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Case Report

Primary malignant melanoma of the duodenum: a rare case report

Villalan Ramasamy*, Anand Lakshmanan, Senthilkumaran, Govindaraj Raman, Kannan Devigounder

Department of Surgical Gastroenterology, Institute of Surgical Gastroenterology, Rajiv Gandhi Government General Hospital, Madras Medical College, Chennai, Tamil Nadu, India

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***Correspondence:**
Dr. Villalan Ramasamy,
E-mail: rexvillalan@yahoo.co.in

ABSTRACT

Malignant melanoma especially primary melanoma is uncommon in the gastrointestinal (GI) tract. GI melanomas are usually metastatic from skin melanoma. Anorectal region and small bowel are the most common sites of primary and metastatic GI melanoma respectively. Various theories have been put forward to explain the origin of Primary Melanoma of Small intestine. Melanomas of the GI tract can produce range of symptoms based on their location and size. Surgery is the main stay of treatment for localized disease. We reported a case of primary duodenal melanoma which was managed by surgically.

Keywords: Duodenum, Primary melanoma, Pancreaticoduodenectomy

INTRODUCTION

Primary malignant tumours of small intestine are rare; most of the small bowel tumours are metastatic and less than 2 %originate in the small bowel.[1] Melanoma originates from melanocytes which are usually found in skin, retina, anal canal and meninges. Most of the melanomatous lesions of the Gastrointestinal tract (GI) are metastases from the skin. Anorectal region and small bowel are the most common site of primary and metastatic GI melanoma respectively. At autopsy, small bowel is involved in 58% of patients with metastatic melanoma.[2] Primary small bowel malignant melanoma, particularly of duodenal origin is rare with paucity of published reports. Overall GI melanoma has poor prognosis in comparison with cutaneous melanoma because of the good blood supply of mucosa and late detection.[3] Here we report a case of large Primary duodenal melanoma in a middle-aged patient.

CASE REPORT

A 43 years old male presented with abdominal pain, vomiting and palpable lump in the right side of the abdomen. He had jaundice with loss of weight and passing high coloured urine of 4 months duration. No history of fever, anorexia, haematemesis or radiating pain was elicited. He had no previous history of surgery. Routine hematological and renal function tests were normal. His Liver function test showed elevated Bilirubin (Total Bilirubin 10, Direct 7) and serum alkaline phosphatase (305) and liver transaminases were within normal limits. Upper GI scopyrevealed Multiple raised blackish spots starting from the duodenal bulb extending upto third part of duodenum. Ampulla appeared prominent and ulcerated with central blackish pigmentation. Endoscopic biopsy was negative for malignancy. Magnetic Resonance imaging showed a 11*9.5*7.3 cm hetereogenously enhancing exophytic T2

heterointense lesion with haemorrhage and necrotic components noted arising from medial wall of second part of duodenum compressing the ampullary region causing dilatation of proximal biliary system and main pancreatic duct, suggestive of Gastro intestinal Stromal tumour (GIST) (Figure 1).

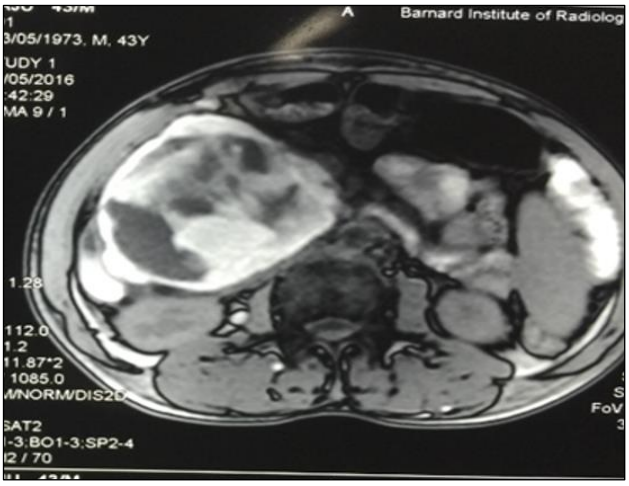

Figure 1: 11*9.5*7.3 cm hetereogenously enhancing exophytic t2 hyperintense lesion with haemorrhage and necrotic components noted arising from medial wall of second part of duodenum.

