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Intra Abdominal Aggressive Fibromatosis: A Rare Soft Tissue Tumor

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Abstract: ***Abdominal fibromatosis is rare locally aggressive benign soft tissue tumour result of monoclonal fibroblastic proliferation derived from deep connective tissue characterised by high local recurrence rate and no metastasis. This is a case report of 28 year male with complaints of abdominal distension with mild dull aching pain since 5 months. CECT suggestive of large mesenteric mass extending from subdiaphragmatic space up to pelvis. Histopathological studies confirmed it to be a desmoid tumor. The mass was surgically excised and patient was discharged with stable vitals. On followup 12 months until now there is no recurrence reported.***

Keywords: abdominal fibromatosis, benign tumor, desmoid tumor, surgical excision, no recurrence

1. Introduction

Abdominal Fibromatosis (AF) also known as desmoid type fibromatosis is uncommon locally invasive tumor with estimated annual incidence of two to four new cases per million people in general population. Giant mesenteric DTs are even rare [(1)] AF can affect almost all parts of body, most commonly affected locations are abdominal wall neurovascular bundle of extremities, mesenteric root and head and neck. We report a case of gaint desmoid tumor arising from root of mesentry which was surgically resected due to mass effect.

2. Case Presentation

A 28 year man presented with complaints of progressive abdominal distension associated with dull aching pain for 5 months. On physical examination abdomen was grossly distended and mass was felt extending from xiphisternum to pelvis which was firm, non tender, non mobile. Moderate pitting edema was present on bilateral lower limbs. Bladder and bowel habits were normal. There was no history of fever, chills, weight loss. There was no family history of colon cancer or FAP and no personal history of trauma. CECT revealed large ill defined heterogeneous density lesion in left lumbar region extending from periumblical region to suprapubic region displacing stomach and bowel loops on right side, abutting tail and body of pancreas, greater curvature of stomach and spleen. Size of the lesion 31 x 18 x 37 cm.

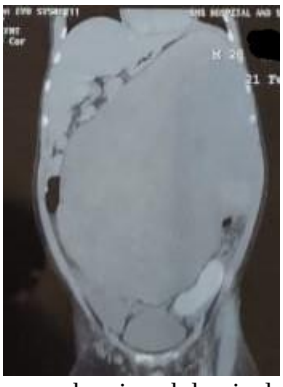

Figure 1: CT image showing abdominal mass extending from left hypochondrium to pelvis

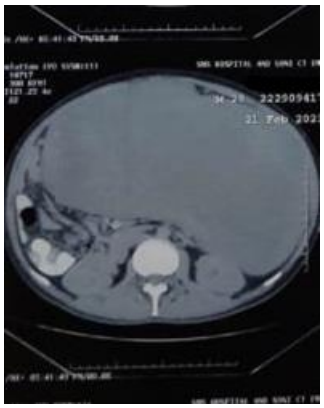

Figure 2: Tansverse section

Histopathological examination of USG guided biopsy revealed desmoid tumor. Patient underwent explorative laparotomy and the mass was found to be attached to mesentery displacing whole of the gut to one side. Mass was excised in toto. The dimension were 30 x 15x 35 cm and it weighed 24 kg.

