ORIGINAL RESEARCH PAPER

Surgery

INTRA-ABDOMINAL DESMOID TUMOUR- A RARE CASE REPORT WITH REVIEW OF LITERATURE.

KEY WORDS: Desmoid Tumour-Intra abdominal-resection

Dr. Bhimanagouda Marigoudar Junior Resident of Surgery, Department of Surgery, Gauhati Medical College Hospital, Guwahati-781032.

Dr. Rajiv Paul Assistant Professor of Surgical Oncology, Department of Surgery, State Cancer Institute, Gauhati Medical College., Guwahati-781032.

Dr. K Bhuyan Professor of Surgery, Department of Surgery, Gauhati Medical College Hospital, Guwahati-781032. *Corresponding Author

Dr. Soham Som Senior Resident of Surgery, Department of Surgery, Gauhati Medical College Hospital, Guwahati-781032.

ABSTRACT

Introduction: Desmoid tumours (DTs) are benign, slowly growing deep seated monoclonal myo-fibroblastic neoplasms. It arises from musculoaponeurotic stromal elements. The incidence in the general population is 2-4 cases per million people per year. DTs are typically sporadic; and can occur anywhere in the body. A 30 year male patient presented with a giant intraabdominal DT is discussed here with review of available literature. **Discussion:** DTs are benign, deep-seated slowly growing monoclonal myo-fibroblastic neoplasms arise from musculoaponeurotic stromal elements. Diagnosis may be confirmed with biopsy and immunohistochemistry. Preoperative assessment with MRI, CT with angiogram are imperative for definitive surgical resection. There are roles of chemotherapy and radiation therapy in management of primary and recurrent lesions. The surgical resection remains the mainstay of its management. **Conclusions:** The management of DT is challenging. Surgical resection is the cornerstone of its management. Chemotherapy and radiation therapy in management of primary and recurrent lesions may be considered.

CASE REPORT:

A 30-year-old male presented with swelling of his abdomen for six months with vague abdominal pain. The swelling covering whole epigastrium, left hypochondrium and left lumber region was found to be firm and fixed to abdominal wall. The clinical diagnosis was a giant intra- abdominal desmoid tumour. CT abdomen showed a giant tumour occupying the majority of the abdominal wall with intra-cavity extension (Fig 1). MRI showed a lesion with low signal density. Subsequent CT angiogram showed the tumour drawing its blood supply from the splenic artery, SMA, IMA with multiple collateral formation. Surgery was performed for definitive diagnosis and improvement of the symptoms.

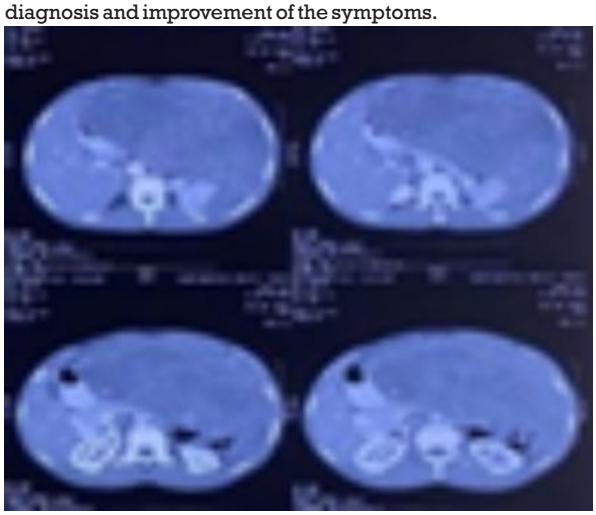

Fig1: CECT Whole Abdomen showing a giant tumour occupying the majority of the abdominal cavity.

A midline laparotomy incision, extending into left side found a large abdominal tumour involving left side of diaphragm, left lower 3 ribs, spleen, large bowel, omentum and mesentery.

A wide local excision was performed with resection of left side of diaphragm, left lower 3 ribs, left hemicolectomy, splenectomy and partial left lobe hepatectomy followed by reconstruction of left diaphragm with biological mesh (Fig 2,3). Abdomen closed in layers with intercostal drain in situ.

Fig 2: Intra-operative Photograph Of The tumour.

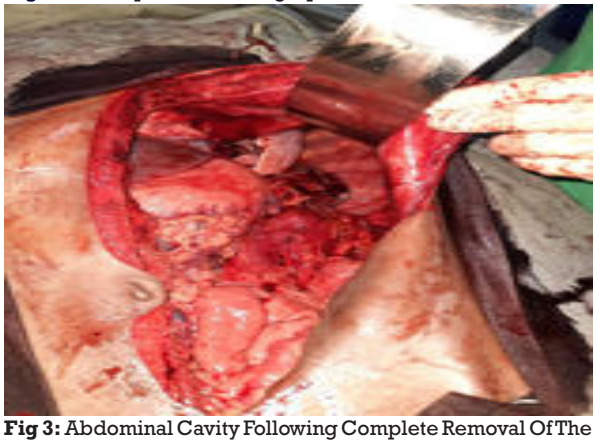

Fig 3: Abdominal Cavity Following Complete Removal Of The tumour.

