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RESEARCH ARTICLE

DESMOID FIBROMATOSIS OF THE ABDOMINAL WALL: SURGICAL RESECTION AND RECONSTRUCTION WITH MESH

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Abstract

Introduction: Desmoid tumor also known as aggressive fibromatosis are rare monoclonal fibroblast proliferation that are not regarded as malignant due to inability to metastasize but locally aggressive in their ability to locally invade the adjacent structures.

Case Report: A 29-year-old woman presenting with left iliac fossa region swelling, initially small in size then progress on and reached up to present size around 6×5cm in size. Wide local excision of tumor and meshplasty was done. In histopathology and IHC it was diagnosed as desmoidfibromatosis.

Conclusion: The clinical management of desmoid tumors is becomes complex due to high recurrence rates and requires a multidisciplinary approach because of the unpredictable disease course.

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Introduction:-

Desmoidtumors also called aggressive fibromatosis, are very rare with an estimated incidence of 2–5 cases per million [1, 2]. Desmoidtumors lack the capacity to metastasize but may behave in a locally aggressive fashion and possess a high risk of local recurrence despite adequate surgical resection with negative margins [1, 3]. Desmoidtumors can develop in any musculoaponeurotic structure and they may be located at virtually any anatomical site. They are divided into three general anatomic locations: extra-abdominal, intra-abdominal and abdominal wall. The principal sites of involvement for extra-abdominal desmoid tumors are the shoulder, chest wall and back, thigh and head and neck region. Intra-abdominaldesmoid tumors often arise in the mesentery or pelvis while abdominal tumors arise from musculoaponeurotic structures of the abdominal wall, especially the rectus and internal oblique muscles and their fascial coverings [4, 5]. With most favorable prognosis is for the lesions in abdominal wall and trunk. Wide local excision with negative margins is standard treatment for desmoid tumors. In the surgically treated patients who experience recurrence (local recurrence rates are about 25–65%) a wide re-excision is needed [6]. Synthetic mesh is often used in extensive abdominal wall reconstruction. It may present with complications, including infection, bowel adhesion, extrusion, and fistula according to the prosthetic material used [7]. A prosthetic material with more favorable properties than traditional mesh could have a major effect on surgical practice and patient outcomes, avoiding chronic inflammation and resisting infection after implantation [8].

Case Presentation:

A 29-year-old woman presenting with left iliac fossa region mass of size around 6×5cm in size. She had a history of caesarian section two years back following which after six months she noticed mass at the left lateral end of scar

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which was small and has progressed to present size in last two years. Normal bowel and bladder habit, no addiction history, normal menstrual history. On examination solitary swelling around 6×5cm, hard, non-tender with restricted mobility, fixed to underlying abdominal wall muscle on leg raising test. All routine blood investigation was normal, ultrasonography shows a homogeneously hypoechoic mass with vascularity on color Doppler of size approximately 5.5×5cm. On CECT abdomen a well-circumscribed mass, focally hyper attenuating size 5.5×5cm, not involved underlying muscle, it was suggestive of fibroma. Wide local excision of tumor was done [FIGURE 1] with onlay mesh plasty [FIGURE 2] and a negative suction drain was placed over it. Post-operative recovery was uneventful and discharged after third postoperative day after drain removal. On histopathology examination specimen was 8 x5x 4cm, base formed by muscle, on cut section well circumscribed grey white to white glistening nodular growth measuring 3.5x 3x3 cm seen with margins free from tumor. On microscopy sections showed an infiltrative tumor arranged in broad fascicles comprising of spindle shaped cells with eosinophilic cytoplasm, plump to wavy nuclei and indistinct cytoplasmic borders embedded in fibrovascular and abundant fibro-collagenous stroma, occasional pericytoma like blood vessels are also seen. Muscle cells bundle and lymphoid aggregates were seen. Cellular atypia or prominent mitotic activity was not seen. Features were suggestive with fibromatosis (Desmoid-tumour). On immunohistochemistry tumor was SMA: Positive, CD-34: positive, Vimentin: positive, Beta catenin: positive, Desmin: negative, Ki67: less than 01%, S-100: negative, CD34: negative and it was diagnosed as desmoid fibromatosis. Patient is on follow up with no complaints at six months.

