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Carcinoid Tumour of the Terminal Ileum-A Case Report

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Abstract: *Carcinoid tumors are classified as neuroendocrine tumours. It is the most common tumour of the small bowel and its incidence is rising. Most carcinoid tumours grow slowly and mostly asymptomatic or may present with nonspecific abdominal pain or carcinoid syndrome.*

We came across one such case who presented with mass per abdomen since 1 year, on examination mass felt in the umbilical region. On admission patient was hemodynamically stable. Exploratory laparotomy segmental resection and anastomosis done. Postoperative period uneventful. Histopathology Section features are suggestive of Ileal ''Carcinoid tumor''.

Age and complete resection were identified as independent prognostic factors for survival in patients with small bowel carcinoid tumors. The importance of achieving R0 resection is therefore emphasized.

Keywords*: Adult Intestinal obstruction, Ileal Carcinoid, complete resection.*

I. Introduction

Carcinoid tumors are classified as neuroendocrine tumours[1]. It is the most common tumor of the small bowel and its incidence is rising. Most carcinoid tumors grow slowly and mostly asymptomatic or may present with nonspecific abdominal pain or carcinoid syndrome[2]. 70% of carcinoid tumors are in the appendix. They often regarded as benign. After appendix small bowel (ileum) is the most common location and has metastatic potential and therefore classified as malignant carcinoids arise from pleuripotent cells in the crypts of leibeurkhunn[3].

II. Case report

A 58 year old female patient was admitted to the department of general surgery with chief complaints of mass per abdomen since 1 year. Mass in the right lower quadrant of abdomen appears intermittently once or twice in a month and there is h/o pain, vomiting and diarrhea when the mass appears. H/o melena present. No h/o loss of appetite or loss of weight. On examination a mass is felt in umbilical region towards right side of size 6*3 cm, shape is ovoid, surface is smooth, Borders are rounded, Consistency is hard, non tender, does not move with respiration. Intrinsic mobility present in both horizontal and vertical movements. No hepatomegaly, no splenomegaly No free fluid in the peritoneal cavity, resonant note is heard over the mass. Normal bowel sounds present.

On admission patient was haemodynamically stable. Routine blood tests are within normal limits. Ultrasound reveals ileo ileao colic intussusception with hypo echoic mass lesion acting as a lead point. Colonoscopy not able to negotiate beyond sigmoid colon.

Exploratory Laparotomy done intra operatively hard nodular intra luminal mass measuring 1x1 cm noticed in the ileum 2 feet away from ileocaecal junction. Tortuous and stellar pattern of arteries in the region of tumour. Mesentery is thickened, fibrous, and there were few velvety adipose lesions in the mesentery. No free fluid in peritoneal cavity. No mesenteric lymph node enlargement. Liver and spleen normal. Segmental resection and end to end anastomosis done.

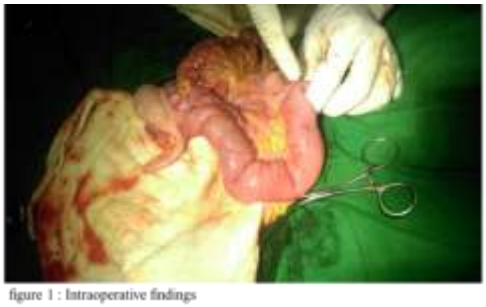

figure 1 : Intraoperative findings

Figure 2 : Gross resected specimen

Patient tolerated the surgery well. Post operative period was uneventful. Patient was discharged Follow up after 15 days.

Histopathology Section studied from ileum shows intact mucosa. The sub mucosa is infiltrated by the tumor cells arranged in trabecular, nests, sheets, rosettes and pseudo glandular pattern. The tumor cells are round to oval with abundant eosinophilic granular cytoplasm and round to oval nucleus have salt and pepper chromatin with inconspicuous nucleoli. Section studied from proximal ileum is free of tumor. Section studied from distal end is free of tumor. Features are suggestive of Ileal "Carcinoid tumor''.

