Case Report

A Large Abdominal Desmoid Tumor Associated with Pregnancy

Vichit Viriyaroj MD*, Narongchai Yingsakmongkol MD*,
Passakorn Pasukdee MD*, Naparat Rermluk MD**

** Department of Surgery, Faculty of Medicine, Srinakarinwirot University, Bangkok, Thailand*
*** Department of Pathology, Faculty of Medicine, Srinakarinwirot University, Bangkok, Thailand*

Desmoid tumors are rare, benign tumors with locally aggressive behavior which originate from fascial or musculoaponeurotic structure. They are more common in young women than young man. The etiology of desmoid tumors are uncertain but may be related to operation, trauma or hormonal factors. The authors report a 17-year old woman, gravida 1, para 1 with a mass at her lower abdominal wall during the fifth month of gestation. She was biopsied during delivery in another hospital but was not given a definite diagnosis. The mass had grown rapidly after biopsy and delivery. The tumor measured 28 x 21 x 18 centimeters in size and 4,900 g in weight. Complete surgical excision was performed. The pathological report was desmoid tumor (aggressive fibromatosis). She had no post-operative complication and no recurrent tumor in the 8 months after operation.

Keywords: *Abdominal wall, Fibromatosis, abdominal, Fibromatosis, aggressive, Pregnancy complications, neoplastic*

J Med Assoc Thai 2009; 92 (Suppl 3): S72-5
Full text. e-Journal: *http://www.mat.or.th/journal*

Desmoid tumors are rare, benign tumor with locally aggressive behavior. Desmoid tumors proliferate from mesenchymal cells that arise from fascial or musculoaponeurotic structure. The incidence of sporadic desmoid tumor is estimated to be 2-4 per million individuals per year[1,2]. Desmoid tumors are classified according to their location into extra abdominal and abdominal type. The abdominal type can be sub-classified according to their location as in the abdominal wall or intra-abdomen. The most common sites affected are the shoulder girdles and rectal muscles[2,3]. Desmoid tumors are more common in young women than young men[2,3]. The causes of desmoid tumor are uncertain and may be related to operation, trauma or hormonal factors. This case report shows a rare occurrence of large abdominal desmoid tumor during pregnancy.

Correspondence to: Viriyaroj V, Department of Surgery, Faculty of Medicine, Srinakarinwirot University, Nakhon-Nayok 26120, Thailand. Phone: 037-395-085 ext. 11201-2, Fax: 037-395-271, E-mail: vivichit@yahoo.com

Case Report

A 17-year old woman, gravida 1, para 1 was presented with a mass at her suprapubic region during the fifth month of gestation. She completed a full-term pregnancy and delivered by cesarean section. She was biopsied during delivery in another hospital but given no definite diagnosis. The mass had grown rapidly after biopsy and delivery. She developed frequency and difficulty in micturition. She has no family history of these symptoms and signs. Physical examination found a lower abdominal wall mass measuring 20 x 20 centimeters in diameter, which was fixed to pubic bone, rubbery to hard in consistency, not tender. Magnetic resonance imaging (MRI) revealed a large lobulated mass in the pelvis. The tumor was minimal invasion into the lower part of the left rectus abdominis muscle and extended into the true pelvis and under the pubic symphysis. The urinary bladder, uterus and left ovary were displaced to the right side. There was moderate bilateral hydronephrosis from bilateral distal ureteric obstruction by this tumor. There was no evidence of intra-abdominal, pelvis, retroperitoneal

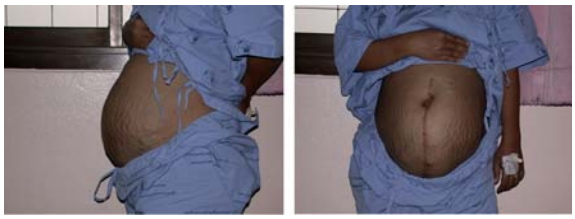

Fig. 1-2 The patient presented with a large lower abdominal wall mass which was fix to pubic bone, rubbery to hard in consistency

