

SUCCESSFUL SURGICAL REMOVAL OF A HUGE PELVIC DESMOID TUMOR

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Pelvic fibromatosis, also known as desmoid tumor, is a benign neoplasm with no metastatic potential [1]. It accounts for about 0.03% of all neoplasms [2]. Pathophysiologically, it is a deep, infiltrative fibroproliferative process involving the fascia and muscle aponeurosis. The mean age at the time of diagnosis is 41.3 years [3]. The most common sites are the anterior abdominal wall and shoulder girdle. Spread of aggressive fibromatosis can cause compression of the intestine, bladder, ureter, vagina, and pelvic nerves and vessels, leading to its symptomatology, morbidity and mortality. It is highly associated with female sex [3], surgery or trauma (4-fold risk), pregnancy, familial polyposis coli (10–15% of the cases) [4,5] and Gardner's syndrome. The reported recurrence rate after surgery is 45% [6].

A 40-year-old woman weighing 79 kg presented with a 2-month history of abdominal distension. The distension was sudden in onset and painless, but had been associated with indigestion and epigastric pain and urinary symptoms including frequency, urgency, and dull aching pain in the left lumbar region for 1 week. She was the mother of four children, the youngest of whom had been delivered by lower segment cesarean section 3 years before. Her menstrual cycles and bowel habits were normal, and there were no neurologic symptoms. Examination revealed a huge (38 × 36 cm) immobile, non-tender and solid mass, extending from the suprapubic area to the xiphisternum. There was no fluid thrill or shifting dullness. Speculum examination revealed a healthy-looking cervix and a hard, immobile mass filling all the fornices on bimanual palpation. The uterus was not felt separately. All routine blood investigations and tumor markers were within normal limits. Abdominopelvic ultrasonography revealed a large mass of mixed echogenicity that was mainly solid, with a few cystic lesions arising from the pelvis and extending to the epigastrium. The uterus and ovaries could

not be visualized because of the large mass that was clearly separate from the liver, gall bladder, pancreas, and spleen. The kidneys, ureters, and bladder were all normal. Computed tomography (CT) examination of the abdomen and pelvis confirmed a soft-tissue density mass of 26 × 13 × 24 cm completely filling the pelvis, and extending into the abdomen with mild to moderate hydroureter and hydronephrosis (Figures 1 and 2). There

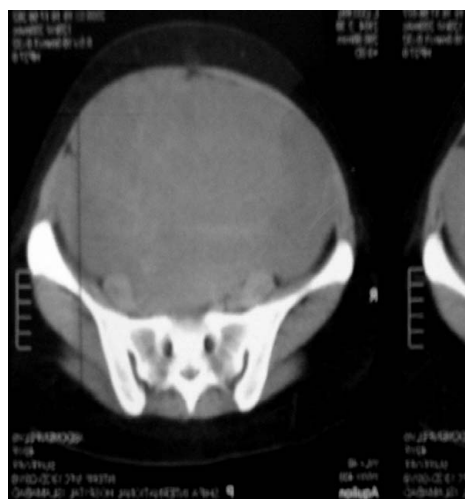


Figure 1. Computed tomography scan shows tumor (transverse view).



Figure 2. Computed tomography scan shows tumor (sagittal view).



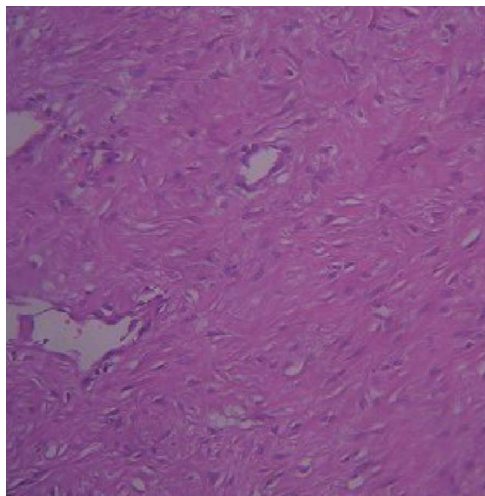
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Table. Outcome of various treatment modalities

