

(Desmoid Tumor) 7

1 . 2 . 1 . 2 . 2

= Abstract =

Seven Cases of Desmoid Tumor of Trunk

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Background: A desmoid tumor is a rare neoplasm of a mature fibroblast within an extensive collagen matrix. These tumors are classified pathologically as aggressive fibromatosis with local invasiveness, showing recurrences after surgical treatment, although they do not metastasize. Complete surgical excision without leaving a remnant tumor seems to be the primary treatment option, but these tumors often recur after resection. Therefore, alternative treatment strategies, including radiation therapy, nonsteroidal anti-inflammatory agents, anti-estrogen compounds, and cytotoxic chemotherapy, have been employed.

Method: The authors report the clinical characteristics and prognoses after treatment for seven patients who were diagnosed as having desmoid tumors and who underwent surgical excision for therapy during the ten-year period from 1986 to 1997 at Keimyung University Dongsan Medical Center.

Results: There were two males and five females. The most common chief complaint was a slowly growing painless mass. The duration of the symptom before diagnosis was quite different from patient to patient. The locations in the trunk were scattered diffusely: the shoulder, the chest wall, the abdominal wall, the retroperitoneum, and the pelvic cavity. The tumors were well visualized by imaging techniques such as ultrasonograms, CT scans, and MRI. Four of the seven patients experienced recurrent tumors after surgical excision; two are dead. The tumors in the recurrent patients were excised incompletely at initial surgery.

Conclusion: Although the uncommon, solid desmoid tumors which develop in the musculoaponeurosis of the trunk are benign pathologically, they were recurrent after surgical excision when the resection margin is incomplete. Therefore, complete excision is the only curative therapeutic modality.

Key Words: Desmoid tumor, Fibromatosis, Trunk

Desmoid

“tendonlike”

1832 John MacFarlane

¹⁸⁾ 1838 J.

69-7, ☎ 700-100

Muller

‘desmoid tumor’

Tel: 0562-44-2662, Fax: 0562-45-5311

²²⁾

: 1998 4 6 , : 1998 9 7

0.03%,

3 4%

144 : 56 1 1999

2 5 1 :
15) 4 가 ; 가
1 4) 가 ,

ner's syndrome Gardner's syndrome ;
33% . 2% Gard-

가 가 .

8%가

가 70% 가
5) 가
가 1992 2 3×4 cm
가

2.

28 , , 가 /24
21 , , : 1
7 , , 6

: 1988 (limping
gait)

1.

: , /24
: 2 6

: 1983

가

6×7×5 cm

9.0×6.0×5.0 cm

가

가

가

가

: 17.5×16×6 cm

가

3.

: , /49
: 3 1/2

: 4×3×3 cm

1.5 cm

가

, 4×3×3 cm

가 , 1995 5 (9)
43 mm

, 1995 7

가

가

가

가

1995 10

가
가

(linear streaky density)
가

3

96 6

4.

: , /40

: 1 6

(2) 3×3 cm

가

1

2

Fig. 1. Case 3. CT: Slightly increased size of mass in previous operation site and inguinal area.

Fig. 2. Case 4. A. CT: Well marginated ovoid solid mass in anterior aspect of the right 2nd rib. B. CT: Recurrent mass in the right 2nd rib.

가 , 3 cm
 2 가 : 5×15×0.8 cm
 :
 ; 5×15×0.8 cm
 ; 가
 : 1 4
 follow up
 5.
 : , /66
 : 11 2×
 2 cm 가 가 5×6 cm
 : 1993 12
 가 : 3×4×7 cm 가
 : 4×5 cm
 ; 8.0×5.0×2.5 cm
 ;
 : 1 9
 6.
 : , /32
 : 3 가
 : 3.8×5.1×2.1 cm
 가
 10×5×8 cm
 가 (extravesical space) 가
 : 12×10 cm 가
 ; 가
 가 8.0×6.0
 ×5.2 cm 7.2×6.0×5.5 cm 가
 가 ;

Fig. 3. Case 6. MRI: 10×5×8 cm sized well defined huge lobulating mass in pelvic cavity

Fig. 4. Case 7. A. MRI: Lobulated dumbbell shaped mass in left pelvic cavity. B. The gross specimen is a relatively well-demarcated dumbbell-shaped rubbery mass, C. Cut section shows a glistening pale tan, trabeculated surface with a partial myxoid change

가 .

가 : 9 .

7. 가 , 가

: , /32 , 가

: 3 가

가 : 3 .

9,12,14,27) 1832 1838

J. Muller desmoid tumor 22)

1928 Ewing J. 가 가 가

11) 가 ,

10.5 × 15 × 8 cm .