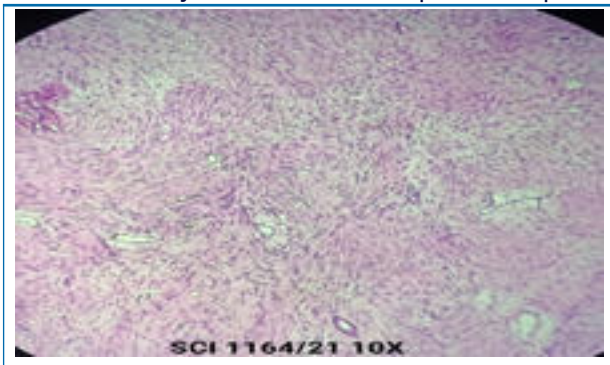

Fig 4: Desmoid Fibromatosis.

The immediate post operative course was uneventful. The post operative SSI was managed conservatively. The patient was discharged on 15th postoperative day. The HPE report (Fig 4) showed "Benign spindle cell tumour favouring desmoid fibromatosis".

DISCUSSION:

DTs are benign, deep -seated monoclonal myo-fibroblastic neoplasms that slowly grow, infiltrate, and arise from musculoaponeurotic stromal elements. DTs are rare; they account for only 0.03% of all neoplasms and less than 3% of all soft tissue tumours(1,2). They can arise from anywhere on the body. The most likely location for intra-abdominal DTs is the mesentery, especially the small bowel (3). Patients with FAP have a more than 800-fold risk of developing DTs compared to the general population. The index patient has no such family history and remained asymptomatic till it appeared as a lump in the abdomen.

DTs are usually appear as a well circumscribed homogenous lesion iso-dense or hyper-dense relative to muscle in CT and MRI. The optimal therapy in a symptomatic patient is complete surgical resection with negative margin (4). In spite of the complete resection of the tumour, recurrence rate of the DTs ranges between 30 and 40% [5]. Radiation and systemic therapy with tamoxifen, doxorubicin, NSAIDS and interferon are suitable in patients with recurrence[1,6].

A high level of clinical suspicion with appropriate imaging(CT, MRI) clinches the diagnosis. Surgery is the mainstay of its treatment modalities. There is high rate of recurrence in DT for which close follow up is essential.

CONCLUSION:

A rare case of giant intra- abdominal DT in a young male who had no history of trauma, surgery or FAP is described. The patients presented with a symptom of extra-abdominal lump. Imaging studies revealed a giant DT with intrabdominal extension. Extensive surgical excision was performed. The patient is being on followed up closely as DTs have a high rate of recurrence.

Conflict Of Interest: None

Consent: written consent obtained from the patient.

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REFERENCES:

[1] C.Escobar, R.Munker, J.O.Thomas,and G. V. Burton, "Update on desmoid tumour." Annals of Oncology, vol.23, no.3, pp.562-569,2012
[2] A.P. Burke, L.H. Sobin, K.M. Shekitka, et al., Intra-abdominal fibromatosis. A pathologic analysis of 130 tumours with comparison of clinical subgroups, Am. J. Surg. Pathol. 14 (April (4)) (1990) 335–341, PubMed PMID. 2321698
3] A.J. Smith, J.J. Lewis, N.B. Merchant, et al., Surgical management of intra-abdominal desmoid tumours, Br. J. Surg. 87 (May (5)) (2000) 608–613, PubMed PMID. 10792318
4] B. Kasper, P. Strobel, P. Hohenberger, Desmoid tumours: clinical features and treatment options for advanced disease, Oncologist 16 (5) (2011) 682–693, Pubmed Central PubMed PMID. 21478276.
5] M.T. Ballo, G.K. Zagars, A. Pollack, et al., Desmoid tumour: prognostic factors and outcome after surgery, radiation therapy, or combined surgery and radiation therapy, J. Clin. Oncol. 17 (January (1)) (1999) 158–167, PubMed PMID. 10458229.
6] S.R. Patel, R.S. Benjamin, Desmoid tumours respond to chemotherapy: defying the dogma in oncology, J. Clin. Oncol. 24 (January (1)) (2006) 11–12, PubMed PMID.;1; 16330666.