CASE REPORT AND REVIEW OF LITERATURE

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Hemihyperplasia with Multiple intestinal Lipomatosis presenting with intestinal obstruction due to intussusception, challenges in treatment- A Rare case

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ABSTRACT

Hemihyperplasia is a syndrome with overgrowth of limbs and associated with various genomic syndromes. Rarely patient presents with intestinal obstruction when it is associated with sub mucosal lipomas. Submucosal lipomas are common in colon. But whole of small and large bowel studded with them is very rare as in our case. It presents dilemma not only in diagnosis but in treatment also. Here we present a case of forty two years females with hemihyperplasia syndrome came to causality with recurrent intestinal obstruction. On CECT Abdomen diagnosis of intestinal obstruction due to intussusception was made. Whole of bowel is studded with submucosal lipomas. Patient was tried to manage conservatively but patient does not respond and Right Hemicolectomy was done of intussusception segment. Anastomosis leaked and on re-exploration end ileostomy was done. Obstruction due to Multiple Intestinal Lipomatosis (as a part of Genetic Syndrome) should be managed conservatively and if operative intervention is required avoid primary anastomosis.

Key words: Hemihyperplasia; Multiple Intestinal Lipomatosis; Intussusception; Intestinal Obstruction

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INTRODUCTION

Hemihyperplasia is overgrowth of one or more limbs or body parts. It is an interesting identity associated with a number of syndromes. The diagnosis of these disorders mainly is on clinical grounds there being no laboratory test to confirm.[1] Hemihyperplasia is a heterogeneous group of disorders which have hemihyperplasia as a predominant finding, but may be associated with others feature as in Ollier's syndrome, Klippel Trenaunay syndrome, Maffucci Syndrome, Multiple Lipomatosis syndrome etc.[2] Sub mucosal lipomas represent the 2nd most common tumour of colon.[3,4] These may remain silent or may present with complication like bleeding, abdominal pain, bowel obstruction or intussusception.

CASE REPORT

A forty-two years female patient came to Emergency Department on 5th January 2019 of Bhagat Phool Singh Government Medical College with complaints of pain in abdomen and yellow colored vomitus for last 7 days. Pain started in right iliac fossa region with sudden onset and associated with multiple bilious/feculent vomits. Pain was intermittent and colicky in nature and gradually spread to whole abdomen. On examination, visible peristalsis was seen along with lump in right iliac region, which was extending up to right sub hepatic region. Lump was non tender and slightly mobile. Patient was dehydrated but vitally stable. Patient was

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also having Limb overgrowth and multiple Lipomatosis (Figure 1,1a).

Provisional diagnosis of intestinal obstruction was made. She was resuscitated and biochemical and radiological investigations were done. CECT abdomen suggests intestinal obstruction with ileo-colic intussusception and multiple lipomas present throughout small and large intestine (Figure 2).

Emergency exploratory laparotomy was done. Intra operatively, intussusception was present involving ileum, ascending colon and hepatic flexure. It was not reducible, so right hemicolectomy was done along with side-to-side ileo-transverse anastomosis. Whole of small bowel and colon was studded with lipomas. These lipomas were so numerous that it was difficult to take clear margins for anastomosis. On Cut section, tip of intussuscept was gangrenous (Figure 3).

Patient did well for 3 days post operatively and passed flatus and stool on 3rd day. On 4th postoperative day, patient developed tachycardia and blood pressure 90/60 mm of Hg. Patient again advised nil per oral and Ryle's tube was inserted and on 6th day abdominal drain showed stool in it. Again exploration was done on 7th postoperative day and anastomosis leak was present. End ileostomy was fashioned with submucosal lipomas present on ileum and distal colonic end is closed (Figure 4).

