

Epithelioid Hemangioendothelioma of Ileum Presenting with Intussusception: Report of a Rare Disease

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Case Report

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Abstract: Intestinal obstruction is frequently encountered in emergency and intussusception is one of the etiology. The cause of obstruction differs in adult from pediatric age groups, where intussusception is one of the causes of bowel obstruction. Here, we present an unusual case of adult intussusception with a polyp as lead point and featuring epithelioid hemangioendothelioma on histological examination. Till now epithelioid hemangioendothelioma has not been reported in bowel. We have discussed about pathology and management of this rare disease.

Keywords: Epithelioid hemangioendothelioma; Intestinal obstruction; Intussusception; Small intestinal polyp.

INTRODUCTION

Intestinal obstruction is a common surgical disorder, very frequently encountered in emergency. The cause of obstruction differs in adult from pediatric age groups, where intussusception is one of the causes of bowel obstruction. In, almost all adults, there will be a pathological lead point, benign or malignant, causing intussusception [1]. Here, we present an unusual case of adult intussusception with a polyp as lead point and featuring epithelioid hemangioendothelioma on histological examination. After extensive search in indexed journals and to the best of our knowledge, we could not find any report of epithelioid hemangioendothelioma arising in bowel.

CASE REPORT

A 33 year old woman, homemaker who was referred to our surgery emergency from a private hospital with chief complaints of pain abdomen, bilious vomiting and inability to pass flatus and feces for the last 8 days. There was central abdominal pain, intermittent, colicky in nature, severe in the initial 4 days which later became mild and without change in its character. Patient gave history of fever for the past 4 days. She was amenorrhoeic for the past 5 years and had no comorbidity. General examination BP: 130/87 mm hg, PR: 139/min and SPO₂: 99% with oxygen by facemask. Abdominal examination showed a distended abdomen without tenderness and guarding, tympanic and absent bowel sound. The air entry was decreased in the right basal region of chest. Digital rectal examination showed an empty collapsed rectum without bloodstaining. Clinical diagnosis of acute intestinal obstruction was made. X-ray Abdomen erect film showed multiple air fluid level in a stepwise pattern, X-ray abdomen supine film showed dilated small bowel loops. The blood parameters were within normal limits.

The patient underwent emergency exploratory laparotomy for acute intestinal obstruction. Intraoperatively there was approximately 150 ml of serous fluid intraperitoneally with ileo-ileo caeca intussusception. Telescoping started from the terminal ileum and went in to caecum through the ileocecal valve (IMAGE: 1&2). The intussusception could be reduced manually by gentle retrograde milking from caecum. There was an intraluminal polyp 20 cm proximal to ileocecal junction and 10 cm of terminal ileum appeared unhealthy distal to the polyp (IMAGE: 3). Hence, 20 cm of ileum was resected from 5 cm proximal to the polyp to 15 cm distal to the polyp and end ileostomy with distal ideal mucous fistula was fashioned. The patient was kept on ventilator support in ICU due to her poor chest condition. The patient developed burst abdomen due to SSI on day 4. She was extubated on postoperative day 9 and was shifted to surgical ward from ICU on postoperative day 13. She was discharged on postoperative day 17 with laparotomy wound being allowed to heal by secondary intention. On histopathological examination the polyp showed features of epithelioid hemangioendothelioma and CD-34 was positive on IHC.

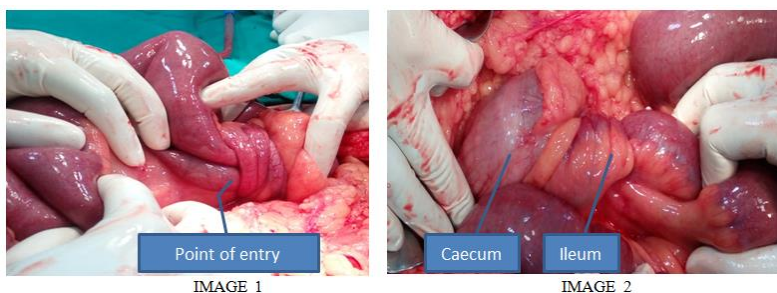


Image-1 & 2: showing ileo ileo caecal intussusception, In IMAGE 1 surgeon's finger pointing to the ileal intussusception and in IMAGE 2 intussusceptions distal ileum and caecum are seen