Ultrasound guided biopsy revealed features suggestive of Malignant melanoma. A complete history and detailed examination of skin, eyes, anal canal and meninges did not reveal any primary lesion. An exploratory laparotomy was performed through a midline incision. Intra operatively a 11*10 cm Lobulated growth arising from the medial wall second part of duodenum splaying the duodenum. The Mass was abutting the superior mesenteric vein and involving the head and neck of pancreas. A dilated Common bile duct with multiple peripancreatic nodes nodes were present. Liver and spleen were normal. No evidence of free fluid or peritoneal deposits. In view of the above findings, Pancreaticoduodenectomy (Whipples procedure) was performed (Figure 2).

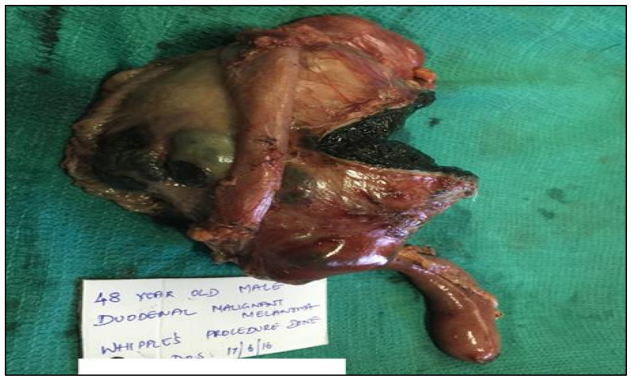

Figure 2: Pancreaticoduodenectomy (Whipples procedure) specimen.

Post-operative course was uneventful. Post-operative Histopathology of the resected revealed duodenal mucosa with ulceration and a underlying neoplasm composed of sheets of spindle to polygonal cells with moderate eosinophilic to densely pigmented cytoplasm containing melanin with round to ovoid nucleus and prominent nucleoli. The tumour is infiltrating the underlying pancreas and suggestive of malignant melanoma of duodenum (Figure 3).

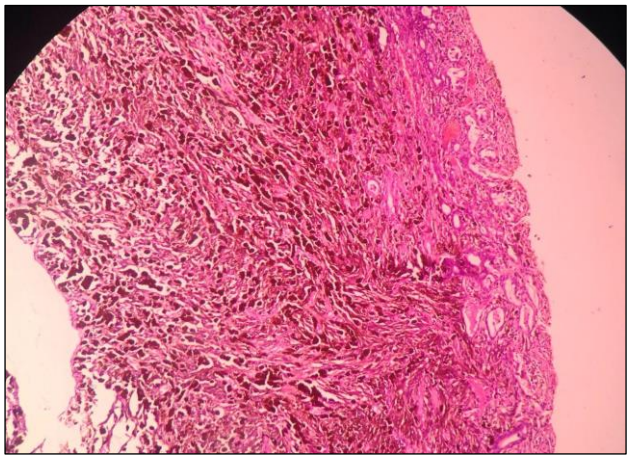

Figure 3: HPE duodenal mucosa with ulceration and a underlying neoplasm composed of sheets of spindle to polygonal cells with moderate eosinophilic to densely pigmented cytoplasm containing melanin with round to ovoid nucleus and prominent nucleoli.

His postoperative course was uneventful and discharged on 9 typist operative day. On follow up patient is doing well with no evidence of locoregional recurrence or distant metastasis.

