CLINICOPATHOLOGICAL FEATURES OF ANAPLASTIC CARCINOMA OF SMALL INTESTINE: A RARE CASE REPORT

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ABSTRACT

In the small intestine, anaplastic carcinoma is a rare variant with few cases reported in literature. We report a case of a 55-year-old male who presented with intussusception and lower abdominal pain. Microscopically, the tumour showed a diffuse growth pattern unlike conventional adenocarcinoma. The present case discusses the histomorphology of anaplastic carcinoma of small intestine, its immunohistochemical profile and the differential diagnosis of this rare but highly aggressive tumour.

KEYWORDS

Anaplastic Carcinoma, Intestine, Intussusception, Immunohistochemistry.

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INTRODUCTION: In small intestine, the Anaplastic Carcinomas are very rare, with an incidence of 0.5 to 0.8 per 100,000 population per year.[1] In small intestine, the Anaplastic carcinoma is a very rare variant, arises most frequently in ileum and distal jejunum.[2] The anaplastic carcinoma follows more aggressive clinical course with high mortality as compared to conventional carcinoma of small intestine. So identification of this rare variant is essential. Here, we report a case of anaplastic carcinoma of the small intestine.[3]

This case report highlights the histomorphological features of this rare and unusual variant of intestinal adenocarcinoma. To arrive at an accurate diagnosis, use of immunohistochemistry is very essential as the tumour morphology mimics other diseases. Providing early diagnosis and optimal clinical management is essential for improving prognosis and survival as the tumour has highly aggressive nature.

CASE HISTORY: A 55-year-old male presented with a history of intestinal obstruction and intermittent lower abdominal pain. Ultrasound of abdomen and CT scan of abdomen showed intussusception, marked thickening of intestinal wall. An exploratory laparotomy was performed which revealed a polypoid tumour measuring 11 cm × 6 cm × 3 cm extending through full thickness wall of ileum.

Gross findings: Ileal segment measuring 11 x 6 x 3 cm, polypoidal growth identified at 8 cm from one resected end, growth partially cut opened measuring 5.5 x 3 cm. Rest of mucosae appears normal (Figure 1).

Fig. 1: Gross Photograph showing Tumour Measuring 8 cm x 5 cm x 4 cm with Polypoid Growth

Microscopic Findings: The tumour tissue arranged in diffuse sheets and discreetly placed. Most of the areas had round to oval to polygonal cells arranged in sheets with focally dissociated cells. The tumour cells had abundant eosinophilic cytoplasm. The nuclei were round to oval with clumped chromatin and 0-2 prominent nuclei (Figure 2).

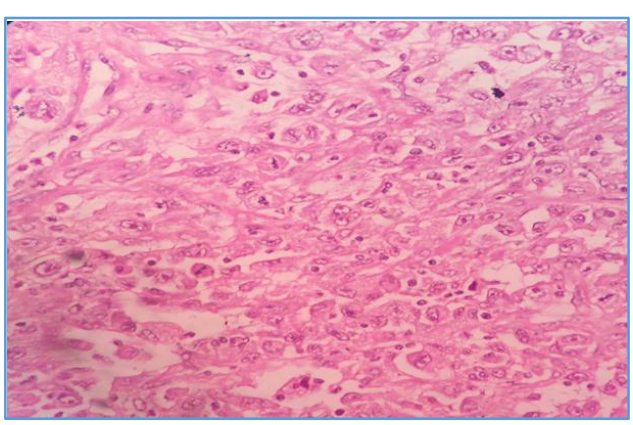

Fig. 2: Microphotograph showing Tumour Cells arranged in Sheets with Tumour Cells having Vesicular Nuclei and Prominent Nucleoli

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Differential diagnosis of malignant epithelioid GIST, leiomyosarcoma, amelanotic melanoma and diffuse large B cell lymphoma (DLBCL).

Immunohistochemistry showed diffuse positivity for both cytokeratin (Pan-CK and CK-20) (AE1-AE3) (IT-Ks20.8) and vimentin (V9) in all tumour cells, in all stages of differentiation (Figure 3 & 4). Focal positivity for epithelial membrane antigen (EMA) was also seen. However, LCA (CD45), CD30, CD10, CD99, BCL2, CDX2, MUC1, Desmin, Chromogranin, DOG1, Alk-1, S-100, CD117, CEA, SMA, HMB-45 and CD34 were all found to be negative.