: 가 , (acellular fibroma)

; 16.0 × 9.8 × 7 cm (cellular low grade fibrosarcoma)

가 가 가

; 가 (fascicle) 3가 가

가 가 가 (girdle muscle)

가 가 가

: 가 49%, 43% 8% 가

가 90%가

²⁴⁾ 가 가

가 가

가 ²⁵⁾ 가 가

20 가 , 가 ,

3 ,

^{15,25)} Fernand (spermatic cord) ¹³⁾ 1 가

1 ¹⁷⁾ 1950

가 (Gardner's syndrome)

가 ^{13,20)} 75

S ¹⁹⁾

1 , ¹⁶⁾

, 가 ,

(Gardner's syndrome) 가

, ²³⁾ 2

가 가 ¹³⁾ 50%

가 가

arm 가 5 long

,

가 가

^{8,10,21)} 가 ^{5,15)} Bauernhofer goserelin acetate tamoxifen

4 (4.3), 가 (27.2), (43.9

), (68.1) , goserelin acetate

, 가 ⁶⁾ Patel doxorubicin dacarbazine

가 ²³⁾ (sulindac), c-AMP modulator(ascorbic acid, theophyllin, testolactone)

가 ²⁸⁾ 7 4 가

^{24,25)} 1 3 5 5

가 89 Reitamo 2

3 ²⁴⁾ 100 2 , 가 가

, ²³⁾ 가

8 11 1986 7 1997
 27 20
 7 , , ,
 7 4
 6
 1
 6
 , 3 10 1
 2 8 ,
 1 1 9 1
 1 3

가 가

REFERENCES

1) , , :
 1 44(1): 146, 1993
 2) , , , :
 10(8): 581, 1968
 3) , , , : 1
 43(3): 466, 1992
 4) , , : 1
 22(11): 67, 1980
 5) Bataini JP, Belloir C: Desmoid tumor in adults. The role of radiotherapy in their management. *Am J Surg* 754
 6) Bauernhofer T, Stoger H, Schmid M, Smola M: Sequential treatment of recurrent mesenteric desmoid tumor. *Cancer* 77: 1061, 1996
 7) Bülow S: Incidence of associated disease in familial polyposis coli. *Semin Surg Oncol* 3: 84, 1987
 8) Bridge JA, Sseekantaih C, Mouron B, Neff JR, Sandberg AA, Wolman SR: Clonal chromosomal abnor-

malities in desmoid tumors. Implication for histopathogenesis. *Cancer Res* 69: 430, 1992
 9) Conran RS: Robbins pathologic basis of disease. WB Saunders, Philadelphia, 1994, p1265
 10) Coon H, Jensen S, Hoff M, Holik J, Plaetke R, Remherr F, et al: Agenome-wide search for genes predisposing to manic depression, assuming autosomal dominant inheritance. *Am J Hum Genet* 52: 1234, 1993
 11) Ewing J: Neoplastic disease. 3rd ed. Philadelphia, WB saunders, 1928
 12) Gupta TK, Brasfield RD: Extraabdominal desmoid tumors. *Arch Surg* 98: 109, 1969
 13) Karakousis CP, Bejian RA: Mesenteric fibromatosis in Gardner's syndrome. *Arch Surg* 113: 998, 1978
 14) Khorsand J, Karakousis CP: Desmoid tumor and their management. *Am J Surg* 149: 215, 1985
 15) Kiel KD, Suit MS: Radiation therapy in the treatment of aggressive fibromatosis(desmoid tumor). *Cancer* 54: 2051, 1984
 16) Kransdorf MJ, Jelinek JS, Moser RP, et al: Magnetic resonance appearance of fibromatosis. Areport of 14 cases and review of the literature. *Skeletal Radiol* 19: 495, 1990
 17) Lai FM, Allen PW, Chan LW, Chan PS, Cooper JE, Mackenzie TM: Agressive fibromatosis of the spermatic cord. *Am J Clin Pathol* 104: 403, 1995
 18) MacFarlane J: Clinical report on the surgical practice of Glasgow Royal infirmary. Glasgow: D. Robertson, 1838, p63
 19) Magid M, Fishman E, Jones B, et al: Desmoid tumor in Gardner's syndrome: Use of computed tomography. *AJR* 1141, 1984
 20) McAdam WAF, Goligher JC: The occurrence of desmoids in patients with famillial polyposis coli. *Br J Surg* 57: 618, 1970
 21) Miyaki M, Konishi M, Kikuchi-Yanosshita R, Economoto M, Tanakas K, Takahashi H, et al: Coexistence of somatic and germ-line mutation of APC gene in desmoid tumors from patients with familial adenomatous polyposis: *Cancer Res* 53: 5079, 1993
 22) Muller J: Ueber den feiem Bau and die Formen der kranfhafteu Geschwulate. Belrin, G. Reimer, 1838, p60
 23) Patel SR, Evans HL, Benjamin RS: Combination chemotherapy in adult desmoid tumor. *Cancer* 72: 3244, 1993
 24) Reitamo JJ: The desmoid tumor I. Incidence, sex-, age-and anatomical distribution in the Finnish popu-

- lation. Am J Clin Pathol 77: 665, 1982
- 25) Reitamo JJ: The desmoid tumor IV. Choice of treatment, result, and complications. Arch Surg 118: 1318, 1983
- 26) Reitamo JJ, Scheinin TM, Hayry P: The desmoid syndrome. New aspects in the cause, pathogenesis and treatment of the desmoid tumor. Am J Surg 151: 230, 1986
- 27) Schwarz SI: Principles of surgery. 6th ed., McGraw-Hill, New York, 1994, p 1485
- 28) Waddell WR: Treatment of intraabdominal and abdominal wall desmoid tumor with drugs that affect the metabolism of cyclic 3,5'-adenosine monophosphate. Ann Surg 181: 299, 1975