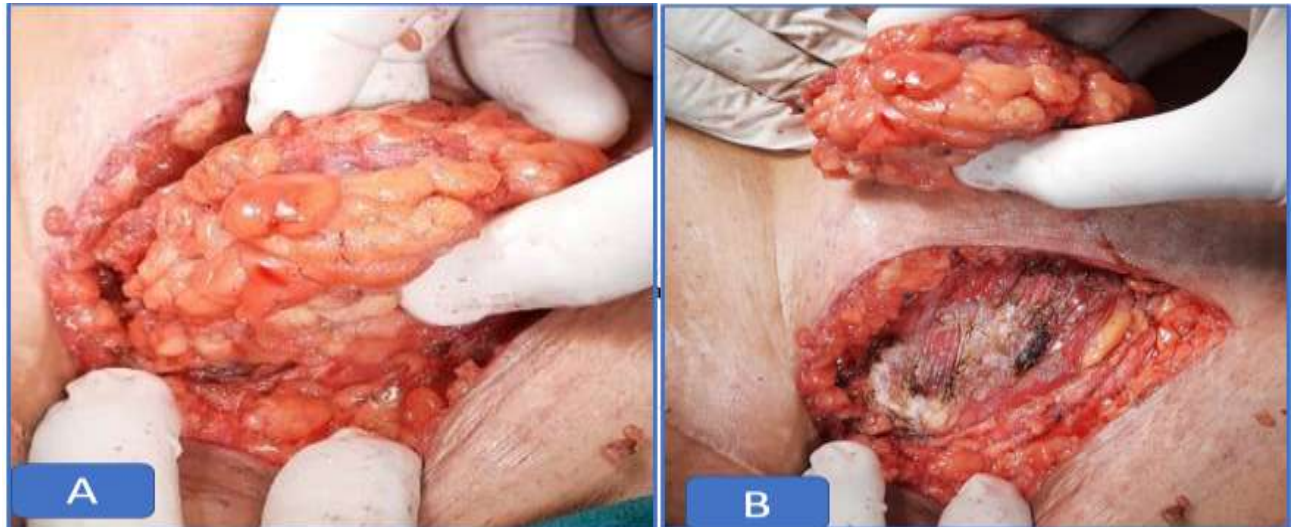


Figure 1:- A- Intraoperative image showing tumour mass, B- Wide local excision of tumor.



Figure 2:- Mesh placement of tumour bed.

