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REVIEW ARTICLE

PRIMARY MALIGNANT MELANOMA OF SMALL INTESTINE PRESENTING AS INTESTINAL OBSTRUCTION

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ABSTRACT

Primary malignant lesion of gut is a rare tumor, only few cases have been reported in the litreature for which different criteria have been mentioned by different authors. We report a case of 70 years old female admitted with complaints of pain and abdominal distention presenting as intestinal obstruction. She was operated in emergency and found to have intussusception hence segmental intestinal resection was performed. Gross, microscopic, histochemical, Immunohistochemical finding confirmed the diagnosis of malignant melanoma. No other primary site was found on clinical examination. On follow up secondaries were found after 10months in the liver and retroperitonium, which was confirmed by FNA.

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INTRODUCTION

The small intestine is a common site of metastasis because of its rich blood supply. Only few cases of primary malignant melanoma have been reported in the small bowel. It is commonly believe that most cases of malignant melanoma are metastatic, originating from an occult primary cutaneous and ocular lesion (Chaung *et al*., 2008). In most of the cases diagnosis is difficult to establish because symptoms are nonspecific and the tumors are usually undetectable by gastroscopy or colonoscopy. Its diagnosis is difficult and important because at the time of presentation the patient is already in advanced stage and has poor prognosis. This is the case report of a primary small intestinal malignant melanoma, complicated by intestinal obstruction which is due to intussusception caused by growth.

Case Report

A 70 year old female presented with pain abdomen and distention, diagnosed as a case of intestinal obstruction, in emergency at govt. hospital in the year 2006. X-ray showed multiple fluid and gas level. The case was operated in emergency. On laparotomy small gut was intussuscepted hence segmental resection along with draining lymph nodes, were done. The post operative period was uneventful and patient was recovered in time. Grossly the specimen of small intestine ms 10cm in length and have a polypoidal growth ms 3x2x2 cm. c/s is grey brown to black (Fig-1). Microscopy shows highly anaplastic polypoidal growth envolving the wall of the small intestine The cells are polygonal to spindle, arranged in sheets, acinar and trabecular pattern. Nucleoli are large eosinophilic and shows abundant mitotic figures. There were focal aggregates of brown-black pigment intra as well as extracellularly (Fig-2, 3&4). Special stains prussian blue excludes the haemosiderin pigment while massons-fontana favours the melanin pigment.IHC was positive for HMB-45 and S-100. The case was followed up and after 10 months she devloped mild hepatomegaly and left sided lumber mass.USG revealed a metastatic lesion while guided FNAC showed metastatic melanoma.

DISCUSSION

The primary malignant melanoma is relatively a rare tumor comprises of 1-3% of all tumors and it is present at different sites primarily on the skin, orbit, transition zone between skin and mucosa. In GIT the primary sites are the oropharynx and nasopharynx (32.8%), anal canal (31.4%), rectum (22.2%), oesophagus (5.9%), stomach (2.7%), small intestine (2.3%) and gallbladder (0.9%) (Chaung *et al*., 2008) Except the oesophagus and rectum where melanocyte normally exist, primary malignant melanoma of GIT is controversial as the small and large gut doesn't contain melanocyte hence some authors believe that primary malignant melanoma of GIT is unlikely (Declore and Friesen, 1993). However there are few concept mentioned in the literature which favours the primary

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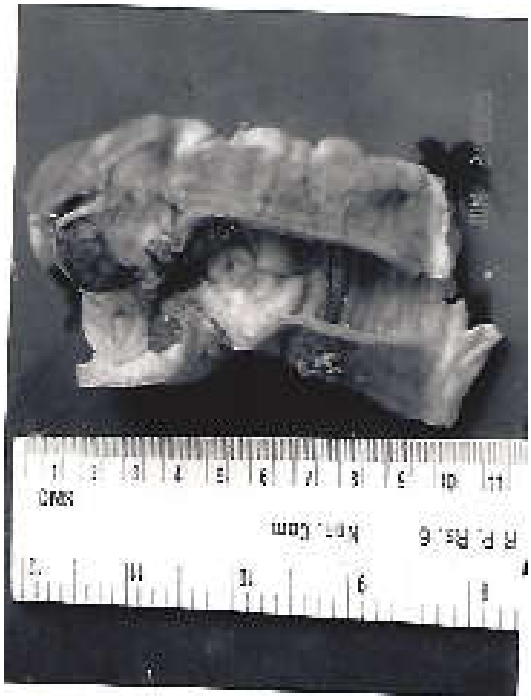

