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Original Research Paper

Pathology

A RARE CASE OF A PRIMARY MESENTERIC FIBROMATOSIS IN A PERIPHERAL HEALTH CARE CENTRE

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ABSTRACT **Background:** - Mesenteric fibromatosis (MF) is a monoclonal, myofibroblastic, neoplastic proliferations that tend to show a locally aggressive behaviour, which is prone to aggressive local recurrences, but lacks metastatic potential. Primary or spontaneous MF is rare and occurs in the absence of any predisposing conditions. **Case presentation:** - We report a case of a 35-year-old female who presented with abdominal pain associated with no other symptoms. Her per abdominal examination revealed a firm mobile mass on palpation in the mid-abdomen. Her computed tomography scan revealed an 8.3cm x 7.8cm x 4.7cm non-enhancing hypodense lesion in the hypogastric region. She underwent laparoscopic surgical excision of the abdominal mass which was sent for histopathological examination. The microscopy showed proliferating fibroblasts along with entrapped skeletal muscle fibers, nerve bundles, and a few blood vessels, with no atypia, mitosis, or necrosis, findings were consistent with mesenteric fibromatosis. **Conclusion:** - Mesenteric fibromatosis is a rare tumour that generally grows at a slow rate and is asymptomatic. The final diagnosis depends on the histopathology, which remains the gold standard for diagnosis.

KEYWORDS : Mesenteric Fibromatosis, Soft tissue tumor, Intra-abdominal Mass.

INTRODUCTION: -

Mesenteric fibromatosis (MF) or intra-abdominal desmoid tumor is a rare neoplastic, monoclonal myofibroblastic proliferative disease affecting the mesentery. They account for 0.73% of all abdominal tumours with an incidence rate of 2-5/ million/ year. Despite its rarity, mesenteric fibromatosis is the most common mesenteric tumour [1]. Although mesenteric fibromatosis account for less than 10% of sporadic fibromatosis, 70% are intra-abdominal, and most involve the mesentery. They can arise spontaneously after surgical trauma or abdominal surgery [2]. The deep fibromatosis is classified by anatomic location, the first group usually originating intraabdominally (mesenteric, pelvic, retroperitoneal fibromatosis), the second arising from the deep soft tissues of the abdominal wall (abdominal fibromatosis), and the third originating within extra-abdominal soft tissues (extra-abdominal fibromatosis) [3]. The objective of this report is to present a case of mesenteric fibromatosis in a female patient without any of the predisposing factors.

Case Report: -

We report a case of a 35-year-old female who presented with complaint of vague abdominal pain of six months duration. Her past medical history was unremarkable and there was no notable family history. She had no alterations in her bowel or bladder habits. Abdominal examination revealed diffuse tenderness with a mobile mass on palpation in the mid-abdomen. Her computed tomography scan revealed an 8.3cm x 7.8cm x 4.7cm non- enhancing hypodense lesion in hypogastric region extending along the vertebral body of the first lumbar vertebra to the fifth lumbar vertebral body which was suggestive of a mesenteric cyst. She underwent laparoscopic surgical excision of the abdominal mass under general anaesthesia. The postoperative course was uneventful.

Pathological Findings: -

Gross: - Gross examination revealed a globular soft tissue mass measuring 10 cm in diameter, externally it was capsulated and grey white in colour with a glistening surface. Cut surface was grey white, homogenous coarsely trabeculated with firm consistency. [Fig.1]

Microscopy: - Microscopy showed long fascicles of myofibroblasts arranged in parallel with uniform spacing, in a hypocellular collagenous stroma proliferating fibroblasts along with entrapped skeletal muscle fibers, nerve bundles, and a few blood vessels. The cells have small ovoid nuclei with punctuate nucleoli and indistinct cell boundaries. Pleomorphism and necrosis are not seen, findings were consistent with mesenteric fibromatosis.

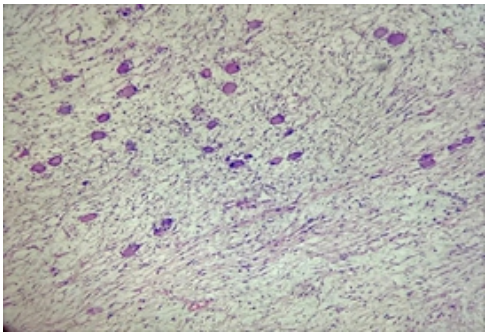
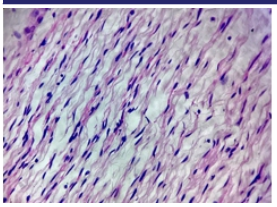


Fig. 2: Photomicrograph Showing fascicular growth pattern of fibromatosis with interlacing bundles of fibroblasts along with entrapped skeletal muscle fibers separated by variable amounts of collagen (H and E, 10X).



