Giant Abdominal Wall Desmoid Tumor: Surgical Resection and Combined Reconstruction with Biological and Synthetic Prostheses

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Abstract

Introduction: Desmoid fibromatosis is an extremely rare but locally aggressive soft tissue tumor. When radical resection is indicated, the surgical team faces the challenge of abdominal wall reconstruction, for which optimal technique is still debated. We present the case of a giant abdominal wall desmoid tumor identified in the ante-partum period, showing a growth in the post-partum period which was subsequently resected.

Presentation of the case: 37 years old woman suffered from rapidly grew desmoid tumor of abdominal wall explored by CT scan and a biopsy which revealed a large abdominal wall tumor invading completely the right part of abdominal rectus and oblique muscles without developing in the abdominal peritoneal cavity. The 30cm×25cm mass was successfully resected with negative margins. The formed defect was repaired following a combined method using a synthetic bi face mesh Ventra light ST and an absorbable biological mesh Phasix®. The patient had an uncomplicated post-operative course and was discharged on day 14.

Discussion: There is no consensus regarding the surgical technique to close the abdominal defects. Nowadays, in the most cases, surgeons use different types of synthetic meshes. The advent of biological matrices has added a valuable option to the field of abdominal wall reconstruction.

Conclusion: We present a case of large abdominal wall desmoid tumor excision followed by reconstruction with biological implant.

ABBREVIATIONS

FAP: Familial Adenomatous Polyposis; APC: Adenomatous Polyposis Coli

INTRODUCTION

Desmoid tumor is a benign and slow-growing soft tissue tumor of intermediate grade with a potential for local recurrence and invasiveness without metastatic spreading[1-3]. Although first described by MacFarlane in 1832 [4], the designation desmoid was given in 1838 by Muller in Berlin, who first used the term “desmos” relating to the Greek word – which means “similar to tendon” [5]. Such neoplasia is extremely rare, it accounts for 0.03% of all documented neoplasms and approximately 3% of all soft tissue tumors, with an incidence of 2–4 per million per year [6].

Although the etiology of these tumors is unknown, certain factors play a role in their development and growth, such as Familial Adenomatous Polyposis (FAP), including a mutation in the APC gene [7], endocrine and physiologic factors, including estrogen hormonal stimulus and pregnancy [8-10]. There is no generally accepted protocol for treatment. Nevertheless extremely thorough surgical resection is essential. Radiotherapy can be beneficial in case of residual or inoperable tumors.

This is the report about the combined use of absorbable biological implant and synthetic mesh in case of abdominal wall reconstruction after extensive giant desmoid tumor successfully resection.

CASE PRESENTATION

In 2015, during pregnancy delivery by a cesarean, a 37 years old woman has been diagnosed with an abdominal wall neoformation: a solid mass up to 5 cm in diameter. After diagnosis this patient did not seek any medical examination and treatment. In her history she reported 4 pregnancies with cesarean births. The patient had no prior medical history, and no family history of a similar condition, colorectal cancer, or familial adenomatous polyposis.

In January 2017 the patient was taken to the Poitiers
University Hospital due to increasing of abdominal growth. Clinical examination of the anterior abdominal wall revealed tumor of huge dimensions; medial border was the white line, lateral border was the anterior-superior iliac spine, upper border was a margin of the 12th rib, and lower border was the pubic bone. She did not experience any change in bowel habits. A computed tomography was performed; solid tumor sized 211mm × 237mm on the right abdominal wall, noninvasive in abdominal peritoneal cavity (Figure 1). Result of percutaneous biopsy determined a desmoid tumor.

The patient was taken to the operating room for surgical exploration and resection. Considering a large tumor and a wide resection, the operation was performed together with specialists of plastic and reconstructive surgery.

After paramedian laparotomy focused on the lesion (Figure 2), exploration confirmed the tumor local extension to the whole right abdominal rectus muscle and partially to oblique muscles with a measuring size of 30cm × 25cm. The rectus muscle and a large portion of the oblique and transverse muscles were removed en bloc with 1,5cm of healthy margin from the neo formation. Intraoperatively the absence of tumor invasion into the abdominal cavity was confirmed. The formed defect of the anterior abdominal wall sized 15x20cm, which was repaired combining method using different types of meshes and muscles aponeurosis.

