Case Report

Indian Journal of Pathology: Research and Practice
Volume 5 Number 1, January - April 2016
DOI : https://dx.doi.org/10.21088/ijprp.2278.148X.5116.8

Abdominal Musculoaponeurotic Fibromatosis

Banushree C. Srinivasamurthy*, Nagarajan K., Kaushik Saha***, Arpita Saha*****

*Assistant Professor *** Senior Resident, Department of Pathology **Assistant Professor, Department of Radiodiagnosis, All India Institute of Medical Sciences, Bhubaneswar.

Abstract

Desmoid tumor (musculoaponeurotic fibromatosis) is an aggressive monoclonal fibroblastic proliferation arising in musculo-aponeurotic structure at any site. It is a distinct pathological entity which can locally infiltrate and recur posing therapeutic challenges to a surgeon. We are reporting a rare case of abdominal desmoid tumor in a young patient.

Keywords: Desmoid; Abdomen; Fibroblast.

Introduction

The term desmoid is derived from the Greek word desmos, meaning a band or tendon[1]. McFarlane, was the first to report the disease occurring in the abdominal wall of a young woman after delivery in 1832[2]. Later, Mueller in 1838 coined the term desmoid tumor[3]. The fibromatoses are a group of benign fibroblastic proliferations that vary from benign to intermediate in biological behavior. The World Health Organization (WHO) now groups desmoids tumor under the term deep fibromatoses [4]. The deep fibromatoses are fibroblastic proliferations that arise within the deep musculo-aponeurotic structures and are traditionally divided into extraabdominal, abdominal wall, and intraabdominal types. They demonstrate infiltrative growth and local recurrence but do not metastasize. It is to have considered intermediate biological behavior due to frequent local recurrence[5]. Desmoid tumor is a rare lesion representing < 3% of all soft tissue tumors[6]. More frequently seen in the female population , with a female to male ratio ranging from 1.4 to 1.8. The peak incidence is between the ages of 25 and 35 years suggesting an association of this disease with the endogenous hormonal environment and exogenous sex hormones [7,8].

Corresponding Author: Banushree C.S., Assistant Professor, Department of Pathology, All India Institute of Medical Sciences, Sijua,Patrapada, Bhubaneshwar, Odisha 751019.

E-mail: drbanushree15@hotmail.com

Case Report

A 23-year-old female patient presented to surgery department with a painless gradually increasing mass in the lower abdomen for two years . The patient had no history of similar condition in family, previous abdominal surgery or any abdominal trauma.

On examination, a single lower abdominal mass involving right lumbar, hypogastric, and iliac regions and extending vertically 10cm below xiphisternum to pubic symphysis and transversely from right anterior axillary line to mid-line.

Lump was fixed to the anterior abdominal wall, nontender, globular in shape of size 10 × 8 × 8 cm with smooth surface, and firm in consistency. Ultrasonography demonstrated a large solid heterogeneous hypoechoic mass showing internal vascularity seen in pelvic cavity extending to right lumbar region displacing the bowel loops superiorly.

After preoperative workup, patient was planned for operation, and complete excision of the tumor with wide surgical margins along with the anterior abdominal wall down to the peritoneum was performed, resulting in a large wall defect which was repaired after mobilization and release of rectus

abdominis and reconstructed with polypropylene + Monocryl mesh . Skin was closed after keeping vacuum suction drain beneath the subcutaneous space.

The postoperative course was uneventful. On Histology, fascicles of spindle cells were seen arranged in herring bone pattern with uniform nuclei and Proliferative spindle cells entrapping the skeletal muscle. The periphery of the lesion was showing haemorrhage and perivascular lymphocytic infiltrate. Histopathological reports were consistent with desmoid tumor with negative surgical margins. At 1 year of followup, patient did not have any recurrence or incisional hernia.