Discussion:-

Desmoid tumor are neoplasms with infiltrating growth and with a tendency toward local recurrences; nevertheless, they lack metastatic potential. Although the morphologies of these tumors have been well characterized, their nature and pathogenesis have remained obscure for many years [6, 9]. According to the literature, the median age at the diagnosis of desmoid tumor is about 35 years and the majority of patients are women [10]. In particular, in these patients the abdominal wall is the preferred site of involvement [4]. Supposed risk factors of desmoids are previous surgical interventions, pregnancy, and hormonal treatment with estrogens [6]. Because the tumor biology is notoriously unpredictable, periods of rapid tumor growth can be followed by stability or even regression [11]. The treatment with tamoxifen, as well as chemotherapy and radiation, is controversial, since the long-term clinical improvement is minimal [6]. Wide local excision with negative margins is standard treatment and is crucial for reducing the recurrence rate. Abdominal wall integrity after full-thickness surgery can be restored with direct suture but the occurrence of postoperative incisional hernia is highly reported. For this reason one stage reconstruction with prosthetic abdominal wall reinforcement increases the chance of definitive cure, enhancing the patient's perceived quality of treatment [12]. Indeed, Luijendijk et al. [13] in a multicenter, randomized controlled trial observed a double rate of hernia recurrence in the primary suture group compared with the mesh reinforcement group. Synthetic meshes are usually associated with an increased risk of extrusion, adhesion, and following obstruction and enterocutaneous fistula formation, especially when placed in an overlay fashion [12]. Moreover, patients who have had radiation to the abdominal wall prior to reconstruction are at increased risk for wound healing complications and subsequent mesh exposure. For that reason, Butler et al. [14] recommend avoiding synthetic meshes in patients with radiated abdominal walls. The advent of biological matrices has added a valuable option to the field of abdominal wall reconstruction. The inherent ability of biological matrix to turn into patient self-tissue and therefore resist infection, allows to implant it in direct contact with the bowel, resulting in fewer adhesions than prosthetic mesh [8, 12]. These bioprosthetic devices, derived from human or animal dermis, are chemically and enzymatically cleaned to remove all cellular components while maintaining the extracellular matrix, which can be cross-linked or not. It is hypothesized that cross-linking treatment adds strength to the matrix, theoretically resulting in lower rates of hernia recurrence as compared to non-cross-linked products. In contrast Butler et al. [14] in a comparative study had not appreciated any mechanical differences between cross-linked and non cross-linked matrices. Moreover cross-linked matrices revealed delayed revascularization and higher percentage of adhesions resulting in poor integration regarding noncross-linked acellular dermal matrices. Despite the great advantages reported about the biological matrices, the high price ranging from USD 8.60/cm² to USD 22.00/cm², remains a high deterrent to their use [15]. So for a resource constraint country like ours conventional proline mesh is still most widely used and onlay repair with short duration suction drain helps to minimize the complications. Systemic therapies for desmoid tumor have been tried traditionally in unresectable, recurrent or a progressive disease. Anti oestrogen agents and NSAIDs are the first line of medical therapy used. Most of the desmoid tumors are strongly positive for estrogen receptors and these are the candidates for SERMs, commonly used ones are tamoxifen and reloxifen. Response rates being 40% to 51%.^[16] Sulindac is most commonly used NSAID with a response rate of about 28%.^[16] However an objective response may take several months to achieve stability and decrease the associated symptoms. Chemotherapy is considered second line of treatment owing to more side effects but it can be first line treatment in cases with rapidly progressive disease, impending destruction of critical anatomic structures or symptomatic unresectable tumors. commonly used regimen is are anthracycline in combination with methotrexate, vinblastine or cisplatin. With these French sarcoma group got a partial response in 54% cases.^[16] Tyrosine kinase inhibitors like imatinib and sunitinib have been tried with limited response after a prolonged treatment. Radiotherapy another modality used as primary treatment in unresectable tumors. 70% - 80 % chance of local control has been reported in retrospective European studies.^[16]

Conclusion:-

The abdominal wall desmoidfibromatosis should be considered in a young female patient presenting with abdominal wall mass, especially during pregnancy or within the 1st year of childbirth with the previous history of abdominal trauma or surgery (cesarean section) scar. The surgery has been the main stay of treatment but hormone therapy has resulted in the tumor regression, only in estrogen-receptor positive tumors, which could avoid mutilating surgery with multiple recurrences. Adjuvant radiotherapy could also reduce the incidence of local recurrence after complete resection. Mesh reconstruction helps in maintaining the integrity of the abdominal wall following surgery in large tumor.