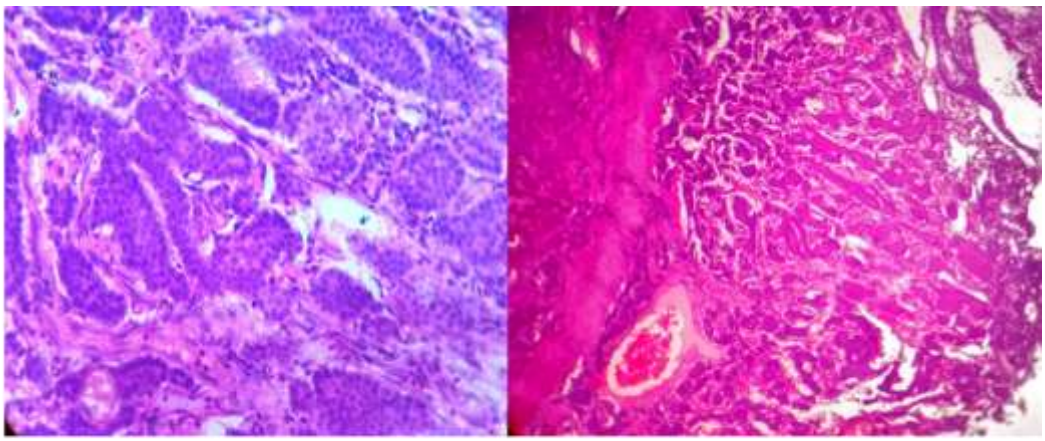

Figure 3 & 4: Histological features are those of Ileal Carcinoid tumour

III. Discussion

Oberndorfer coined the term "karzinoid''[4]. Carcinoids are relatively rare tumors, incidence 4.7:100, 000, median age at diagnosis is63years, with male predominance. They arise from the enterochromaffin cells (kulchitsky cells) found in the crypts of lieberkuhn, also known as argentaffin cells because of staining with silver compounds. These cells are neuro ectodermal in origin and capable of amine precursor uptake and decarboxylation (APUD) and tumours derived from these can secrete biologically active peptides responsible for the carcinoid syndrome[5].

Eighty percent of Carcinoids arise in the gastrointestinal tract, 10% in the bronchus or lungs and others in rare sites including the ovaries, testicles, pancreas, and kidneys.Appendix is the most common site in the GI tract for primary carcinoid tumours, followed by the small bowel.30% of GI Carcinoids arise in the jejunum or ileum and have the most aggressive clinical features[5].

Carcinoid tumours are mostly asymptomatic, Growth of the tumour is very slow, vast majority are < 1cm (5% > 2cm). It grows outward leaving the mucosa intact. As it reaches the serosa it can cause a desmoplastic reaction and lead to kinking of the bowel and intestinal obstruction[5].

Carcinoid syndrome is almost uniquely associated with metastaticmidgut Carcinoids (neuro-endocrine tumours of the gastrointestinal tract). Carcinoid syndrome was first described by Thorson and co-workers in 1954.Systemic symptoms are caused by an excess of biogenic amines, peptides and other factors (serotonin, tachy- and bradykinins and histamine) in the circulation only after liver metastases. Clinical Features of carcinoid syndrome are Cutaneous flushing, Venous telangiectasia, Diarrhoea, Bronchospasm, Cardiac valvular lesions. The patient in this case didn't have symptoms of carcinoid syndrome[5].

Diagnostic proceduresinclude measurements of serum chromogranin A and urinary excretion of 5-HIAA. Urinary levels in excess of 10mg/dl are highly suggestive of diagnosis. Tumour imaging with Transabdominal ultrasound is the initial imaging procedure in most patients with metastasized carcinoids.Contrast-enhanced three-phase CT or MRI followed by needle biopsy for pathology can be performed. In-pentetreotide scintigraphy is positive in 80-90% of patients.Echocardiography is mandatory in patients with carcinoid heart disease[5].

TreatmentincludesSurgery Based on site, size and presence/absence of metastasis. That less than 1 cm with no nodal involvement can be treated with local excision. Those tumours >1cm treated with segmental bowel and mesenteric resection. For tumours with widespread metastasis, surgery is still of benefit. Metastasis to liver can be treated by hepatic artery ligation, Based on the observation that tumor cells get nearly all their nutrients from the hepatic artery. Hepatic artery embolization or chemoembolization occludes the blood flow to the tumor, achieving significant shrinkage in over 80% of cases. Selective internal radiation therapy deliver radioactive microsphere by injection into the hepatic artery. Final surgical option is liver transplantation[5].

Complications include carcinoid crisis an immediate onset of a debilitating and life-threatening condition associated with carcinoid syndrome. May occur spontaneously or may be precipitated by anaesthesia,

chemotherapy, infection, stress, catecholamines, and tumour manipulation or embolization procedures. Symptoms include prolonged severe flushing, diarrhoea, hypotension, tachycardia, severe dyspnoea, peripheral cyanosis and sometimes hemodynamic instability. Appropriate precautions include immediate therapy and close monitoring before, during and after surgical treatment. Carcinoid crisis prevented by prophylactic administration of octreotide given by continuous intravenous infusion at a dose of 50 μg/h for 12 hours prior to and at least 48 hours after the procedure to prevent a cardiovascular carcinoid crisis[6].

Medical therapy is directed to patients with malignant carcinoid syndrome and those with widespread metastasis where surgery not possible. Somatostatin analogues (octreotide, pasiriotide) given subcutaneously 2 or 3 times daily or intravenously with total parenteral nutrition. It decreases circulating levels of serotonin and urinary 5-HIAA and cause relief of symptoms (diarrhoea + flushing) and also tumour regression (17%) [5].

Radiotherapy has no benefit. Chemotherapy can achieve partial response 30-40%.Cytotoxic chemotherapy agents used Streptozocin + 5-FU/ Cyclophosphamide (33%), Doxorubicin, cisplatin[5].

Best prognosis of all small bowel tumours. Resection of localized tumours 100% survival rate.65% for patients with regional disease.25-35% for patients with distant metastasis[5].

IV. Conclusion

Age and complete resection were identified as independent prognostic factors for survival in patients with small bowel carcinoid tumours. The importance of achieving R0 resection is therefore emphasized.

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