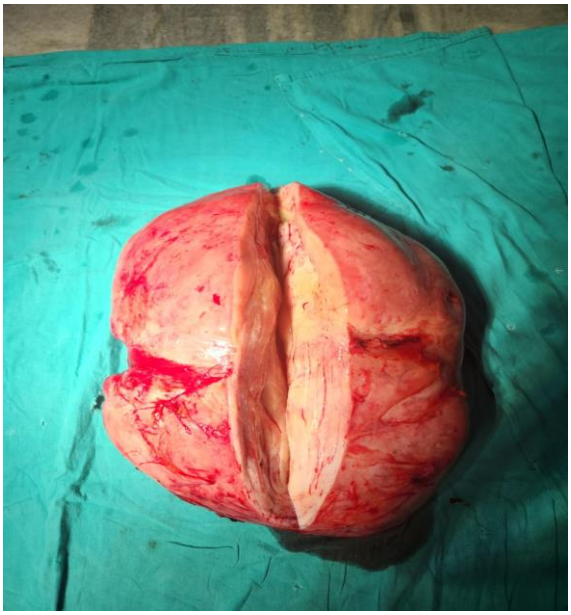

Figure 3: Cut section of gross specimen

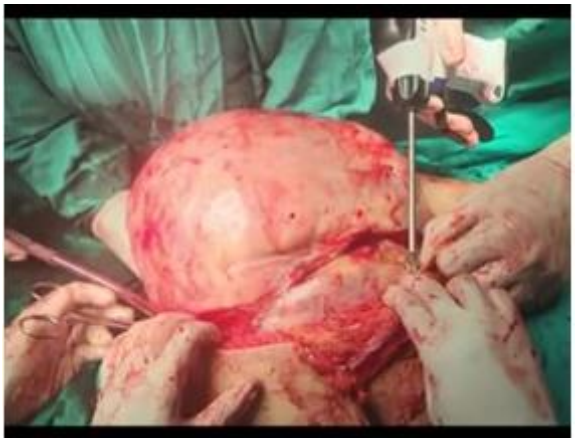

Figure 4: Intra operative image

External surface was smooth and lobulated. On cut section it appeared smooth, whitish, less vascular and firm to touch all findings in favour of benign pathology [Figure 3]. Macroscopic pathological examination revealed spindle cells. IHC of tumor cells positive for vimentin and beta catenin and negative for CK, SMA, CD34, CD117, Desmin and calretinin.

Patient's postoperative course was uneventful. Patient was discharged on post op day 4 with stable vitals. On follow - up 4 months until now there is no recurrence reported.

3. Discussion

AF are rare and involve a monoclonal fibroblastic proliferation derived from deep connective or muscle tissue, which is characterized by a benign pathology, invasive growth, a high local recurrence rate and no metastasis. The incidence of AF is very low (~4–6 cases/1, 000, 000 population per year) and accounts for only 0.03% of all tumors and 3% of soft tissue tumors. [(1)]

AF commonly develops between the age of 15 and 60 years (average: 30 years) and it is more likely to involve female patients (male: female ~1: 3). The 5 - year recurrence is ~50%, and recurrence is related to age, tumor location and resection margin status. Intra - abdominal AF is the least common subtype (~15%) and typically involves the small mesentery and retro peritoneum, with multiple lesions observed in ~10% cases.

The aetiology and pathogenesis of AF remain unclear. The first set of factors include genetic. In one the studies it was found that about 7.5% cases were associated with FAP, especially patients below 60 years with abdominal wall or intra abdominal desmoid tumour. Moreover, the Gardner syndrome is associated with a higher local recurrence rate than the other AFs.

The second set of factors is trauma or surgery, as ~30% patients with AF have a history of trauma [(2),] which indicates that the wound healing process may induce fibroblastic proliferation and promote AF formation. In one of the studies 37.5% patients had history of abdominal surgery or trauma.

Clinical presentation of AF can be variable in many cases it can go asymptomatic chronic progression leading to abdominal distension only as in this case. In some cases, it may lead to obstruction, perforation or symptoms realted to specific organ involvement. In some cases, the AF may stabilize or regress over time, with ~20% AF cases exhibiting natural regression [(3)]. This variable clinical course may also indicate the need for non-surgical modalities of treatment.

For investigation ultrasonography, CT and MRI are the most common modalities. USG reveals round or oval, smooth, solid soft tissue mass with variable echogenicity. However, due to lack of its specificity it is only the primary modality and not the investigation of choice. CT and MRI can provide more detailed anatomical information, which is critical for assessing tumor resectability and planning any surgical treatment. The first choice for diagnosis and evaluation is typically CT, especially for intra - abdominal AF, which generally reveals a soft - tissue mass that is slightly denser than the skeletal muscle, with intra - tumor collagen components that can exhibit high density. Mild - to - moderate enhancement can be observed, while necrosis and calcification are rare. [(4)]

MRI provides better resolution of soft tissue and is recommended for evaluation in cases involving extra - abdominal AF and recurrence. Heterogeneous MRI signals are commonly observed because the AF contains spindle cells, collagen fibers and extracellular matrix, which typically exhibit an isointense signal on T1WI and a hyperintense signal on T2WI. Richer cellular components are associated with higher signal. There is also a significant enhancement in the arterial phase and delayed enhancement in the portal venous phase, which is more obvious when the tumor has a higher proportion of cellular components. [(5, 6)]

Surgery remained the main modality of treatment but the variable biological behaviour of tumor indicates for other treatment modalities like active surveilliance as this tumor may remain stable or may undergo natural regression. Surgery and radiotherapy remaining local treatment method for symptom relief. In one of the cohort studies of 771 patients no major difference was found in overall survival of patients between the surgery and non surgery groups of patient with no or mild symptoms.[(7)]

Surgery remains the main modality of treatment in patients with severe symptoms, rapid progression, major organ involvement and serious complications. Complete resection is recommended with negative margins to reduce the local recurrence. As in this case we could achieve complete resection and histopathological examination found the margins to be negative. The 3 - year local recurrence rate after surgery is ~40–50% [(8),] with long - term local recurrence rate of ~25–70% [(9),] although negative margins are associated with a much lower recurrence rate relative to positive margins (~10 vs.80%) [(10)]

Radiotherapy is recommended in patients with severe symptoms and contraindications to surgery like very old patient. Nuyttens et al. performed a retrospective study that revealed that radiotherapy or radiotherapy after surgery

achieved a local control rate of up to 70%, which was significantly higher than the rate for surgery alone, regardless of margin and recurrence. [11]

4. Conclusion

Intraabdominal AF are rare finding with variable presentation from being asymptomatic to severe symptoms as a result of organ involvement. Genetic association with FAP should be ruled out. Most of the cases can be managed surgically achieving negative margins to reduce the rate of local recurrence. In recent years systemic modalities of treatment has been recommended looking at the variable biological behaviour of the tumor.

