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Mesenteric fibromatosis mimicking GIST and presenting as a rare cause intestinal obstruction in a 1 year old boy: A rare case report

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ABSTRACT

Mesenteric Fibromatosis (MF) is a rare myofibroproliferative tumour that may present as a common clinical entity like intestinal obstruction or abdominal mass. They are exceedingly rare in the pediatric population. They often mimic Gastrointestinal Stroma Tumours (GIST) in their presentation, imaging and even histological features. Here we present the case of a one year, 9 months old boy who had a right hemicolectomy for an obstructing ascending colon tumour whose initial histological analysis was suggestive of GIST. A repeat analysis including Immunohistochemistry staining confirmed the diagnosis of MF. No further treatment was given and the child is asymptomatic one year later. MF though rare may present as common clinical entities and tends to mimic MF. Detailed analysis is required to distinguish it from GIST as the prognosis and treatment may differ significantly.

Keywords— *Mesenteric fibromatosis, Gastrointestinal stromal tumour, Intestinal obstruction*

1. BACKGROUND

Mesenteric Fibromatosis (MF) belongs to a spectrum of rare tumours called Desmoid Tumours (DT) that occur in 2-4 cases per million population. They account for 0.03% of all tumours and 3% of all soft tissue tumours. DTs are myofibroblastic proliferative and infiltrative tumours that lack metastatic potential. MF is the commonest of these otherwise rare intra-abdominal desmoid tumours [1].

Although generally rare, these mesenteric fibromatoses may present as common clinical entities e.g. abdominal mass or intestinal obstruction thus presenting a diagnostic dilemma in these situations. They are very rare in the paediatric age group [2]. They often mimic the more common Gastro-Intestinal Stromal Tumours (GIST) in clinical presentation, imaging findings and even histology [3]. It is important to distinguish MF from GIST because there is a distinct difference in their prognosis and several aspects of their management. Here, we present a rare case of intestinal obstruction caused by MF in a one year old boy, which was initially diagnosed as GIST.

2. CASE PRESENTATION

A 1 year, 9 months old boy of African origin presented at the age of 1 year and 3 months with features of intestinal obstruction at a hospital in his home country. He underwent an exploratory laparotomy in which an obstructing ascending colonic tumour was discovered and a right hemicolectomy was performed. This led to resolution of symptoms and the patient was discharged in good condition after an uneventful post-operative hospital stay.

Five months later, he presented with features of intestinal obstruction similar to the previous episode. An exploratory laparotomy was performed but it did not identify any obvious mechanical obstruction. The patient was then referred to our institution for further assessment with a clinical and histopathological diagnosis of GIST. He was stabilized and investigated. An abdominopelvic CT scan revealed dilated loops of small bowel with air fluid levels but no obvious transition or mass lesion seen. The patient was managed non-operatively with a resolution of the obstructive symptoms and later discharged in stable condition.

Histopathology was repeated at our institution on the tissue block available from the first laparotomy. This showed colonic tissue with a myofibroblastic proliferation on the serosal surface entrapping the adipose tissue. The lesional cells were elongated with fine nuclear chromatin, small nucleolus and pale eosinophilic cytoplasm and loose myxoid stroma. Ropy Collagen bundles were noted. Nuclear atypia and necrosis were not seen. The lesional cells expressed SMA and did not express c-kit, DOG-1, Desmin, S-100 protein, Alk-1 & CD 34. Findings are consistent with the diagnosis of MF.

A multidisciplinary discussion decided in favour of a watchful waiting strategy and the Child is asymptomatic one year after the first resection of the obstructing mesenteric tumour.

3. DISCUSSION

MF is the commonest of the otherwise rare Desmoid tumours. MF is exceedingly rare in the paediatric population with a peak age of presentation between 5-8 years; making this case of MF in a one year old an exceedingly rare occurrence. MF may present as a progressive abdominal mass or less commonly as an infiltrating tumour leading to intestinal obstruction as seen in this case [2]. On the other hand, diagnosed tumours may have an indolent clinical course or even spontaneously regress as has previously been reported.

MF is a myofibroploriferative tumour that may proliferate and infiltrate the intervening segment of the gastrointestinal tract, thus mimicking primary intestinal tumours in clinical presentation and radiology. The proliferation and infiltrating fibrosis may lead to a significant narrowing of the intestinal lumen producing intestinal obstruction as seen in this case.

MF commonly mimics the more common GIST in clinical presentation, imaging and even histology [3]. In one study, up to 52% of MF cases were initially diagnosed histologically as GIST [4]. Histology is often inadequate, necessitating more elaborate immunohistochemical staining to clinch the diagnosis.

At histology, the presence of infiltrating elongated/wavy spindle cells with scant cytoplasm and intervening collagen bundles or myxoid stroma is typical of MF. Mitotic activity is low. No necrosis was seen. The intervening muscles may show regeneration and giant cell formation. This is in contradistinction to GIST which has pushing rather than infiltrating spindle cells with frequent mitoses and cellular atypia. Immunohistochemistry (IHC) of MF is usually positive for SMA and nuclear Beta catenin but negative for C-Kit, Desmin, S 100 and CD 34. The opposite holds true for GIST [3, 4]. The typical findings at histology accompanied by consistent IHC, in this case, led to the confirmation of a diagnosis MF as opposed to GIST.

Surgical excision with negative margins (R0 resection) where feasible remains the best treatment option [5]. Tumour free margin is the strongest predictor of long term outcomes of treatment, especially tumour recurrence. Other predictors include multifocal disease or having surgery as the precipitating factor [6]. Recurrence may occur in 30-76% of cases [5]. Our patient had a right hemicolectomy with negative margins (R0 resection). The absence of other foci of disease or surgery as precipitating factor all confer our patient a low risk for recurrence according to the currently available literature.

In our patient, a Multidisciplinary team discussion between the surgeon, radiologist, pathologist, oncologist together with child's parents opted for a watchful waiting strategy going forward. The role of adjuvant therapy for R0 resections is not yet established. The child remains asymptomatic one year after the initial surgery.

4. CONCLUSION

MF though rare may present as a cause of a common clinical entity e.g. intestinal obstruction and should, therefore, be considered among the less common differential diagnoses. It is critical to conclusively distinguish MF from GIST as their prognosis and management may differ significantly. A multidisciplinary approach is necessary for successful management of MF on which literature is still scanty.

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