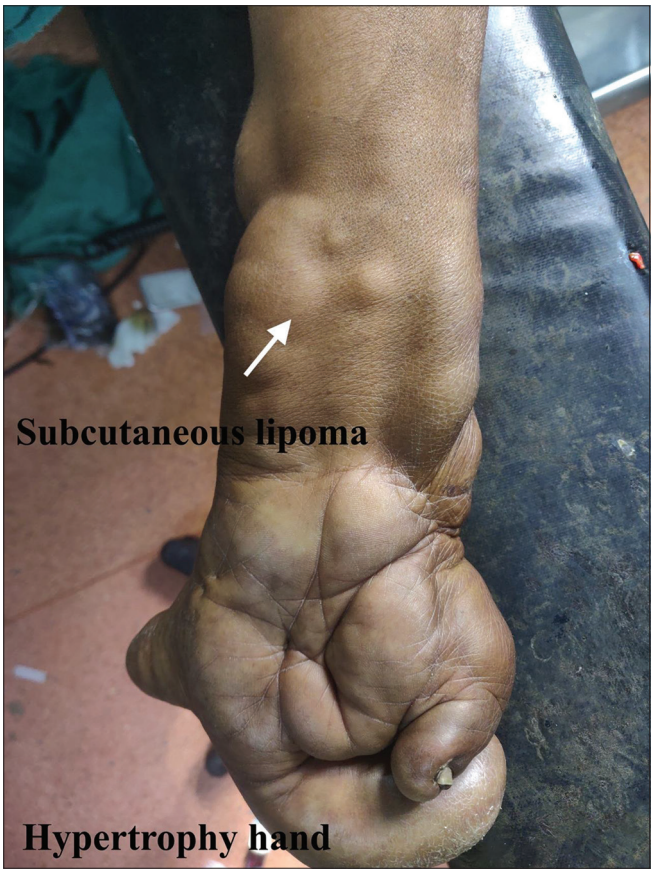

Figure 1: Overgrowth of upper limb with subcutaneous lipoma

Peritoneal cavity was thoroughly washed and abdominal drain kept. Postoperatively, she started complaining of pain in abdomen and ileostomy started functioning intermittently. Patient developed pneumonia and condition started deteriorating with intermittent obstruction along with burst abdomen. Conservative management was continued but patient succumbed on 24th Jan 2019.

DISCUSSION

Hemihyperplasia is a heterogeneous group of disorders, which have hemihyperplasia (overgrowth of one or more limbs or body parts) as a predominant finding. Various clinical criteria had been laid down to label these syndromes. Connective tissue nevi are the hallmarks of Proteus syndrome are not found in present case.[5,6] It is associated with multiple intestinal lipomatosis

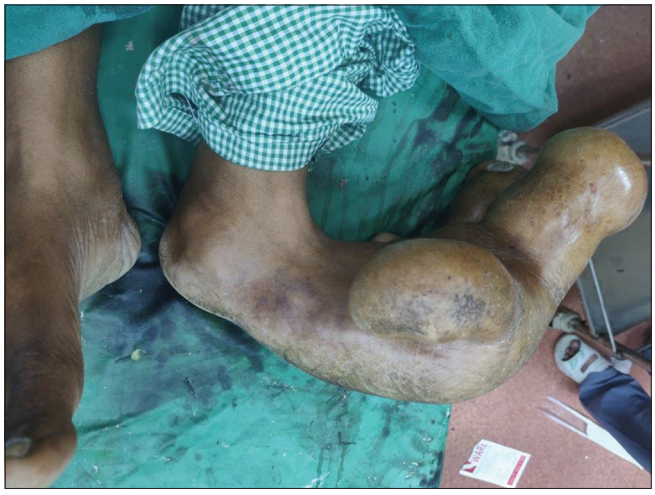

Figure 1a: Hemiperplasia syndrome (Lower Limb)

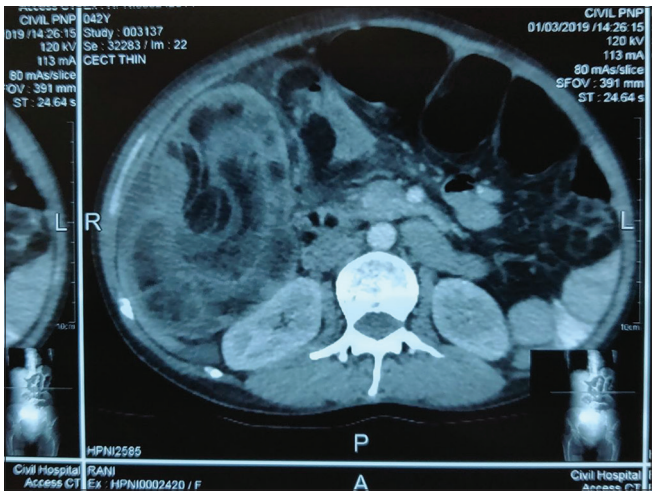

Figure 2: CECT (Abdomen) showing Intussusception with submucosal lipomas

that are present throughout alimentary tract starting from duodenum to colon. Hemihyperplasia-multiple Lipomatosis (HHML) has been described by Biesecker et al.[2] The association may prove to be useful for classifying those patients with moderate asymmetry and overgrowth with subcutaneous lipomatas, who do not fulfill criteria for Proteus syndrome. Present case is not only associated with subcutaneous lipomas but with multiple intestinal lipomas also.