Fig. 1. Gross of resected ileum showing pigmented polypoidal growth

etiology of the small bowel melanoma like-1. melanocyte are present in the alimentary tract as well as in the respiratory tract and lymph node (Kruger *et al*., 2005) 2. It may arise from neural crest cells. These cells are multipotential cells which became amine precursor uptake and decarboxylation (APUD) cells, migrate through the body and come to the intestine via umblical -mesentric canal where they differentiate into APUD cells. Their neoplastic transformation leads to production of tumors like carcinoid and gastrinomas. Melanomas may potentially orignate from APUD cells although they don't produce any hormones. According to APUD theory the ileum which represent the distal end of the umblical mesentric canal should be the most common site of primary malignant melanoma of the small intestine (Kruger *et al*., 2005). The case has been presented here because of its rarity and unusual presentation. Patient presented to us with abdominal emergency due to a mass which is present in the small intestine and lead to intussusception and later on diagnosed as malignant melanoma by routine, histochemistry and and immunohistochemistry. For the diagnosis of small intestinal malignant melanoma as a primary lesion, different diagnostic crieteria was proposed from time to time. According to Sachs *et al*. (2004) these are:

1. Presence of biopsy proven melanoma cells in the small intestine at single focus.
2. No evidence of disease in any other organ and absence of draining lymph nodes.
3. Disease free survival of at least one year after diagnosis (Blecker *et al*., 1999)
4. In situ changes in the overlying or adjacent GIT epithelium (Christova *et al*., 1996) This feature is recognised histologically by the presence of atypical melanocytic cells in the basal layer of the epithelium and extending in a pegetoid fashion into the more superficial epithelium reported in 40-100% of primary melanoma (Logondianakis *et al*., 2006)

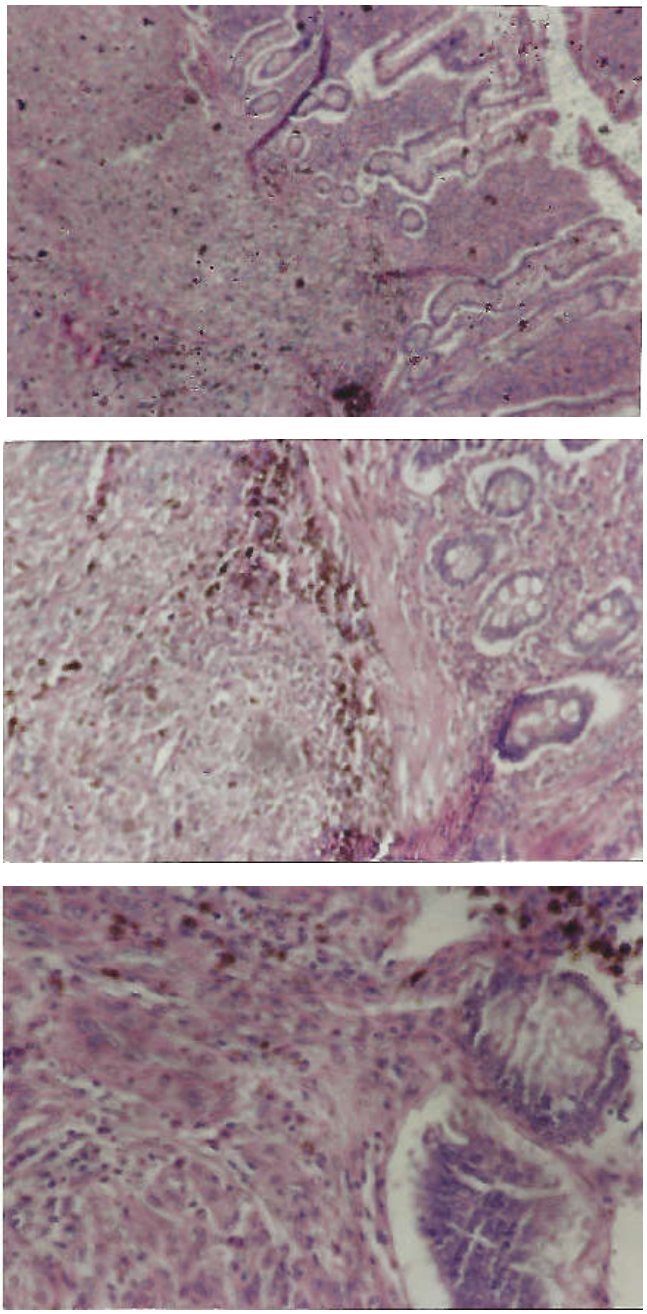

Fig- 2,3 & 4 Histopathology of the growth showing mucosal pigmented malignant cells infilterate the lamina propria (4x, 10x & 40x)

Melanomas arising from the mucosal surfaces appears to be more aggressive and are associated with worse prognosis, compared with the cutaneous melanomas. The poorer prognosis may be associated with the (8) delay in diagnosis, more aggressive behavior, earlier dissemination through lymphatics and vascular supply.

Conclusion

Considering the above mentioned criteria, the primary malignant melanoma may be exist in this particular case, diagnosed histopathologically and confirmed by immunohistochemistry favouring the concept of APUD cell origin.

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