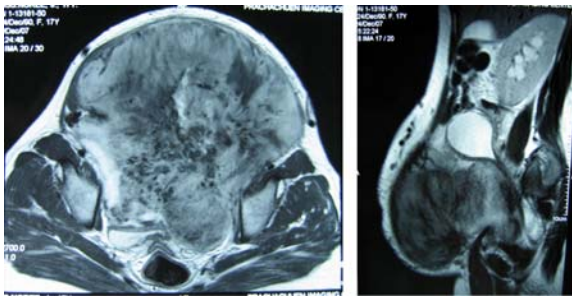

Fig. 3-4 MRI revealed large lobulated mass in the pelvis

Fig. 5-6 The gross pathology revealed multilobulated mass and cut surface revealed white whirling texture without necrosis myxomatous appearance

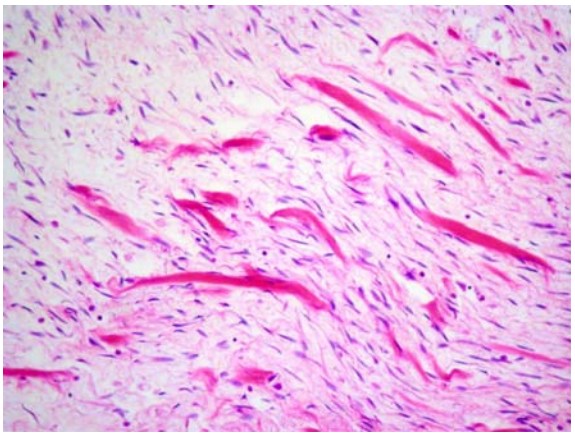

Fig. 7 The histological examination revealed cellular proliferation composed of ill-defined fascicles of band spindle cells and keloidal collagen deposition in the mass (H&E stain)

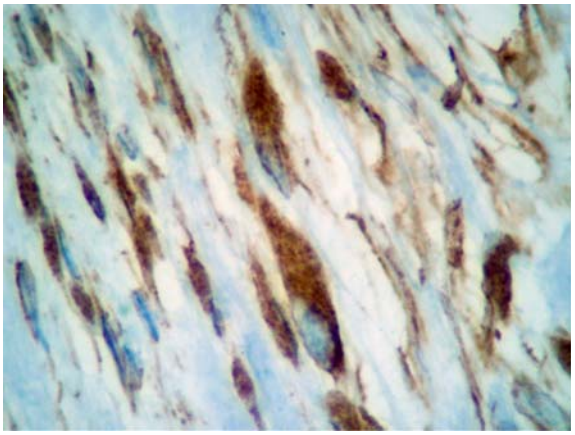

Fig. 8 The immunohistochemistry is positive for Vimentin

lymphadenopathy or peritoneal nodule. She underwent a cystoscopy with double-J stent placement in both kidneys before the operation. The operative finding found a large extra-peritoneal tumor mass at prevesical space. The tumor occuppied in the pelvis and infiltrated the rectus abdominis muscle and pelvic floor muscle. The tumor mass was removed completely. Her abdominal wall was closed by direct repair without using any prosthesis.

She underwent colonoscopy on the 7th post operative day which revealed no colonic polyp. She was discharged from the hospital without post operative complication. No evidence of recurrent tumor has been found in the 8 months after operation.

The pathological report revealed the resected specimen measuring 28 x 21 x 18 centimeters in size and 4,900 grams in weight. The histological examination revealed cellular proliferation composed of ill-defined fascicles of band spindle to stellate cells with dispersed small to medium sized vessels. The nuclei are fusiform to oval shape and one-pale stained. There was also keloidal collagen deposition in the mass. The appear-ance of which was consistent with desmoid tumor (aggressive fibromatosis). The immunohistochemistry is positive for Vimentin, focal positive for smooth muscle actin, negative for CD34, S100 and desmin.