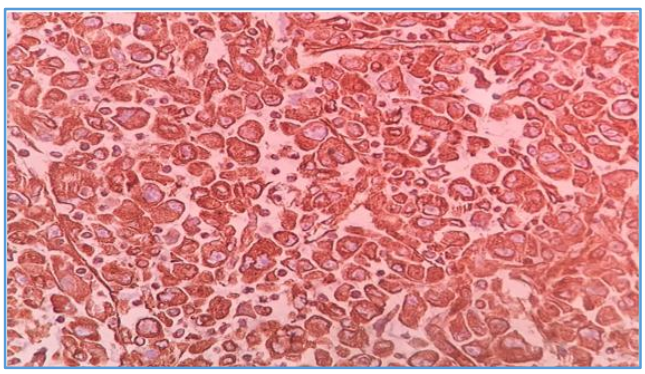

Fig. 3: Microphotograph showing Vimentin Positivity in Tumour Cells

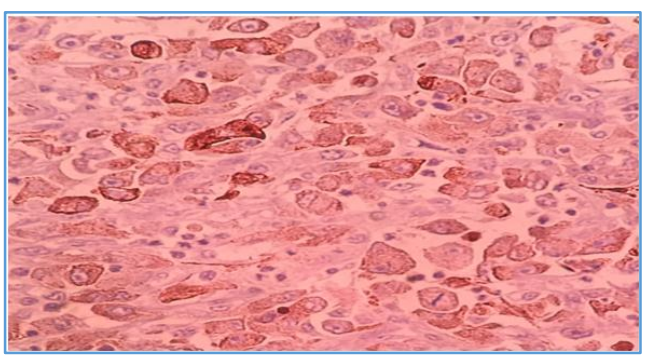

Fig. 4 Microphotograph showing Cytokeratin Positivity in Tumour Cells

DISCUSSION: In the small intestine, anaplastic carcinomas are very rare with only 18 cases reported in literature till date.[4] 58 years (range, 38-81 years) is the mean age of presentation with a slight male predominance, M: F ratio 1.5:1.[5] Ileum is the most common location for these tumours followed by jejunum and rarely duodenum, presenting as endophytic tumours.[5] Anaplastic carcinoma may appear biphasic or monophasic on routine histological examination with a mixture of epithelial and mesenchymal cells.[5] The epithelial component may be arranged in sheets, clusters or glands like structures. The mesenchymal component consists of spindle cells with tapering cytoplasm. In both morphologically different areas, nuclei are round to oval, vesicular with prominent nucleoli. Foci of anaplasia and tumour giant cells may be seen. Sometimes anaplastic cells may predominate to the exclusion of spindle cell component. Such tumours have been called pleomorphic giant cell carcinoma.[6] Our case showed a predominantly epithelial component with focal spindle cell areas. Our case showed similar findings. Immunohistochemical, anaplastic carcinomas showed positivity for CK (75% of cases), focal positivity for EMA and diffuse positivity for vimentin (90% of cases).[5] Typically, conventional small intestinal and colorectal adenocarcinomas are positive for CK20 and negative for CK7.[7]

Differential diagnosis of anaplastic carcinomas of small intestine includes anaplastic large cell lymphoma (ALCL), epithelioid gastrointestinal stromal tumour (GIST) and metastatic carcinomas as these tumours also showed round to oval to polygonal cells arranged in sheets with vesicular nuclei and prominent nuclei. Multinucleation can also be seen. Hence, use of a wide immunohistochemical panel becomes essential for arriving at correct diagnosis. ALCL shows positivity for LCA, CD30 and Alk-1. No epithelial dysplasia or mucin is seen. Our case was also negative for LCA, CD30 and Alk-1. GISTs are negative for CK and positive for CD117 and CD34. Our case showed CK positivity and negativity for CD117 and CD34.

To conclude, in the small intestine, anaplastic carcinomas are extremely rare, with aggressive clinical course and extensive local spread. The patient who has undergone surgical procedure like complete tumour resection continues to have a poor prognosis with 3-year survival rate of 10.8% and median survival time of 5 months, despite treatment by surgical procedures.[4] Most of the patients with anaplastic carcinoma are in their late stages and presentation with obstructive symptomology. Anaplastic carcinomas are best treated by complete surgical resection; unlike lymphomas and GISTs, its closest differentials, which are amenable to chemotherapy and radiotherapy respectively, anaplastic carcinomas when treated with chemotherapy and radiotherapy are not having any significant effect on the rate of survival.[8] No specific type of mutations have been detected in the tumour which would allow for the development of specific targeted therapies. Hence, clinical awareness of this entity is essential for early diagnosis and appropriate treatment and management thereby increasing the survival rate and reducing disease morbidity and mortality.[4]

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