Author's Contribution
SG and DK wrote the draft of manuscript. SG and JM were the operating surgeons. All authors contributed the literature review and approved the final manuscript for submission.

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Patient's Consent: Obtained

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Abbreviations
AF, aggressive fibromatosis; DTs, desmoid tumors;

References

[1] Jianchun Xiao, J. M. (2020, january 31). *frontiers in medicine*. Retrieved may 29, 2023, from frontiers in medicine: https: //www.frontiersin. org
[2] Schlemmer M. Desmoid tumors and deep fibromatoses. *Hematol Oncol Clin North Am.* (2005) 19: 565–71. doi: 10.1016/j. hoc.2005.03.008
[3] Bonvalot S, Ternes N, Fiore M, Bitsakou G, Colombo C, Honoré C, et al. Spontaneous regression of primary abdominal wall desmoid tumors: more common than previously thought. *Ann Surg Oncol.* (2013) 20: 4096–102. doi: 10.1245/s10434 - 013 - 3197 - x
[4] Braschi - Amirfarzan M, Keraliya AR, Krajewski KM, Tirumani SH, Shinagare AB, Hornick JL, et al. Role of imaging in management of desmoid - type fibromatosis: a primer for radiologists. *Radiographics.* (2016) 36: 767–82. doi: 10.1148/rg.2016150153
[5] Guglielmi G, Cifaratti A, Scalzo G, Magarelli N. Imaging of superficial and deep fibromatosis. *Radiol Med.* (2009) 114: 1292–307. doi: 10.1007/s11547 - 009 - 0458 - 7
[6] Lee JC, Thomas JM, Phillips S, Fisher C, Moskovic E. Aggressive fibromatosis: MRI features with pathologic correlation. *AJR Am J Roentgenol.* (2006) 186: 247–54. doi: 10.2214/AJR.04.1674
[7] Penel N, Le Cesne A, Bonvalot S, Giraud A, Bompas E, Rios M, et al. Surgical versus non - surgical approach in primary desmoid - type fibromatosis patients: a nationwide prospective cohort from the French Sarcoma Group. *Eur J Cancer.* (2017) 83: 125–31. doi: 10.1016/j. ejca.2017.06.017
[8] Crago, Aimeé M. MD, PhD*; Denton, Brian MS†; Salas, Sébastien MD, PhD‡; Dufresne, Armelle MD§; Mezhir, James J. MD*; Hameed, Meera MD¶; Gonen, Mithat PhD†; Singer, Samuel MD, FACS*; Brennan, Murray F. MD, FACS*. A Prognostic Nomogram for Prediction of Recurrence in Desmoid Fibromatosis. Annals of Surgery 258 (2): p 347 - 353, August 2013. | DOI: 10.1097/SLA.0b013e31828c8a30
[9] Cassidy, Michael R. MD∗, †; Lefkowitz, Robert A. MD‡; Long, Niamh MD‡; Qin, Li - Xuan PhD§; Kirane, Amanda MD∗; Sbaity, Eman MD∗; Hameed, Meera MD¶; Coit, Daniel G. MD, FACS∗, ||; Brennan, Murray F. MD, FACS∗, ||; Singer, Samuel MD, FACS∗, ||; Crago, Aimeé M. MD, PhD, FACS∗, ||. Association of MRI T2 Signal Intensity With Desmoid Tumor Progression During Active Observation: A Retrospective Cohort Study. Annals of Surgery 271 (4): p 748 - 755, April 2020. | DOI: 10.1097/SLA.0000000000003073
[10] Skapek SX, Ferguson WS, Granowetter L, Devidas M, Perez - Atayde AR, Dehner LP, Hoffer FA, Speights R, Gebhardt MC, Dahl GV, Grier HE; Pediatric Oncology Group. Vinblastine and methotrexate for desmoid fibromatosis in children: results of a Pediatric Oncology Group Phase II Trial. J Clin Oncol.2007 Feb 10; 25 (5): 501 - 6. doi: 10.1200/JCO.2006.08.2966. PMID: 17290057.
[11] Nuyttens, J. J., Rust, P. F., Thomas Jr, C. R., & Turrisi III, A. T. (2000). Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: a comparative review of 22 articles. *Cancer*, *88* (7), 1517 - 1523.

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