Successful treatment of recurrent pelvic desmoid tumour with tamoxifen

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The case report of a young woman with recurrent pelvic desmoid tumour successfully treated with tamoxifen is described. The desmoid tumour recurred within 6 months after the initial exploratory laparotomy. Tamoxifen therapy led to complete relief of ascites within 2 months and complete tumour regression by the end of the fourth month, and the patient has remained stable for 6 years. Without sacrificing pelvic organs or major vessels and preserving reproductive ability, tamoxifen should be considered as the first drug of choice in such a recurrent condition.

Key words: desmoid tumour/tamoxifen

Introduction

Desmoid tumour or musculoaponeurotic fibromatosis is a rare neoplasm of spindle-shaped fibroblasts and collagen which infiltrates muscle and can become adherent to adjacent organs. Pelvic desmoid tumours are common in women of reproductive age (Adcock \textit{et al.}, 1989). The spread of aggressive fibromatosis can cause compression of the intestine, ureters, bladder, vagina, pelvic nerves and major vessels and fatal cases have been reported (Simon \textit{et al.}, 1985). Although surgery has been the primary treatment, complete local excision is difficult and is associated with high recurrence and complication rates (Adcock \textit{et al.}, 1989). To preserve the fertility of women of reproductive age, various alternative treatments have been used, including steroids and trimephenylethylenes (tamoxifen and toremifene) (Timmons \textit{et al.}, 1994). This report describes a case of recurrent pelvic fibromatosis that was successfully treated with tamoxifen; the patient has remained disease-free for over 6 years.

Case report

This 19 year old female high school graduate had suffered from distension of the abdomen for 3 months beginning in early January 1992. She was unmarried, and had no sexual experience or any significant medical or surgical history. Routine gynaecological ultrasonography revealed a cystic lesion with some solid components and irregular lining over the right adnexal region. Ascites was also present. She was admitted and underwent tests for possible gynaecological malignancy. There was no elevation in serum tumour markers including α-fetoprotein and CA-125. Computed tomography (CT) scan showed a lobulated mass with homogenous enhancement in the right lower abdomen. The mass involved the right adnexa, right lower ureter and extended to the pelvic wall (Figure 1). Intravenous pyelogram (IVP) showed marked right hydronephrosis with obstruction at the lower ureter. However, a retrograde pyelogram could not be performed because of inability to insert a 5 Fr. ureteral catheter beyond 5 cm from the right ureteral orifice. An exploratory laparotomy was performed and 500 ml of bloody ascitic fluid was drained. A pseudocystic lesion was found over the right paracolic gutter, and a dense fibrous plaque was found over the right paracolic gutter, which extended to the pelvic brim and down into the pelvis and involved the right parametrium and round ligament. No abnormalities were found in the uterus or bilateral tubes. The right ovary was of normal size but was densely adherent to the fibrous plaque. Attempts to perform lysis failed due to excessive intra-operative blood loss (about 1000 ml) and the dense nature of the adhesion. Retropertitoneal soft tissue biopsy was performed. Microscopically, soft tissue specimens revealed a picture of fibromatosis characterized by proliferation of uniform slender spindle-shape fibroblasts arranged in bands and fascicles and surrounded by varying amounts of collagen. No pleomorphism or mitosis was seen. The patient refused further treatment and was released on the seventh postoperative day.

Abdominal distension recurred 6 months later, and she was referred to our hospital due to poor oral intake for a month. Physical examination revealed cachexia with a body weight of 35 kg. Her abdomen was distended with a circumference of 85 cm. CT scan showed massive ascites and obstruction of the right ureter at the L3–L5 level. Fibrous adhesion bands were seen arising from the right pelvic wall which formed a cystic lesion adjacent to the right ovary. An antegrade pyelogram showed the leakage of contrast medium at the distal right ureter into a loculated pocket in the retroperitoneum. The ascitic fluid was serosanguineous in colour. Analysis revealed a pH of 7.9, osmolarity 285 mosmol/kg, specific gravity 1.030, blood urea nitrogen (BUN) 16 mg%, creatinine <10 mg, red
well for a follow-up period of more than 6 years after the cessation of tamoxifen therapy.

Discussion
Pelvic fibromatosis or desmoid tumour is a rare, non-metastatic tumour that tends to be locally invasive. It is always recurrent and arises from the fascial sheath and musculoaponeurotic structures (Adcock et al., 1989). These tumours form large mass lesions that are poorly circumscribed and often infiltrate the adjacent organs. Clinically, desmoids may be mistaken for ovarian, mesenteric or retroperitoneal tumours. However, histological examination readily distinguishes these tumours from sarcomas due to the lack of pleomorphism, cellularity and mitotic activity (Simon et al., 1985). Desmoids are a common manifestation of Gardner’s syndrome and have a predilection to develop in women of reproductive age. Until recently, surgery has been the mainstay of treatment for fibromatosis. However, total resection often proves difficult, as microscopic tumour is frequently present beyond the disease. Reconstructive procedures may be necessary following radical excision. Incomplete resection explains the high recurrence rate of desmoid tumour. In cases of recurrent tumours or widely involved tumours, radiation or chemotherapy may be attempted (Kinzbrunner et al., 1983; Timmons et al., 1994). Although these alternative options of therapy may be beneficial, they may also cause serious side-effects. Surgery was not considered as the first treatment option in the present case, because the clinical diagnosis and pathology had already been confirmed. In 1983, cases of desmoid tumours were reported which dramatically responded to tamoxifen (Kinzbrunner et al., 1983; Waddell et al., 1983). In the present case, tamoxifen therapy was preferred to high dosage of steroid or cytotoxic drugs because our patient was a young woman and the use of tamoxifen caused relatively fewer side-effects. Immediate extensive urological procedures were not arranged due to the risk of aggravation of the disease by trauma. Percutaneous nephrostomy was performed to resolve hydronephrosis. A second exploratory laparotomy was performed once the remission of disease was achieved and the right ureteral stricture caused by infiltration of desmoid was repaired smoothly.

The oestrogen receptor appears to be implicated in the pathogenesis of desmoid tumour, since fibromatosis is common in women of childbearing age and regresses following menopause. Reports have shown that trials of different agents that counteract the effects of oestrogen can produce partial or complete regression. Tamoxifen has anti-oestrogen actions and this was shown to be partially responsible for its effects on desmoid tumour (Mukherjee et al., 1995). However, tamoxifen has also been shown to produce a response in desmoid tumours without oestrogen receptors. In this case, decreasing fibroblast growth factor or inducing an increase in transforming growth factor β concentrations by tamoxifen presumably led to the inhibition of aberrant fibroblasts in desmoid tumour (Benson and Baum, 1993). This hypothesis has led to a debate on the inter-relationship between the biology of fetal and adult tissue development, wound healing and the progression of tumours. It is not known how long tumour remission persists after
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therapy nor how long tamoxifen therapy should last (Timmons et al., 1994; Shulman et al., 1995). In this case, tamoxifen was given for 7 months and the patient remained disease-free for more than 6 years with normal ovarian function.

Overall, this case demonstrates that a trial of tamoxifen therapy is indicated in case of recurrent pelvic fibromatosis rather than extensive surgical re-exploration or radiation therapy. In young women with recurrence, tamoxifen should be considered as the first choice of treatment as it may avoid the risk of massive haemorrhage and resection during surgery, and provide better chances for preservation of fertility.

Reference


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