Case Report

Primary jejunal malignant melanoma presenting as intussusception: a rare case report

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ABSTRACT

A 60-year-old male patient presented with complaints of pain abdomen and vomiting. Clinical examination revealed a mass in hypogastrium extending up to right iliac fossa. Exploratory laparotomy showed a jejuno-jejunal intussusception with lead point mass lesion presenting as sub-acute intestinal obstruction. Per-operative findings revealed a blackish mass. A provisional diagnosis of melanoma was made, which was subsequently confirmed on histopathology and immunohistochemistry. Gastrointestinal (GI) melanomas are rare tumors, involving ileum and anus. Jejunum is a rare site amongst the small intestinal melanomas. Intussusception is common in the pediatric age group. We report a rare case of primary jejunal malignant melanoma, presenting with sub-acute intestinal obstruction due to jejuno-jejunal intussusception. Primary melanomas have a pre-dilection for males, involve ileum and have a poor prognosis than the metastatic GI or primary cutaneous melanomas. Surgery is the treatment of choice.

Keywords: Melanoma, Small intestine, Intussusception, Immunohistochemistry

INTRODUCTION

Tumors of a small intestine are rare, mostly malignant and commonly metastatic in nature.^{1,2} Commonest primary tumors arising in the small intestine are adenocarcinoma, carcinoid, lymphomas and gastro-intestinal (GI) stromal tumors.³ Malignant melanomas arising from skin and eye have propensity for a wide spread metastasis, involving any organ in the body including GI tract (GIT).⁴ Although small bowel involvement by malignant melanoma is most frequently metastatic, a primary origin at this site has been reported in rare instances. Primary melanomas of the GIT involve particularly the ileum and anus.

Around 1% of all bowel obstructions are due to intussusceptions.^{5,6} It is a rare occurrence in the adult population accounting for <5% of all intussusceptions.⁶ Surgery is the treatment of choice since the etiology

involves an underlying pathologic, malignant process that acts as a lead point.⁵⁻⁷ Diagnosis is invariably made post-operatively.

CASE REPORT

A 60-year-old male patient was admitted to the surgical department with complaints of pain abdomen and vomiting, which were insidious in onset. General examination revealed severe dehydration. All routine investigations including serological tests were within normal limits. Local examination of the abdomen revealed a palpable, oblique - oval mass measuring 10 cm × 4 cm extending from hypogastrium up to right iliac fossa, across the umbilicus. Swelling had well defined borders, bosselated surface and is intrinsically mobile. A clinical diagnosis of sub acute intestinal obstruction was made.

X-ray - erect abdomen showed multiple dilated small bowel loops with air fluid levels. Ultrasonography (USG) abdomen revealed jejunal loop along with the mesentery projecting into the jejunal loop with 'Whirlpool sign' with a 3 cm × 2.5 cm hypoechoic lesion noted at the tip of intussusception with color uptake suggestive of a lead point. Dehydration was corrected.

Exploratory laparotomy showed dilated proximal bowel loop with jejuno-jejunal intussusception with blackish mass as the lead point, about 40 cm from duodenojejunal flexure along with multiple enlarged mesenteric lymph nodes. Reduction of intussusception was done, and 30 cm of jejunal segment was resected with 10 cm of margin of jejunum on either side of the lead point. End to end anastomosis done and abdominal drains were placed. Specimen was submitted to the department of pathology for histopathological examination. Post-operative period was uneventful.

Gross examination

Received a jejunectomy specimen measuring 30 cm in length. Cut opened specimen revealed a jet black, intraluminal polypoidal mass measuring 5 cm × 5 cm × 3 cm, at a distance of 17 cm from the proximal resected margin, occluding the lumen (Figure 1). Few small satellite nodules were observed in the adjacent mucosa. Rest of the bowel and resected margins were unremarkable. A total of 10 lymph nodes were dissected ranging in size from 0.5 to 2 cm in diameter in the mesentery. Cut sections of these lymph nodes revealed blackish tumor deposits. Multiple sections were taken from the intestinal segment including tumor and lymph nodes.

Histopathological examination

H and E stained sections revealed all the four layers of native jejunum showing focal ulceration with tumor arising in the mucosa, extending up to serosa. Tumor is cellular, arranged diffusely in a vague fascicular fashion and in sheets, composed of oval, spindle-shaped to polygonal cells having characteristic brown intracytoplasmic pigment with a vesicular nucleus and prominent eosinophilic nucleoli in most of the cells (Figure 2). Mitoses were appreciated. Focal areas of necrosis and hemorrhages were seen.

Sections from all the lymph nodes revealed tumor deposits having similar histological features as described above. Sections from both resected margins were free from tumor infiltration.

A provisional diagnosis of malignant melanoma of jejunum with secondary metastatic deposits in mesenteric lymph nodes was considered.

Panels of immunohistochemistry markers were done which showed positivity for S100, HMB45, Melan-A and negative for Pan CK and p63.

It was advised to search for primary in the skin, eye, esophagus and anus.

On follow-up, a thorough clinical examination and investigations including upper GI endoscopy, proctoscopy and fundoscopy were done to rule out the possibility of primary melanoma arising from esophagus, rectum, anus and eye. Computed tomography (CT) scan of chest and brain, USG of liver, bone scintigraphy were performed to rule out metastatic deposits from cutaneous melanoma. There was no history of removal of any pigmented skin lesion or its spontaneous regression.