DISCUSSION

Small bowel tumours are rarer than tumours of stomach and colon. Various explanations have been offered regarding the lower incidence of small bowel tumours but the most plausible one is effective local immune response.[4]

Malignant melanomas originating from melanocyte, is not a common tumour and accounts for 1-3% of all malignant tumours of GI tract. Metastasis to the GI tract is seen most frequently in the small intestine followed by colon, stomach, rectum and esophagus. In the GI tract, melanoma can also be found in the rectum and sigmoid colon by the local migration of primordial skin melanocytes.

Primary melanoma originating in the small bowel, particularly in the duodenum is extremely rare and the presence of such an entity is itself very controversial. Different theories have been offered to explain the origin of Primary Melanoma of Small intestine. First theory postulates that small bowel melanomas are thought to

originate from melanoblastic cells of the neural crest, which migrate to the distal ileum through the umbilico mesenteric canal.[2] Second theory states that Melanomas may potentially originate from APUD cells, although they do not produce any hormones. Based on APUD theory, Ileum is the most common site of tumour which represents the distal end of umbilici mesenteric canal.[5] Few studies deny the very existence of primary melanoma of the GI tract. Some authors states that primary tumour regress before the occurrence of metastasis or is too small to be identified.[6] We together with other authors claim that primary small bowel melanoma does exist.[7]

Thorough systemic examination must be done to rule out possibilities of metastasis from other sites such as skin, retina, anal canal, esophagus, penis and vagina. Primary small bowel melanoma is an extremely rare lesion which must be differentiated from other intestinal tumours. Melanoma itself is a great mimicker of other neoplastic lesions and may create diagnostic challenge when presenting as intra-abdominal tumour. So, it should be confirmed by histopathological examination.[8] Majority of metastatic lesions are asymptomatic. Although 60% of the patients who died from malignant melanoma had metastasis in some part of the GI tract, only 4% had this diagnosis during their life time.[9]

Clinically melanomas of the GI tract can cause wide range of symptoms depending on their location and size. The clinical feature of this malignancy is the same as of any other primary tumours of small intestine. They usually present with symptom such as abdominal pain (70%), anemia (20-50%), Loss of weight (50%), Palpable mass (25%), Intussusception, Massive Rectorrhagia and perforation.[10] It is very difficult to differentiate between a primary GI melanoma and a metastatic melanoma of the GI tract from a unknown or regressed cutaneous primary melanoma. According to Blecker, primary melanoma of small intestine is diagnosed when there is a lack of concurrent or previous removal of melanoma or melanocytic lesion from the skin and lack of any other organ involvement and in situ change in the overlying or adjacent epithelium.[11]

Primary GI melanomas are recognized histologically by in situ changes in the overlying and adjacent GI epithelium depicting atypical melanocytic cells in the basal layer of epithelium and extension in a pagetoid fashion in to the more superficial epithelium. Younger patients have very poor prognosis because of very aggressive tumour biology and rapid metastasis. In elderly patients the course is somewhat indolent and less metastatic.[12] Surgical treatment is the standard of care for both primary and metastatic disease because chemotherapy, Radiotherapy and immunotherapy cannot offer definitive treatment. The tumour should be resected with sufficient Proximal and distal margin and regional lymphadenectomy . The median survival after curative resection of Primary malignant melanoma of the small intestine is 48.9 months and the longest survival has been reported as 21 years.[6,13] Our case met all the following criteria for a Primary malignant melanoma 1). Large single lesion located in Duodenum 2). Thorough history, clinical examination, endoscopic assessment and radiological imaging failed to reveal any other lesion 3). meets Blecker criteria and Confirmed by histopathological examination. Only few published data claims that radical surgery i.e. whipples procedure offers both symptom palliation and long-term survival for primary melanoma of duodenum.[14]

CONCLUSION

Inconclusion, primary malignant melanoma of the Duodenum is an extremely rare condition with varied presentation. Strong clinical suspicion and a thorough evaluation, to rule out a primary melanoma in other organ systems, lead to the diagnosis of primary GI melanoma. Surgery alone remains the primary modality of therapy both for cure and for palliation of symptoms with an improved quality of life.

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