Discussion

Desmoid tumors are uncommon, benign tumor. They account for 0.03% of all neoplasm and less than 3% of all soft tissue tumors[1,2] but the incidence

in familial adenomatous polyposis (FAP) patient ranges from 3.6-34%[4]. They arise from fascial or musculoaponeurotic structure. These tumors frequently infiltrate local tissue but they do not have the ability to metastasize. The majority of tumors is found in women ages 20-40 years[2,3]. In sporadic cases, the most common sites affected are abdominal wall and shoulder girdle[2,3]. In contrast to sporadic cases, familial adenomatous polyposis (FAP) associated desmoids (Gardner's syndrome) are mainly intra-abdominal type[4].

The etiology of desmoid tumors is uncertain. Trauma, hormonal and genetic abnormality is factors that may contribute to the pathogenesis of these tumors. Desmoid tumors often develop during, after pregnancy or following uptake of contraceptive pills[1]. Spontaneous regression is reported to occur after menopause, oophorectomy and after anti-estrogen treatments. Katsuyuki K et al reported a 27-year-old woman presenting with an abdominal desmoid tumor involving the xyphoid and costal chondrium during the eighth month of gestation[5]. Magnetic resonance imaging indicated a 9 x 7.5 cm mass arising from the chest wall. The tumor was radically excised with the xyphoid, a lower part of sternum and the chondrium of the bilateral sixth and seventh ribs. The incision was closed primary without prosthesis. Shashi K et al reported a 22-year-old woman presenting with lower abdominal desmoid tumor during fifth month of gestation[6]. The tumor measured 12.5 x 9.23 cm in size at the suprapubic region. The tumor removal was completed and prolene mesh graft was used afterward. Franco DC et al reported a 42-year-old woman presenting with a 4 x 6 cm abdominal desmoid tumor arising from a cesarean section scar[7]. The tumor was completely resected with double-layer prosthetic mesh. The information from the preceding case report is compatible with this speculation.

Microscopically, there is dense collagenous material with interspersed spindle cells and fibroblasts, which have an abundant eosinophilic cytoplasm[8]. Immunohistochemistry is positive for muscle cell markers such as viementin, alpha smooth muscle actin, muscle actin and desmin. The findings in our patient are in line with the literature.

Imaging study (CT and MRI) may delinate the tumor sites, determine their extent, relationships with adjacent organs and vital structures[9]. This information is important prior to operating. Colonoscopy is indicated to investigate for the presence of Gardner's syndrome.

Complete surgical excision is the treatment of choice for desmoid tumors. Positive surgical margin is a high risk for tumor recurrence. The large soft tissue defect after resection can be achieved by direct repair or by using synthetic materials (Mesh). The other modality of treatment is anti-hormonal drug, NSAIDs and cytotoxic drug. Lim et al. found 33% of desmoid tumors expressed estrogen receptors[10]. Waddell et al reported 2 cases, in which tamoxifen was efficacious, achieving regression of tumors[11]. Non-steroidal, anti-inflammatory drug such as indomethacine, sulindac and anti-estrogens such as tamoxifen are considered as first line pharmacological therapy. Cytotoxic drug such as doxorubicin, dacarbazine, actinomycin-c should be considered for symptomatic, unresectable, clinically aggressive desmoid tumor which do not respond to conventional treatment[12].

In conclusion, the authors report a case of large abdominal desmoid tumor. She was a sporadic case of desmoid tumor associated with pregnancy. This tumor may be associated with trauma and hormonal factors. The patient underwent successful treatment with complete surgical excision. She had no post-operative morbidity, mortality or recurrent tumor at the 8 months follow-up.

References

1. Reitamo JJ, Scheinin TM, Hayry P. The desmoid syndrome. New aspects in the cause, pathogenesis and treatment of the desmoid tumor. Am J Surg 1986; 151: 230-7.
2. Reitamo JJ, Hayry P, Nykyri E, Saxen E. The desmoid tumor. I. Incidence, sex-, age- and anatomical distribution in the Finnish population. Am J Clin Pathol 1982; 77: 665-73.
3. Posner MC, Shiu MH, Newsome JL, Hajdu SI, Gaynor JJ, Brennan MF. The desmoid tumor. Not a benign disease. Arch Surg 1989; 124: 191-6.
4. Gurbuz AK, Giardiello FM, Petersen GM, Krush AJ, Offerhaus GJ, Booker SV, et al. Desmoid tumours in familial adenomatous polyposis. Gut 1994; 35: 377-81.
5. Kunieda K, Saji S, Tanaka Y, Katoh M, Fukada D, Shimokawa K. An abdominal desmoid tumor involving the xyphoid and costal chondrium associated with pregnancy: report of a case. Surg Today 1999; 29: 927-30.
6. Khare S, Hafeez S. Desmoid tumor with pregnancy. J Obstet Gynecol India 2005; 55: 363-4.
7. De Cian F, Delay E, Rudigoz RC, Ranchere D, Rivoire M. Desmoid tumor arising in a cesarean