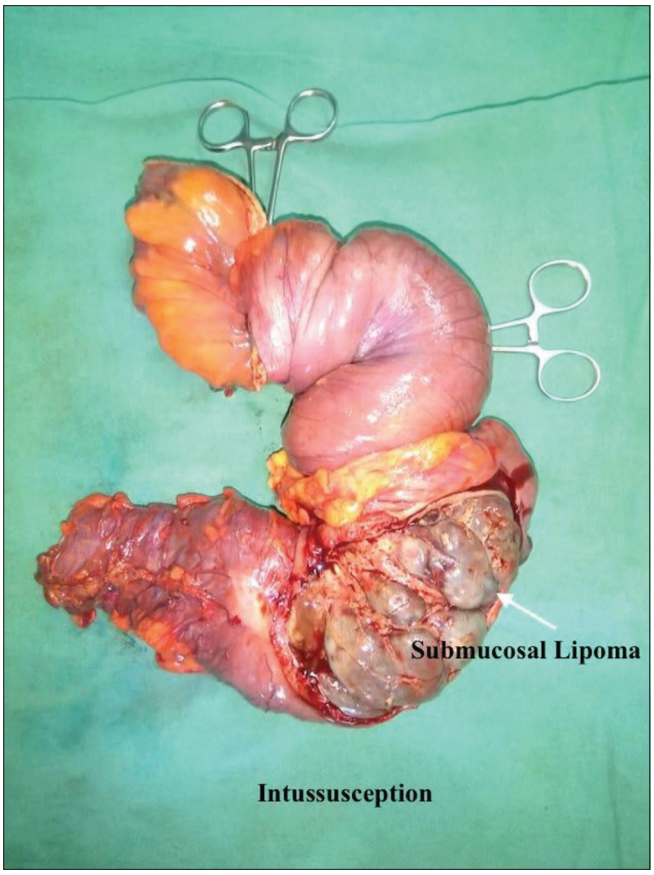

Figure 3: Specimen of right hemicolectomy with multiple submucosal lipomas with Gangrenous part of intussusceptum

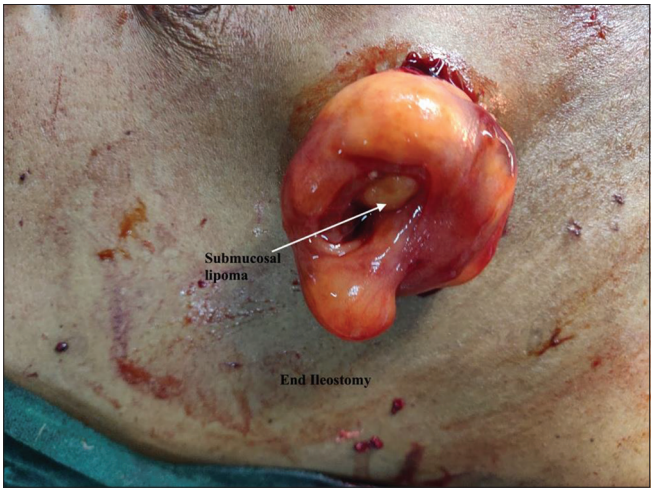

Figure 4: End Ileostomy with multiple submucosal lipomas

Multiple systemic lipomatosis (MSL) is a rare disorder with unknown etiology, characterized by the accumulation of nonencapsulated adipose tissue at face, head, neck, upper and lower extremities, trunk, abdominal cavity, and pelvis. Involvement of gastrointestinal (GI) tract and abdominal cavity is very rare. Intestinal lipomatosis refers to having multiple submucosal lipomas at small intestine and colon.[7]

Although rare, lipomas represent 4% of benign lesions of the gastrointestinal tract and can occur throughout its length. The incidence has been reported between 0.15% and 4.4%.[4,8]

The colon is most commonly affected, followed by the small bowel and stomach. Ninety per cent of colonic lipomas arise from the submucosa.[5] Approximately 275 cases have been reported in literature till date. The most common age of presentation is in the 5th or 6th decade and there is a slight female preponderance.[9]

Most lipomas are found in the right side of the colon. Sarcomatous change in theses lesions has not been reported.[10] Therefore, accurate preoperative diagnosis of these lesions may avoid unnecessary extensive surgery.

Abdominal and intestinal involvements may present with abdominal pain, obstruction, intussusception, and gastrointestinal bleeding. Abdominal lipomatosis might also cause distension, via intermittent obstruction and altering bowel transit time.

As in present case, high suspicion of Multiple Intestinal Lipomatosis is key in diagnosing the condition pre operatively. CECT abdomen is the investigation of choice in suspected cases. Patient is to be managed conservatively until sign of complications (gangrene in intussusceptum- in present case) develop.

TAKE AWAY POINTS FROM THIS CASE

Whole of small bowel studded with submucosal Lipomas. Cases had been reported for Lipomas present in ascending colon. Some in descending colon and transverse colon also. Ours is the case in which Lipomas are present not only in small bowel but at whole of large bowel also.

There is no virgin space available for anastomosis; still anastomosis was done with best possible margin available. So many submucosal lipomas made it difficult for anastomosis but it leaked later on, what to do then??

Single layer anastomosis should be preferred over two layer anastomosis.

CECT abdomen should be done in all cases preoperatively to assess extent of disease and the extent of lipomatosis will determine the extent of resection in emergency.

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