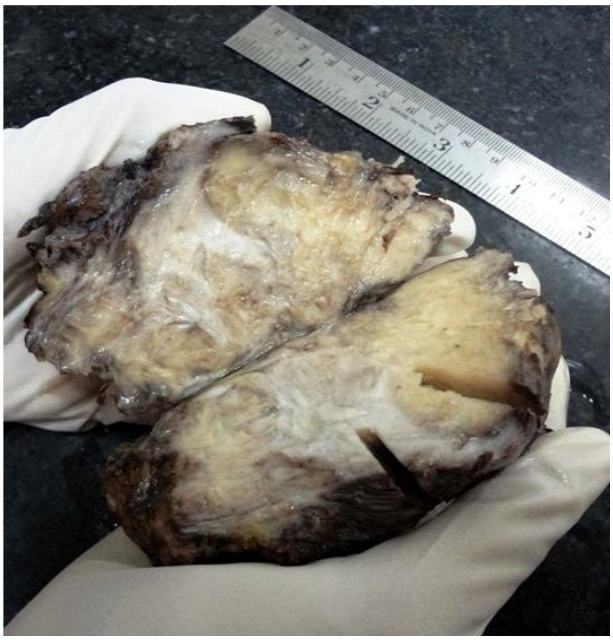

Fig. 1: Cut section showing glistening white and trabeculated tissue, resembling scar tissue

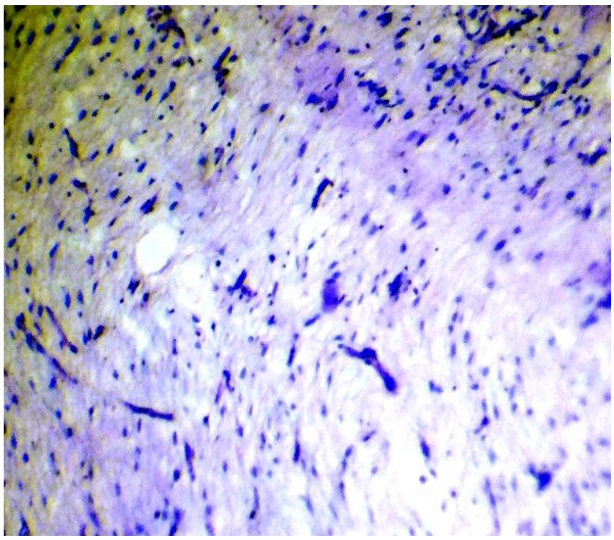

Fig. 2: Microscopic view of the excised tumor showing fascicles of fibroblastic spindle cells with abundant intercellular collagen

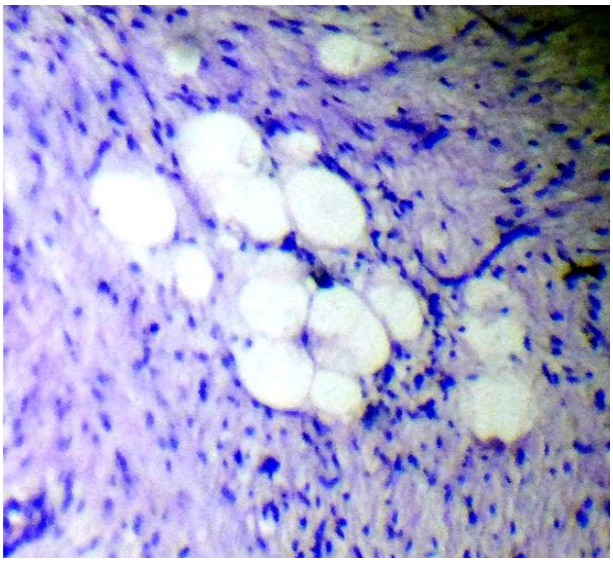

Fig. 3: Microscopic view of the tumor showing fascicles of spindle cells entrapping the muscle bundle and adjacent adipose tissue