A final diagnosis of primary jejunal malignant melanoma with metastatic melanoma deposits in all the 10 regional mesenteric lymph nodes was confirmed.

DISCUSSION

Although small bowel accounts for 75% of the length and 90% of the mucosal surface of GIT, the incidence of primary malignant GI tumors constitutes for 3% at this site. Metastatic tumors are more common, arising from lung and breast. Approximately two-thirds of small bowel tumors are malignant and arise mostly in jejunum. More than 95% of these primary tumors are constituted by adenocarcinomas, carcinoids, lymphomas, and GI stromal tumors.³

Malignant melanomas account for 1-3% of all malignant lesions of the GIT. Small intestinal melanomas can be primary or metastatic. Primary intestinal melanoma is extremely rare. Metastatic melanoma of the small bowel

Figure 1: Intra-operative images. (1) Segment of J-J intussusception after reduction with mesenteric lymph nodes. (2) Cut open jejunectomy specimen - intraluminal polypoidal mass and lymph node. (3) Cut section of bowel mass and of lymph nodes showing blackish tumor with metastatic deposits respectively.

Figure 2: Histopathology images. (4 and 5): Intestine with native mucosa and tumor with brown pigment (H and E, ×4 and ×10). (6) Tumor with nuclear pleomorphism and prominent eosinophilic nucleoli (H and E, ×40). Atypical mitoses noted. (7-9) Secondary metastatic deposit of melanoma in the lymph node (H and E, ×4, ×10 and ×40). (10 and 11) H and E, HMB45 stains showing native mucosa along with tumor (×10). (12 and 13) H and E, HMB45 stains showing tumor infiltrating the muscle coat (×10).

is more common, because of the tendency for cutaneous melanomas to metastasise to the GIT. Other primary sites of melanoma that can metastasize to the small bowel are from eye, esophagus and anus. Symptoms are nonspecific and include weight loss, abdominal pain, GI bleeding, mass, intestinal obstruction, and intussusception.⁸ Non-specific symptomatology, inaccessibility of jejunum and ileum to routine endoscopic procedures and their low diagnostic yield, pose a challenge for early diagnosis. High-resolution CT scan and CT-enteroclysis are helpful in detecting small intestinal melanomas.

Morphology

Morphologic growth patterns of intestinal melanomas include: Intra-luminal/polypoidal obstructing mass, cystic/cavitary, target lesions, infiltrating and exocentric/intra-mural lesions with fistulous tract.

Histologically, the tumors show a variety of architectural patterns like fascicles, diffuse sheets, and nests, pseudo-glandular and around the blood vessels. Various cell types seen in melanomas are spindle cell, epithelioid, mixed and pleomorphic. Microscopically GI melanomas may be pigmented or amelanotic.

Among the differential diagnosis of GI melanomas, especially of amelanotic variant are clear cell sarcoma and GI stromal tumor.

Primary small intestinal melanomas have a worse prognosis than the metastatic GI/cutaneous melanomas.⁹

Various theories have been proposed regarding the origin of small intestinal primary melanomas like from intestinal schwann cells,¹⁰ melanocytes in the GIT or from neural crest cells.^{11,12} These multi-potent cells migrate into the bowel via the umbilical-mesenteric canal,¹¹ where they later differentiate into specialized cells. In the gut, neural crest cells differentiate into amine precursor uptake and decarboxylation cells, which can undergo neoplastic transformation and produce tumors such as carcinoids, gastrinomas and also melanomas.^{11,12}

It is difficult to distinguish between primary and secondary small intestinal melanomas. Criteria for diagnosing primary jejunal melanoma include: Absence of melanomas in other primary sites like skin, eye, upper or lower GI tract. No history of removal or surgery or spontaneous regression of any pigmented lesion/melanoma/atypical melanocytic lesions from the skin or other organs.

Secondary melanomas of the small intestine are multiple, present with non-specific symptoms, are in-accessible to routine endoscopic investigative modalities and are thus diagnosed late. Growth patterns of secondary or metastatic intestinal melanomas are varied and present as extra luminal, intra-mural lesions, polypoidal, target lesion, cavitary, infiltrating and exo-enteric growth with fistulous tract.¹³

Primary intestinal melanoma should be solitary, intra-luminal mass, with no other metastasis, excepting for regional secondary mesenteric lymphnodal deposits and a disease free survival period of atleast 12 months after the diagnosis without any evidence of recurrence.^{14,15}

As the primaries present with obstructive symptoms, they are diagnosed earlier, but have a wide spread metastasis due to the rich vascular and lymphatic supply and thus have a worse prognosis.

CT scan of chest and brain, USG of liver, bone scintigraphy should be done to rule out possible metastatic deposits from cutaneous melanoma and upper GI endoscopy, proctoscopy and fundoscopy to rule out primaries at these sites.

TP63 is a homologue of TP53. TP63 has an antiapoptotic role in melanoma and is responsible for mediating chemoresistance. Expression of p63 in both primary and metastatic melanoma clinical samples significantly correlated with melanoma-specific deaths in these patients.¹⁶ p63 over expression is associated with poor prognosis.

CONCLUSIONS

GIT melanomas are usually metastatic in origin. Primary intestinal melanomas are rare, solitary, intra-luminal mass, having aggressive course, with widespread dissemination and worse prognosis. Surgery is the treatment of choice.

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