

# A giant, rapidly growing intra-abdominal desmoid tumor of mesenteric origin in an adolescent male: A case report and literature review

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**Abstract.** A desmoid tumor is a fibroblastic proliferation of mesenchymal origin, which has no metastasizing potential but is locally aggressive. Although treatment has shifted to observation and active surveillance for newly diagnosed patients with desmoid tumors, intra-abdominal mesenteric tumors or tumors that persistently grow and provoke symptoms may need prompt surgical treatment. There have only been a small number of case reports that illustrate large sporadic intra-abdominal mesentery-deriving desmoid tumors in which the longest diameter was  $\geq 19$  cm. In the present study, an adolescent male patient with a rapidly growing 38-cm long sporadic intra-abdominal desmoid tumor of mesenchymal origin is reported. The patient was treated with chemotherapy followed by surgical resection due to non-responsiveness and progression of symptoms, then with maintenance adjuvant chemotherapy to prevent recurrence due to the large size of the tumor. Despite the rapid growth of the tumor and its high occupancy in the intra-abdominal cavity, an R0 resection was successful with organ preservation. The patient has been recurrence-free for 2 years, and further follow-up is expected in the future.

## Introduction

A desmoid tumor, also termed desmoid-type fibrosis or aggressive fibromatosis, is a fibroblastic proliferation of mesenchymal origin, which has no metastasizing potential but is locally aggressive. It is a rare disease, and in 2013, the incidence was reported as 5-6 newly diagnosed cases/1 million

individuals/year (1), with the peak incidence occurring in individuals aged 30-40 years old, with a slightly higher incidence in women (2,3). Desmoid tumors are known to occur in women taking oral contraceptives, women during or after pregnancy, patients with a history of previous trauma or surgery, and in patients with genetic conditions such as familial adenomatous polyposis (FAP) and Gardner's syndrome (4,5). The mortality is rarely reported; however, in patients with FAP and intra-abdominal desmoid tumors, the overall mortality has been reported to be  $\leq 76\%$  for stage IV tumors in a single center cohort study (6). The tumor develops from the deep soft tissue that is located throughout the body, including the abdominal wall, intra-abdominal cavity, mesentery, gastrointestinal tract, extremities, chest wall, breast, and the head and neck (5). Surgical tumor resection has been used as the primary management for several decades alongside systemic therapy or radiotherapy (7). However, previously, treatment has shifted to active surveillance of the tumor after the initial diagnosis due to its indolent course and possibility for spontaneous regression (SR) (8,9). While prompt medical or surgical treatments are reserved for symptomatic and progressive tumors, intra-abdominal mesenteric tumors may also require early intervention due to the high morbidity risk (9). The present report describes an adolescent male patient with a rapidly growing 38-cm long intra-abdominal desmoid tumor of mesenchymal origin.

## Case report

An 18-year-old Asian male patient with no known past medical history was referred to a tertiary academic medical center (Severance Hospital, Seoul, Korea) in July 2020 via the outpatient clinic with an intra-abdominal mass. The patient had been having upper abdominal discomfort for 2-3 months, which gradually became worse and was accompanied by abdominal pain. The patient had not experienced any traumatic events or a familial history of cancer. As the patient was obese at the first visit (height, 166 cm; weight, 88 kg; body mass index,  $31.9 \text{ kg/m}^2$ ), the mass was not observed by sight but it was firmly palpable. Abdominal sonography and abdomino-pelvic computed tomography (APCT) initially revealed a 26-cm long heterogeneously enhanced, lobulated

