one or more individual authors), all members of the group must meet the full criteria and requirements for authorship described in the following paragraphs. One or more authors may take responsibility 'for' a group, in which case the other group members are not authors, but may be listed in an acknowledgement.

Authorship requirement
When the BIMJ accepts a paper for publication, authors will be asked to sign statements on (1) financial disclosure, (2) conflict of interest and (3) copyright transfer. The correspondence author may sign on behalf of co-authors.

Authorship criteria and responsibility
All authors must meet the following criteria: to have participated sufficiently in the work to take public responsibility for the content; to have made substantial contributions to the conception and design, and the analysis and interpretation of the data (where applicable); to have made substantial contributions to the writing or revision of the manuscript; and to have reviewed the final version of the submitted manuscript and approved it for publication. Authors will be asked to certify that their contribution represents valid work and that neither the manuscript nor one with substantially similar content under their authorship has been published or is being considered for publication elsewhere, except as described in an attachment. If requested, authors shall provide the data on which the manuscript is based for examination by the editors or their assignees.

Financial disclosure or conflict of interest
Any affiliation with or involvement in any organisation or entity with a direct financial interest in the subject matter or materials discussed in the manuscript should be disclosed in an attachment. Any financial or material support should be identified in the manuscript.

Copyright transfer
In consideration of the action of the BIMJ in reviewing and editing a submission, the author/s will transfer, assign, or otherwise convey all copyright ownership to the Clinical Research Unit, RIPAS Hospital, Ministry of Health in the event that such work is published by the BIMJ.

Acknowledgements
Only persons who have made substantial contributions but who do not fulfill the authorship criteria should be acknowledged.

Accepted manuscripts
Authors will be informed of acceptances and accepted manuscripts will be sent for copyediting. During copyediting, there may be some changes made to accommodate the style of journal format. Attempts will be made to ensure that the overall meaning of the texts are not altered. Authors will be informed by email of the estimated time of publication. Authors may be requested to provide raw data, especially those presented in graph such as bar charts or figures so that presentations can be constructed following the format and style of the journal. Proofs will be sent to authors to check for any mistakes made during copyediting. Authors are usually given 72 hours to return the proof. No response will be taken as no further corrections required. Corrections should be kept to a minimum. Otherwise, it may cause delay in publication.

Offprint
Contributors will not be given any offprint of their published articles. Contributors can obtain an electronic reprint from the journal website.

RARE ENTITY OF TWO-POINT INTUSSUSCEPTIONS IN AN ADOLESCENT WITH PEUTZ-JEGHERS SYNDROME.

Jien Yen SOH, Iqtidaar OARIS , Siti Rahmah HASHIM ISA MERICAN, Zaidi ZAKARIA.

Department of Surgery, Hospital Universiti Sains Malaysia and School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.

ABSTRACT

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder characterized by hamartomatous polyposis and mucocutaneous pigmentation. We report a case of a 16-year-old adolescent with chronic abdominal pain and anemia who subsequently developed sudden severe abdominal pain. Clinical examination showed intestinal obstruction, and ultrasound suggested intussusception. He underwent immediate exploratory laparotomy. Intraoperatively, there were two points of jejunal intussusception with intraluminal polyps. Segmental bowel resection was performed with primary anastomosis. The histological examination came back as Peutz-Jegher polyps. We discuss difficulties in diagnosing PJS during the initial presentation, and the diagnosis was only made after the complication occurred. Clinical suspicion with an earlier endoscopic investigation can provide preventive intervention to avoid complication. In the case of intussusception with PJS, a complete assessment of the entire length of the bowel is necessary intraoperatively as there might be additional pathology or additional points of intussusception.

Keywords: Adolescent, Anemia, Intussusception, Peutz-Jegher Syndrome, Polyp, Surgery.

INTRODUCTION

Intussusception is more common in the pediatric age group and rare in adults. It occurs when a loop of the bowel, the intussusceptum, telescopes into an adjacent segment, the intussuscipiens.[1] In the adult, the process occurs because of a pathological lead point lesion due to an infection, tumour, Meckel's diverticulum, and intestinal duplication, causing mechanical disruption of motility.[2] Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder characterized by hamartomatous polyposis and mucocutaneous pigmentation.[3] This syndrome is usually encountered in infancy and late adolescence, and the polyps mainly occur in the jejunum.[4] Due to the rarity of the disease, PJS is not usually diagnosed until complications arise from the disease. We report a case of a young man with two points of small bowel intussusception due to the hamartomatous polyps in PJS.

Correspondence: Siti Rahmah Hashim Isa Merican, Master of Medicine (MMED) Surgery USM, Hospital Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.
E-mail: sitimerican@gmail.com
Contact: +609-767 6774; Fax number: +609-764 5731

CASE REPORT

A 16-year-old teenager presented to Accident and Emergency Department, Hospital Universiti Sains Malaysia, with an acute episode of left-sided abdominal pain associated with recurrent vomiting for the past two days. However, he was still able to pass motion normally. He was previously investigated at the surgical outpatient clinic for chronic intermittent non-specific abdominal pain for more than a years which was associated with anemia. On examination, his vital signs were stable, his abdomen was soft and not distended with tenderness at the left lumbar and iliac fossa. There was a palpable mass over the right paraumbilical region measuring 10x7cm in size. Bowel sound on auscultation was present and not tinkly. Upon admission to the ward, an urgent abdominal ultrasound was performed which showed presence of a target sign located at the same right paraumbilical region, confirming the diagnosis of small bowel intussusception (Figure 1).