Study	Type of study	Treatment options	Outcome
Huang et al [6]	Case report	Surgical removal	Recurrence after 2 yr
Sportiello & Hoogerland [12]	Case report	Surgery, surgery + radiotherapy, tamoxifen	Recurrence, recurrence, no recurrence after 27 mo
Nuytens et al [5]	Comparative review	Surgery, surgery + radiotherapy, radiotherapy	Cases treated with surgery + radiotherapy or radiotherapy alone had best outcome
Mariana et al [11]	Case series	Surgery alone	Recurrence in 3/6 cases
Bhama et al [14]	Case report	Surgery + doxorubicin & ifosfamide	Complete resolution

**Figure 3.** Myofibroblast proliferation with dense collagenous background. (Haematoxylin & eosin stain. Original magnification, x40)

was no evidence of pelvic lymphadenopathy or ascites. All visible soft tissues were normal. The uterus and ovaries were not visualized separately from the mass. A provisional diagnosis of an ovarian mass was made.

The patient and relatives received counseling with a view to performing exploratory laparotomy. Gut preparation and urologic consultation were performed followed by bilateral DJ stenting, prior to the procedure. Operative findings showed no ascites, but a huge, solid lobulated mass was detected attached to the anterior abdominal wall, omentum, and bladder, apparently arising from the fibrous tissue at the posterior surface of the pubic bone. The liver and under-surface of the diaphragm were free. The para-aortic lymph nodes were not enlarged and the uterus, Fallopian tubes, and ovaries were healthy and separate from the mass. A multidisciplinary team including a general surgeon and a urologist were involved in the complete resection of the mass, followed by repair of the bladder, which was opened during the procedure. The patient was transferred to the intensive care unit in view of the massive surgery, but she remained stable. Drains were removed

on the third postoperative day and the DJ stent and urethral catheter were removed on the fifth postoperative day. The patient was discharged on the seventh postoperative day, in a healthy condition. The tumor weighed 6.5 kg. Histopathologic examination confirmed the mass as intra-abdominal pelvic fibromatosis (Figure 3). The patient remained well with excellent wound healing at her 6-month follow-up. A repeat evaluation was planned after a further 6 months, in light of the high recurrence rate for this type of tumor.

Pelvic desmoid tumors are rare neoplasms with only 31 previously reported cases [7]. Intra-abdominal tumors remain asymptomatic until their growth and infiltration leads to intestinal, vascular, ureteric, and neuronal manifestations. Their etiology is uncertain, but may be related to trauma or hormonal factors, or may have some genetic associations (familial polyposis gene on chromosome 5). They commonly occur in young women during or after pregnancy, and are reported to regress after menopause [8]. Estrogen receptors have therefore been implicated in the pathogenesis of the disease.

The most common differential diagnoses include fibrosarcoma and Gardner's syndrome. The diagnostic workup involves immunostaining with vimentin, α -smooth muscle actin, muscle actin, and desmin. Mutational analysis of the gene for adenomatous polyposis coli is also recommended because of the high association between these diseases. Magnetic resonance imaging is considered to be superior to CT in determining the extent of the tumor and its relationship to nearby structures, and during follow-up to detect any recurrence after surgery. The final diagnosis is based on histopathologic examination.

Tumors are usually 3–7 cm in size [9], though the current patient presented with a short history of abdominal distention and a huge mass filling the whole abdomen. A CT scan showed a bilobed mass filling the whole abdomen and pelvis, but the exact site of origin remained unclear. Tumor markers are of no diagnostic use as previous reports have always found them to be negative, as in this case [10,11]. Wide surgical excision

was performed and the entire mass was removed successfully, in accordance with the standard management for primary tumors [12].

This type of tumor is associated with very high recurrence rate, even after complete surgical removal, and tamoxifen and other antiestrogens have been reported to be successful in such cases [11,13,14]. Although radiotherapy and chemotherapy using dacarbazine, carboplatin and doxorubicin can also be associated with good results [15–17], they are associated with their own side effects and are usually reserved for recurrent aggressive tumors requiring multiple surgeries. Tamoxifen should therefore be the first choice of treatment in recurrent cases, in view of its reduced side effects compared to chemotherapeutic agents. However, further clinical trials are needed to decide the optimal management strategy for these tumors because of their high recurrence rate.

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