



APPENDICULAR MALTOMA PRESENTING AS INTUSSUSCEPTION WITH THROMBOCYTOPENIA– A RARE CASE REPORT

General Surgery

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ABSTRACT

Background: Mucosa-associated lymphoid tissue (MALT) lymphomas comprise a group of indolent B-cell non-Hodgkin lymphomas, which have a low incidence in clinical practice. Thrombocytopenia generally does not develop in maltoma unless the tumour has infiltrated the bone marrow. Intussusception due to appendicular maltoma is extremely rare. Thrombocytopenia resulting from immunologic causes in patients with gastrointestinal (GI) maltoma have been rarely reported in the literature. **Case report:** 53 year old female presented with complaints of abdominal pain for 1 month along with altered bowel habits, fever, malena and decreased appetite. Abdomen examination showed tenderness in right iliac fossa. Ultrasound and CECT abdomen showed features suggestive of ileo colic intussusception. Exploratory laparotomy was done and it showed appendiculo-caecal intussusception with a hard tumour mass at the base of appendix and hence a radical right hemicolectomy was done. Biopsy report of the resected specimen showed high grade maltoma involving the base of appendix. **Conclusion:** The diagnosis of appendiceal maltoma is difficult in most of the cases. This case report is a very unique presentation of maltoma which adds richness to the limited amount of resources available.

KEYWORDS

adult intussusception, appendicular maltoma, maltoma with thrombocytopenia

I:INTRODUCTION:

Adult intussusception is a rare entity which accounts for 1-5% of intestinal obstructions. Most of the cases of adult intussusception are caused by an underlying pathologic condition such as malignancy, lipomas, polyps, diverticula, strictures, or benign neoplasms. Mucosa-associated lymphoid tissue (MALT) lymphomas comprise group of indolent B-cell non- Hodgkin lymphomas , which are extremely rare. The clinical presentation varies according to the location of the maltoma in the GIT(gastrointestinal tract). The incidence of primary appendiceal lymphoma is 0.015% of all gastrointestinal lymphoma. Immune thrombocytopenia of malignancy is most frequently associated with lymphoproliferative disorders. Thrombocytopenia generally does not develop in maltoma unless the tumour has infiltrated the bone marrow. Cytopenia resulting from immunologic causes in patients with GI maltoma were rarely reported in the literature. We also found only 2 cases of appendicular lymphoma in paediatric age presenting with intussusception in the literature.

II: CASE REPORT:

53 year old female with hypothyroidism, came to our department with complaints of diffuse abdominal pain associated with altered bowel habits, infrequent vomiting , fever, malena, decreased appetite for 1 month. There was no loss of weight or similar complaints in the past. Patient was previously admitted in a private hospital and was treated conservatively. There was no contact with tuberculosis or any previous surgeries in the past. On examination, patient was afebrile and adequately hydrated. Her vitals were normal and abdomen was soft with tenderness in right iliac fossa with no palpable mass. Routine blood parameters were normal except a low platelet count of 78,000. Ultrasound abdomen was done which showed bowel within bowel appearance noted in ileocecal region with target like appearance for a length of 8cms suggesting ileo-colic intussusception. Contrast Enhanced Computed Tomography (CECT) abdomen revealed bowel within bowel appearance in appendiculo-caecal junction giving target sign and a soft tissue density noted in the ileocecal junction- suggesting appendico-caecal intussusception.

Patient underwent exploratory laparotomy which showed an appendiculo-caecal intussusception with mass at the base of appendix around 4x4cms, hard in consistency. Hence radical right hemicolectomy was done in view of the tumour in base of appendix and a single layer end-to-end ileo-transverse anastomosis done. Post operative period was uneventful and patient recovered well. Histopathological examination of the resected specimen showed a grey brown proliferative growth measuring 3.5x 2.5cms located in

caecum at the base of appendix, obliterating its lumen and extending into serosa, features suggestive of high grade maltoma. Patient continued to have drop in Platelet counts, going down to 10,000 cells/mm³ in spite of multiple platelet transfusions. Hemato oncology opinion was sought and advised bone marrow biopsy that failed to show lymphomatous infiltration of the bone marrow.

V:FIGURES:



Figure 1: USG abdomen: Target sign s/o ileo colic intussusception



Figure 2: CECT abdomen: Bowel within bowel appearance – s/o appendiculo caecal intussusception

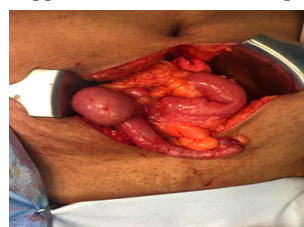


Figure 3: Intraoperatively, intussusception with mass palpated at the base of appendix

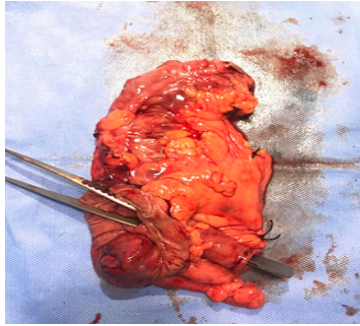


Figure 4: Right hemicolectomy specimen- with mass at base of appendix

III:DISCUSSION:

Intussusception of the bowel is the telescoping of a proximal segment of the bowel within the lumen of the adjacent segment. It is more often reported in children but rare in adults(5%). Most of the cases of adult intussusception are caused by an underlying pathologic condition, such as malignancy, lipomas, polyps, diverticula, strictures, or benign neoplasms. MALT lymphomas comprise a group of indolent B- cell non-Hodkin lymphomas(NHL). The most common site of GI maltoma is the stomach followed by small intestine and colon. MALT lymphoma is a rare cause of intussusception in adults. Thrombocytopenia generally does not develop in maltoma unless the tumour has infiltrated the bone marrow. The literature contains very few reports of cytopenias resulting from immunological causes in patients with MALT lymphomas. We also found only 2 cases of appendicular lymphoma in paediatric age presenting with intussusception in the literature. As in the present case report , maltoma of the base of the Appendix presenting as intussusception with thrombocytopenia in adults is the first case to be described in literature.

IV:CONCLUSION:

Thus appendiceal maltoma is a rare condition and it can also very rarely act as a lead point for intussusception, This should be kept in mind by the practising surgeons. Any suspicion of the lead point being malignant at the time of exploration should dictate a radical procedure. This case report is a very unique presentation of maltoma at the base of appendix which adds richness to the limited amount of resources available.

VI: CONFLICT OF INTEREST: NIL

VII: REFERENCES:

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