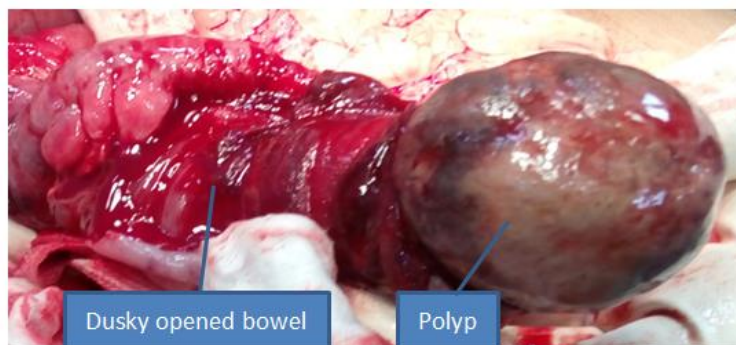


Image-3: showing brownish intraluminal ileal polyps measuring 3×3×3 cm

DISCUSSION

Barbette of Amsterdam first reported 'intussusception' in 1674 [2], and detailed description as 'introssusception' was made by John Hunter in 1789 [2]. Intussusception involves telescoping of bowel segment into adjacent bowel segment leading to obstruction, inflammation, and ischemia [3]. Intussusception is the leading cause of bowel obstruction in children but it is relatively rare in adults where it accounts for 5% of all such cases [3]. Of all the causes resulting in intestinal obstruction in adults only 1-5% is due to intussusception [4]. In children, it is usually primary, benign and pneumatic or hydrostatic decompression is sufficient to treat in 80% of cases [4]. In contrast, almost 90% of adult intussusception is caused by structural lesions such as polyps, benign neoplasms, or carcinoma [4]. In adult intussusception, the lead point is malignant in up to 66% of colonic intussusception and 30% of small intestinal intussusception [5]. In a study of adult intussusception containing 745 cases, 52% were found in the small intestine (39% enter-enteric, 13% ileocolic) and 38% in the large intestine (17% ileocecal, 17% colo-colic, 4% appendicular) [6]. Abdominal CT is the gold standard investigation for intussusception which may show target sign or sausage shaped soft tissue mass. However, these signs on CT are not pathognomonic for intussusception [7]. Excision of segment is practiced due to significant risk of malignancy in adult intussusception [7].

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor originating from vascular

endothelial or pre-endothelial cells with both epithelioid and histiocytic appearance [8]. It represents less than 1% of all vascular tumors and predominantly seen in females with male: female ratio of 1:4. It is usually seen in middle-aged patients and most commonly presents in liver alone (21%), liver plus lung (18%), lung alone (12%) and bone alone (14%) and other sites (35%) including stomach and peritoneum. Distant hematogenous metastases have been reported mainly in the liver and in other sites like skin, serosa, spleen, tonsils, retroperitoneum and kidney; colonic metastases have also been described, but they are very rare [8]. Electron microscopy shows a typical image of endothelial cells similar to those of medium-size vessels, or a large vein, arranged in nests or cords, while immunohistochemistry will reveal Weibel-Palade bodies in the cytoplasm of their cells as typical findings. The typical histopathological feature of a Pulmonary-epithelioid hemangioendothelioma is hyper cellular periphery and hypo cellular center with Coagulative necrosis, Hyalinization, Calcification, Ossification [8]. EHE are positive for Fli-1, CD34, and CD31 in immunohistochemistry. However, none of these markers is 100% specific [9]. There is no consensus on standard treatment due to rarity of this tumor but some authors have recommended surgical resection [8]. Gomez-Arellano *et al.* reported that there is 13% mortality when EHE is located in soft tissue; it increases to 35% when it affects the liver, and 65%, if it reaches the lung [10].

CONCLUSION

Intussusception is a rare cause of intestinal obstruction and adult entity is distinct from pediatric cases in terms of incidence, etiology, and management. As adult intussusception is frequently associated with malignant lesions, we conclude that all of them whenever diagnosed preoperatively should undergo surgical resection of the involved segment and subsequent histopathological examination shall give the final diagnosis and upon which further management would depend.

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