His parents were informed of the diagnosis, and a consent for emergency surgery was obtained. He underwent emergency laparotomy on the same day of admission. Intraoperative findings confirmed two points of jejunal intussusception, 20cm and 40cm from the duodeno-jejunal junction (Figure 2). Upon reducing the intussuscepted bowel, an intraluminal palpable mass at the affected site was noted. The affected bowel was resected, and primary end to end anastomosis

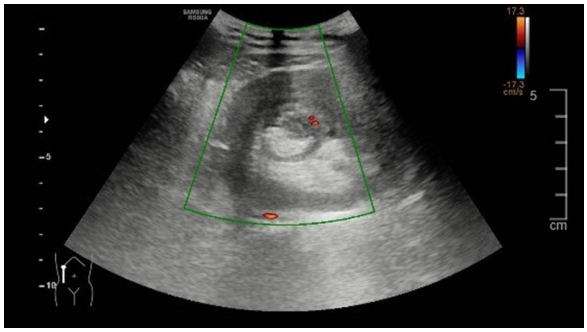

Figure 1: Ultrasound showing the appearance of a target sign. No colour Doppler flow. (Click on image to enlarge)

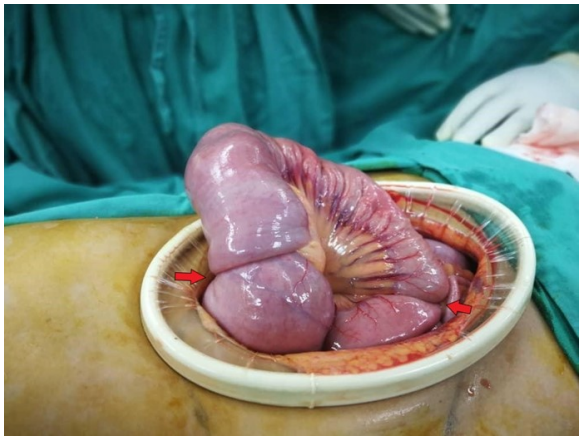

Figure 2:Arrows are showing the two points intussusceptions of the jejunum. (Click on image to enlarge)

performed. The resected bowel was incised and opened to revealed multiple intraluminal pedunculated polyps (Figure 3). The remaining small bowel and large bowel were inspected and found to be normal with no other pathology. The histological report of the resected bowels came back as hamartomatous polyps, which were composed of the central core of smooth muscle fibers, with tree-like branching, covered by intestinal-type epithelium forming villous configurations. There were no dysplasia, intramucosal carcinoma, or features of malignancy were seen. Based on the histological findings, a diagnosis of Peutz-Jeghers syndrome was made. Patient's post-operative recovery was uneventful, and he was discharged home well after a few days.

He was followed up regularly in surgical outpatient clinic and at his six months follow-up, he was noted to be well with no recurrence of abdominal symptoms. He was informed that he will require regular upper endoscopy and video capsule endoscopy at 2-3 years interval.

DISCUSSION

PJS is a rare autosomal dominant disorder, with the presence of familial gastrointestinal hamartomatous polyps and mucocutaneous pigmentation due to a mutant gene localized at 19p34-p36 named STK1, which is a serine/

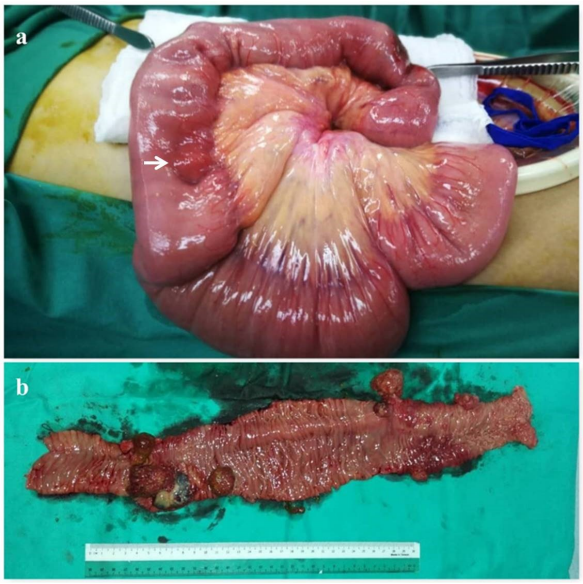

Figure 3: a)The jejunum post-reduction revealed a palpable mass intraluminal at the intussusception point. b) Multiple pedunculated polyps intraluminal in the resected jejunum. (Click on image to enlarge)

threo-nine kinase that regulates growth.[4]