References:-

1. Penel N, Coindre JM, Bonvalot S, Italiano A, Neuville A, Le Cesne A, Terrier P, Ray-Coquard I, Ranchere-Vince D, Robin YM, Isambert N, Ferron G, Duffaud F, Bertucci F, Rios M, Stoeckle E, Le Pechoux C, Guillemet C, Courreges JB, Blay JY: Management of desmoidtumours: a nationwide survey of labelled reference centre networks in France. *Eur J Cancer* 2016;58:90–96.
2. Rastrelli M, Tropea S, Basso U, Roma A, Maruzzo M, Rossi CR: Soft tissue limb and trunk sarcomas: diagnosis, treatment and follow-up. *Anticancer Res* 2014;34:5251–5262.
3. Colombo C, Miceli R, Lazar AJ, Perrone F, Pollock RE, Le Cesne A, Hartgrink HH, Cleton-Jansen AM, Domont J, Bovée JV, Bonvalot S, Lev D, Gronchi A: CTNNB1 45F mutation is a molecular prognosticator of increased postoperative primary desmoid tumor recurrence: an independent, multicenter validation study. *Cancer* 2013;119:3696–3702.
4. Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F (eds): WHO Classification of Tumours of Soft Tissue and Bone. Pathology and Genetics of Tumours of Soft Tissue and Bone, ed 4. Lyon, IARC Press, 2013.
5. Kasper B, Baumgarten C, Bonvalot S, Haas R, Haller F, Hohenberger P, Moreau G, van der Graaf WT, Gronchi A; Desmoid Working Group: Management of sporadic desmoid-type fibromatosis: a European consensus approach based on patients' and professionals' expertise – a sarcoma patientsEuroNet and European Organisation for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group initiative. *Eur J Cancer* 2015;51:127–136.
6. Bonvalot S, Desai A, Coppola S, Le Pécoux C, Terrier P, Dômont J, Le Cesne A: The treatment of desmoid tumors: a stepwise clinical approach. *Ann Oncol* 2012;23(suppl 10):x158–x166.
7. Brown GL, Richardson JD, Malangoni MA, Tobin GR, Ackerman D, Polk HC Jr: Comparison of prosthetic materials for abdominal wall reconstruction in the presence of contamination and infection. *Ann Surg* 1985;201:705–711.
8. Baumann DP, Butler CE: Bioprosthetic mesh in abdominal wall reconstruction. *SeminPlastSurg* 2012;26:18–24.
9. Meneghello C, Ousghir B, Rastrelli M, Anesi L, Sommariva A, Montesco MC, Rossi CR, Hladnik U, Segat D: Nuclear GSK-3 β segregation in desmoid-type fibromatosis. *Histopathology* 2013;62:1098–1108.
10. Van Broekhoven DL, Grünhagen DJ, den Bakker MA, van Dalen T, Verhoef C: Time trends in the incidence and treatment of extra-abdominal and abdominal aggressive fibromatosis: a population-based study. *Ann SurgOncol* 2015;22:2817e23.
11. Enzo MV, Rastrelli M, Rossi CR, Hladnik U, Segat D: The Wnt/ β -catenin pathway in human fibrotic-like diseases and its eligibility as a therapeutic target. *Mol Cell Ther* 2015;3:1.
12. Khansa I, Janis JE: Modern reconstructive techniques for abdominal wall defects after oncologic resection. *J SurgOncol* 2015;111:587–598.
13. Luijendijk RW, Hop WC, van den Tol MP, de Lange DC, Braaksma MM, IJzermans JN, Boelhouwer RU, de Vries BC, Salu MK, Wereldsma JC, Bruijninx CM, Jeekel J: A comparison of suture repair with mesh repair for incisional hernia. *N Engl J Med* 2000;343:392–398.
14. Butler CE, Burns NK, Campbell KT, Mathur AB, Jaffari MV, Rios CN: Comparison of cross-linked and non-cross-linked porcine acellular dermal matrices for ventral hernia repair. *J Am CollSurg* 2010;211:368– 376.
15. Shankaran V, Weber DJ, Reed RL 2nd, Luchette FA: A review of available prosthetics for ventral hernia repair. *Ann Surg* 2011;253:16–26.
16. Grignol VP, Polck R, Howard JH. management of desmoid. *surgclin N am*, 2016;96(5):1015-1030.