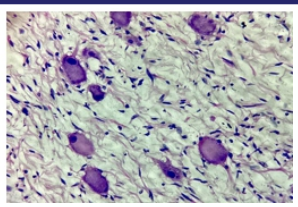


Fig. 3: Photomicrograph Showing long sweeping fascicles with elongated spindle cells of uniform appearance with pale cytoplasm and tapering nuclei. (H and E, 40X).

Fig. 4: Photomicrograph Showing hypocellular collagenous stroma proliferating fibroblasts along with. entrapped skeletal muscle fibers. (H and E, 40X).

DISCUSSION: -

Mesenteric fibromatosis of the intestine is the most common site, followed by the omentum and mesocolon [2]. They are classified into three types: abdominal, extra-abdominal, and intra-abdominal. Mesenteric fibromatosis is rare, locally aggressive, and arises in the mesentery, peritoneal ligaments, retroperitoneum, or pelvis and can become very large. Extra abdominally these are found in the limbs, pelvis, and shoulder girdle [4]. The exact etiopathogenesis of MF is unknown; but various associated factors are (a) Trauma, (b) Estrogen as a growth factor (c) Gardner's Syndrome (d) Crohn's disease (e) Pregnancy (f) Familial adenosis polyposis (FAP) [3]. Mesenteric tumours have an increased incidence in patients with FAP or Gardner syndrome, in whom intra-abdominal fibromatosis develops in up to 15% [5]. Most are asymptomatic, but few present with abdominal mass, abdominal pain, fatigue, vomiting. Mesenteric desmoid infiltrates adjacent organs and causes important complications like intestinal obstruction, ischemia and perforation, hydronephrosis, ureteric fistula, and even aortic rupture [6]. Ultrasonography, Computerised tomography and Magnetic resonance imaging scan are imaging modalities commonly used to diagnose MF [7].

Differential diagnoses of MF include essentially Gastrointestinal stromal tumours (GISTs) and Adult type fibrosarcomas. MF can occasionally mimic GISTs arising from the mesentery or peritoneum because of their morphological similarities [8]. Correct identification of the lesion is of most clinical importance because MF and GISTs are widely diverse processes from biologic, clinical, prognostic, and therapeutic standpoints [9]. MF on gross is greyish and on cut section is homogenous and glistening whereas GISTs are soft in consistency and fatty. These tumours can also be differentiated by their areas of haemorrhage and necrosis. [2]. While GISTs are known to show positive staining for CD117, CD34, DOG 1, and PDGFRA, MF are characterized by positive beta catenin expression and lack of CD34 expression. Hence, staining for beta-catenin and CD34 is recommended whenever there is diagnostic doubt between MF and GISTs [8]. The management of both tumours is completely different. Mesenteric fibromatosis is treated by surgical resection. Treatment of GIST is surgery along with Tyrosine Kinase inhibitors [10].

Adult type fibrosarcomas are a spindle cell type of soft tissue sarcoma, characterized by a solitary, soft to firm, fleshy, rounded or lobulated mass that is grey, white to tan-yellow and measures 3 to 8 cm in greatest dimension. Small tumours are usually well circumscribed and are partly or completely encapsulated. Large tumours are less well defined; they often extend with multiple processes into the surrounding tissues or grow in a diffusely invasive or destructive manner [11]. On microscopy it shows Neoplastic cells arranged in long sweeping fascicles that form a classic "herringbone" pattern. Cells are elongated, spindle shaped with unipolar or bipolar cytoplasm and oval nuclei. Adult fibrosarcomas show a mild to moderate degree of pleomorphism. Fibrosarcomas display varying degrees of mitotic activity. There is a variable amount of stromal collagen present within fibrosarcomas and may mimic fibromatosis in some tumours. Vimentin is a marker that is indicative of a mesenchymal cell origin and is often the only positively stained marker in the diagnosis of fibrosarcomas [11].

The differential diagnosis for intraabdominal fibromatosis also includes sclerosing mesenteritis, a lesion also sometimes referred to as mesenteric panniculitis and mesenteric lipodystrophy. Sclerosing mesenteritis typically involves the small bowel mesentery and presents as a large solitary mass, multiple lesions or diffuse mesenteric thickening may also be seen. Histologically, sclerosing mesenteritis is composed of variable amounts of fibrosis, chronic inflammation, and fat necrosis. IHC staining for -catenin is useful because mesenteric fibromatosis consistently shows strong nuclear -catenin staining and sclerosing mesenteritis does not express this antigen [11].

In our case, complementary therapies were not suggested as the tumour was mesenteric fibromatosis and complete resection of tumour was done. As noted in our case, most of these lesions require complete resection. The patient has shown no signs of recurrence 6 months after surgery.

CONCLUSION: -

Mesenteric fibromatosis is a rare tumour that generally grows at a slow rate and is asymptomatic. Several risk factors may not be present in all cases. Computed tomography and magnetic resonance imaging are the preferred methods for the diagnosis of this condition. The final diagnosis depends on the histopathology, which remains the gold standard for diagnosis.

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