This young man has been complaining of abdominal pain for a year and was investigated for anemia at the outpatient clinic. However, PJS was not suspected as he had no family history nor mucocutaneous pigmentation. The earliest symptom of PJS is mucocutaneous pigmentations which are melanotic pigmented macules, described as dark brown or bluish brown in colour and 1-5 mm in size, commonly at the lip in adult.[4] Without family history, PJS is hard to diagnose until the presentation of severe GI complications.[5]

The hamartomatous polyps in the gastro-intestinal system can cause multiple complications such as bleeding, obstruction, intussusception and an increase risk of malignancy.[1,4] In our case, the patient had anaemia, most likely due to occult bleeding from the polyps, which was undiagnosed during the outpatient follow-up. Hamartomatous polyps have a highly vascular stroma with a tendency to bleed.[6]

Intussusception more commonly occurs in the paediatric age group and rarely in adults. In paediatric intussusception, non-operative image-guided enema reduction is the first-line treatment of choice.[7] However, this does not apply to adult case as intussusception in the adult is due to a pathological lead point lesion which can be infection, tumour, Meckel's diverticulum, and intestinal duplication that causing mechanical disruption of motility.[2]

In our patient, with confirmatory evidence of intussusception from the imaging findings, we immediately informed the patient's parents and obtained consent for emergency laparotomy. Intussusception may be the initial presentation for adult individuals with PJS, and it occurs mostly due to the polyps in the small intestine.[1] Although intussusception can be easily recognized via clinical examination and ultrasound, we would like to highlight the rare occurrence of more than one point of intussusception along the small intestine. This is because the leading points due to hamartomatous polyps can occur at more than one site in PJS. Thus, in adults with intussusception, the whole length of the intestine should be thoroughly inspected and examined during laparotomy for additional pathologies, such as in our case, a second intussusception point.

There is no definitive guideline in the management of Peutz-Jeghers polyps, however resection is generally advised in the cases of bleeding or intussusception.[1] The polyps are most commonly at the jejunum, but they can occur throughout the gastrointestinal tract.[4] The other common location of these polyps is in the colon and stomach.[8] Thus, upper endoscopy, video capsule endoscopy and colonoscopy are recommended in such cases postoperatively every 2-3 years.

CONCLUSION

It is rare to encounter a case of intussuscep-

tion in a young man with undiagnosed PJS. Due to the rarity of the disease, PJS is not usually diagnosed till complications arise. Clinical suspicion with an earlier endoscopic investigation can provide preventive intervention to avoid complication. In the case of intussusception with PJS, a complete assessment of the entire length of the bowel is necessary intraoperatively as there might be an additional pathology or additional point of intussusception.

CONFLICT OF INTEREST

The authors reported no conflict of interest or financial liability.

INFORMED CONSENT

Consent has been obtained from the patient with regards to the use of the pictures for publication.

REFERENCES

1: Shah J, Sunkara T, Xiao P, Gaduputi V, Reddy M, Razia S. Peutz-Jeghers syndrome presenting as colonic intussusception: A rare entity. Gastroenterology Res. 2018;11:150-3. [Accessed on 5 June 2021].

2: Bolivar-Rodriguez MA, Fierro-Lopez R, Pamanes-Lozano A, Cazarez-Aguilar MA, Osuna-Wong BA, Ortiz-Bojorquez JC. Surgical outcome of jejunum-jejunum intussusception secondary to Rapunzel syndrome: a case report. J Med Case Rep. 2018;12:362. [Accessed on 8 June 2021].

3: Nobori Y, Amano T, Ochi M, Kumasaka T, Sunami E. Rectal cancer developing from an anastomotic site 18 years after resection due to intussusception caused by Peutz-Jeghers polyposis in a 31-year-old man: a case report. Surg Case Rep. 2018;4:110. [Accessed on 8 June 2021].

4: Tavusbay C, Acar T, Kar H, Atahan K, Kamer E. The patients with Peutz-Jeghers syndrome have a high risk of developing cancer. Turk J Surg. 2018;34:162-4. [Accessed on 8 June 2021].

5: Zhao ZY, Jiang YL, Li BR, et al. Close and regular surveillance is key to prevent severe complications for Peutz-Jeghers syndrome patients without gastrointestinal polyps: case report of a novel STK11 mutation (c.471_472delCT) in a Chinese girl. BMC Surg. 2018;18:24. [Accessed on 8 June 2021].

6: Yoshizawa N, Yamaguchi H, Kaminishi M. Differential diagnosis of solitary gastric Peutz-Jeghers-type polyp with stomach cancer: a case report. Int J Surg Case Rep. 2018;51:261-4. [Accessed on 8 June 2021].

7: Tang XB, Zhao JY, Bai YZ. Status survey on enema reduction of paediatric intussusception in China. J Int Med Res. 2019;47(2):859-66. [Accessed on 8 June 2021].

8: Plataras C, Christianakis E, Fostira F, et al. Asymptomatic gastric giant polyp in a boy with Peutz-Jeghers Syndrome presented with multiple Cafe Au Lait traits. Case Rep Surg. 2018;2018:1-3. [Accessed on 8 June 2021].