To close peritoneum defect and to achieve better strength we implanted a synthetic bi face mesh Ventralight ST (Bard, USA) with intraperitoneal (IPOM) technique (Figure 3). Because of the large size of the tumor, elasticity of the lateral and left muscular-aponeurosis wall increased, subsequently it was possible to perform an aponeurosis closure above the synthetic mesh using relaxing incisions which granted successful approach of the wound edges (Figure 4). In order to reduce the risk of recurrence, we decided to implement an additional layer under the cutaneous plane. To avoid infectious complications in case of skin necrosis we choose an absorbable biological mesh Phasix® (Bard, USA) with onlay technique (Figure 5). At the end of the complex reconstruction a large flap of epidermis-free redundant skin was used for the last layer laid on the absorbable mesh (Figure 6). The anterior abdominal wall was drained by two suction drains placed at the contact of each prosthesis.

Postoperative course showed no complications, drainages...
were removed on day 7 and 8 respectively; the patient was discharged on the 14th day. There was no evidence of recurrence of the neither tumor nor incisional hernia at 4 months of follow-up.

Histological examination of the resected tumor sized 33cm ×22 cm, weight 8.1kg (Figure 7), was characterized by the proliferation of fibroblasts and myofibroblasts with tumor-free margins. Stroma was fibrous, moderately edematous, with limited necrobiotic sites and hemorrhagic effusions. The diagnosis was confirmed by the presence of vimentin as shown by nuclear β-catenin staining.

**DISCUSSION**

Desmoid-type fibromatosis, also known as aggressive fibromatosis, musculoaponeurotic fibromatosis or desmoid tumor is a rare soft tissue neoplasm characterized by infiltrative growth and a tendency toward local and late recurrence, but an inability to metastasize. According to the literature, the median age at diagnosis of desmoid tumor is about 35 years, and the majority of patients are women [11]. About 15% of desmoid tumor cases are related to FAP [12-13]. Up to 20% of patients with FAP will develop a desmoid tumor, most of which occur intra-abdominally or in the abdominal wall and arise at the site of prior surgical anastomoses or incisions. When located in the abdominal wall, desmoids tumors raised from the rectus abdominal muscle or from previous Cesarean section scars [14].

Intra-abdominal desmoids are usually asymptomatic. They become symptomatic when they compress or have infiltrated surrounding viscera. This may result in intestinal obstruction, ischemic bowel secondary to vascular compression, and hydropneumosis due to ureteric compression. Desmoid tumors can also rarely lead to bowel perforation as well as deep vein thrombosis, pyrexia of unknown origin, gastrointestinal bleeding and intra-abdominal abscess formation [15].

The surgical treatment of desmoid type fibromatosis tumors is still a challenge for surgeons. Although the low incidence, aggressive infiltration and growth towards healthy adjacent tissue implicates in high morbidity associated with radical resection. Currently adequate surgical resection with negative margins is the treatment of choice, except cases when radical surgery would be mutilating or associated with considerable function loss [16].

The literature review about desmoid type fibromatosis within the abdominal wall showed no consensus regarding the surgical technique to close the abdominal defects. Nowadays, in most cases, surgeons use different types of synthetic meshes [17]. However, synthetic meshes are usually associated with an increased risk of extrusion, adhesion, and subsequently obstruction or enterocutaneous fistula formation, especially when placed in an intraperitoneal fashion [18]. 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. The advent of biological matrices has added a valuable option to the field of abdominal wall reconstruction.

One variation of the biological materials, Phasix®, has not been described yet in the literature for abdominal wall reconstruction after tumor excision. Phasix® (Bard, USA) is a naturally derived, absorbable polymer made of poly-4-hydroxybutyrate (P4HB). The choice of a biological mesh was induced by the high risk of infection in onlay position. Our goal was to protect synthetic prosthesis and increase solidity of the abdominal wall even in case of mesh infection without removing it [19]. We report for the first time the successful use of Phasix® in a complex abdominal wall reconstruction following resection of a large desmoid tumor. This device was well accepted without any postoperative complications and no evidence of recurrence of the tumor or incisional hernia has been reported 4 months later.

**REFERENCES**