section scar during pregnancy: monitoring and management. Gynecol Oncol 1999; 75: 145-8.
8. Salloum H, Kanitakis J, Chouvet B, Grimand P, Claudy A. Extra-abdominal desmoid tumor. Microscopic aspects and histogenesis. Ann Dermatol Venereol 1993; 120: 685-8.
9. Sakorafas GH, Nissotakis C, Peros G. Abdominal desmoid tumors. Surg Oncol 2007; 16: 131-42.
10. Lim CL, Walker MJ, Mehta RR, Das Gupta TK. Estrogen and antiestrogen binding sites in desmoid tumors. Eur J Cancer Clin Oncol 1986; 22: 583-7.
11. Waddell WR, Gerner RE, Reich MP. Nonsteroid antiinflammatory drugs and tamoxifen for desmoid tumors and carcinoma of the stomach. J Surg Oncol 1983; 22: 197-211.
12. Kulaylat MN, Karakousis CP, Keaney CM, McCorvey D, Bem J, Ambrus Sr JL. Desmoid tumour: a pleomorphic lesion. Eur J Surg Oncol 1999; 25: 487-97.

ก้อนเนื้องอกเดสมอยด์ขนาดใหญ่บริเวณผนังหน้าท้องสัมพันธ์กับการตั้งครรภ์

วิชิต วิริยะโรจน์, ณรงค์ชัย ยิ่งศักดิ์มงคล, ภาสกร ผาสุกดี, นภารัฐ เริ่มลึก

ก้อนเนื้องอกเดสมอยด์ เป็นเนื้องอกที่พบไม่บ่อย และเป็นชนิดธรรมดาไม่ใช่เนื้อร้ายแต่มีพฤติกรรมชอบลุกลามไปยังอวัยวะข้างเคียง ก้อนเนื้องอกเดสมอยด์กำเนิดมาจากเนื้อเยื่อของผนังกล้ามเนื้อ ส่วนใหญ่พบในผู้หญิงอายุน้อย สาเหตุการเกิดยังไม่ทราบแน่ชัด แต่พบว่าอาจมีความสัมพันธ์กับการผ่าตัด การบาดเจ็บบริเวณนั้น หรือ ภาวะ ฮอร์โมนในร่างกาย ผู้นิพนธ์ได้รายงานผู้ป่วยหญิง อายุ 17 ปี มาโรงพยาบาลด้วยมีก้อนบริเวณผนังหน้าท้องส่วนล่างขณะตั้งครรภ์ได้ 5 เดือน ผู้ป่วยได้รับการตัดชิ้นเนื้อระหว่างการผ่าตัดคลอดจากโรงพยาบาลอื่น แต่ยังไม่สามารถวินิจฉัยได้ หลังจากนั้นก้อนโตขึ้นอย่างรวดเร็วภายหลังจากการตัดชิ้นเนื้อและหลังคลอด ผู้ป่วยได้รับการรักษาโดยการผ่าตัดเอาก้อนออกทั้งหมด พบว่าก้อนมีขนาด 28 x 21 x 18 เซนติเมตร น้ำหนัก 4,900 กรัม ผลการตรวจชิ้นเนื้อพบว่าเป็น เนื้องอกเดสมอยด์ ไม่พบภาวะแทรกซ้อนหลังผ่าตัด และไม่พบว่ามีก้อนขึ้นมาใหม่หลังจากการผ่าตัด 8 เดือน