Discussion

Desmoid tumors have been well characterized histologically , but their nature and pathogenesis have remained obscure for many years [9]. The incidence of desmoid tumors in the general population is 2–4 cases per million per year, with a slight female preponderance and peak incidence in the third and fourth decades . These tumors are more aggressive in younger patients, with recurrence rates up to 87% [10]. Numerous factors strongly associated with development of desmoid. In that, surgical trauma can be associated in approximately 25% of the cases[11]. Trauma, pregnancy and oral contraceptive pills are other risk factors [12]. However, none of the above mentioned risk factors were noted In present case. Abdominal desmoid tumor usually presents as a firm mass with ill-defined margins . Giant desmoid tumor arising from anterior abdominal wall weighing upto 6.5kg has been reported in the literature. Wide local excision with reconstruction of the defect is the treatment of choice. Incomplete tumor removal or positive surgical margins may lead to local recurrence. In our present case after complete removal surgical margins were negative and on one year follow-up, patient was asymptomatic [13]. We conclude that Desmoid tumors should be considered in a patient with painless anterior abdominal mass as it is a rare challenging clinical condition which requires a multidisciplinary approach due to its aggressive behavior and tendency to recur.

References

1. M. Sundaram, H. Duffrin, M. H. McGuire, and W. Vas. "Synchronous multicentric desmoid tumors (aggressive fibromatosis) of the extremities." Skeletal Radiology. 1998; 17: pp. 16–19.
2. MacFarlane J. Clinical report on the surgical practice of Glasgow Royal Infirmary. Glasgow: D. Robertson. 1832: 63.
3. Mueller J. Ueber den feinrn Bau und die Formen der Krankhaften geschwulste. Berlin: G Reimer and Erste Lieferung. 1838: 60.
4. C. D. M. Fletcher, K. Krishnan Unni, and F. Mertens, Eds., Pathology and Genetics of Tumours of Soft Tissue and Bone (IARC WHO Classification of Tumours), 3rd edition, 2006.
5. M. D. Murphey, C. M. Ruble, S. M. Tyszko, A. M. Zbojniewicz, B. K. Potter, and M. Miettinen, "From the archives of the AFIP: musculoskeletal fibromatoses: radiologic-pathologic correlation. "Radiographics. 2009; 29: pp. 2143–2176.
6. Reitamo JJ, Scheinin TM, Häyry P. The desmoid syndrome. New aspects in the cause, pathogenesis and treatment of the desmoid tumor. Am J Surg. 1986; 151: 230–237.
7. Posner MC, Shiu MH, Newsome JL, Hajdu SI, Gaynor JJ, Brennan MF. The desmoid tumor. Not a benign disease. Arch Surg. 1989; 124: 191–196.
8. Lopez R, Kemalyan N, Moseley HS, Dennis D, Vetto RM. Problems in diagnosis and management of desmoid tumors. Am J Surg. 1990; 159: 450–453.
9. Stout AP. Juvenile fibromatoses. Cancer. 1954; 7: 953–978.
10. Atul BS, Nikhil HR, Jyothi PJ, Katherine MK, Angela AG, James EB, et al. Ato Z of desmoids tumors. American Journal of Roentgenology. 2011; 19: 1008-1014.
11. McKinnon JG, Neifeld JP, Kay S, Parker GA, Foster WC, Lawrence W Jr. Management of desmoid tumors. Surg Gynecol Obstet. 1989; 169: 104–106.
12. Dinauer PA, Brixey CJ, Moncur JT, Fanburg-Smith JC, Murphey MD. Pathologic and MR imaging features of benign fibrous soft-tissue tumors in adults. Radio Graphics. 2007; 27: 173–187.
13. Mahim Koshariya, Samir Shukla, Zuber Khan, et al., "Giant Desmoid Tumor of the Anterior Abdominal Wall in a Young Female: A Case Report," Case Reports in Surgery. 2013; vol. 2013, Article ID 780862, 4 pages.