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mass with necrotic components (Fig. 1). The mass appeared to originate from the omento-mesentery with feeding vessels arising from the engorged branches of the superior mesenteric artery, and although a large portion of the small bowel was lateralized to the left, there were no definite signs of organ invasion. Positron emission tomography-CT and whole-body bone scans indicated no evident distant metastasis. After a transcutaneous sonography-assisted biopsy, a spindle cell proliferative lesion in a myxoid background was revealed and immunohistochemical (IHC) stains were negative for anaplastic lymphoma kinase (ALK), S-100, mucin 4, CD34,  $\alpha$ -smooth muscle actin ( $\alpha$ -SMA) and SRY-box transcription factor 10 (SOX10), and positive for  $\beta$ -catenin, which indicated that a desmoid-type fibromatosis diagnosis was a high possibility (Fig. 2). The slide preparation for IHC and H&E staining was performed as follows: Formalin fixation was conducted by submerging tissue in 10% neutral buffered formalin for 24 h at room temperature, followed by dehydration with ethanol, and clearance with xylene of the tissue. Paraffin infiltration was then achieved with an automatic tissue processing machine (HistoCore PELORIS II; Leica Microsystems GmbH), and the block was cut into 4- $\mu$ m wide slices. For IHC staining, deparaffinization, antigen retrieval, primary and secondary antibody reactions, and counterstaining were further performed with an automated IHC staining system (Table I). Hematoxylin and eosin staining was also performed using the automated system (VENTANA HE 600 system; Roche Tissue Diagnostics) as follows: Deparaffinization, rehydration, hematoxylin staining, differentiation of the nucleus, bluing, eosin staining, dehydration and clearing (Table II). The light microscope used was a BX43 microscope (Olympus Corporation). No genetic analysis was performed for  $\beta$ -catenin (*CTNNB1*) or adenomatous polyposis coli (*APC*) mutations.

The upfront surgical resection seemed challenging due to the high occupancy of the tumor in the abdominal cavity, so the patient was first offered treatment with chemotherapy using a regimen of doxorubicin (20 mg/m<sup>2</sup> infusion for 1 h on days 1-3) and dacarbazine (300 mg/m<sup>2</sup> infusion for 1 h on days 1-3) by a pediatric oncologist. Sperm banking was conducted prior to the chemotherapy. After two cycles of the regimen with 3-week intervals, the tumor had increased to 32 cm in diameter in 1 month from the first visit, increasing the abdominal discomfort and pain experienced by the patient. Thus, the chemotherapy regimen was changed to etoposide (100 mg/m<sup>2</sup> infusion for 3 h on days 1-5), carboplatin (200 mg/m<sup>2</sup> infusion for 3 h on days 1 and 2) and ifosfamide (1,800 mg/m<sup>2</sup> infusion for 3 h on days 1-5). However, the non-responsiveness of the tumor and the side effects of the chemotherapy, such as neutropenic fever and general weakness, led to repeated patient re-admissions and made it hard to further delay the surgical management. After the decision for surgical resection of the tumor was made, the patient was injected with antibiotics (piperacillin-tazobactam; 4.5 g mixed in 100 ml saline; three times a day) for 8 days for the neutropenic fever, transfused with platelet concentrates (PCs) for thrombocytopenia and was provided with total parenteral nutrition for 3 days. Preoperative consultations were conducted with upper gastrointestinal, hepatobiliary and vascular surgeons for possible cooperation. On the day before the surgery, percutaneous angiographic embolization was

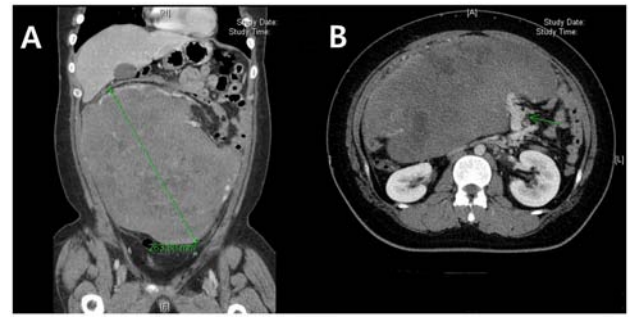


Figure 1. CT scan image of the tumor. Initial (A) coronal and (B) axial views of the intra-abdominal tumor using abdomino-pelvic CT. The feeding vessels emerged from the engorged branches of the superior mesentery artery (green arrow), and definite organ invasion was not observed. CT, computed tomography.

performed on the feeding vessel of the tumor, which branched from the superior mesenteric artery, to prevent intra-operative bleeding. The operation was conducted 2 months after the first outpatient clinic visit.

The surgeon in charge was specialized in colorectal surgery with >10 years of clinical experience in the tertiary academic medical center (Severance Hospital, Seoul, Korea). At the operating field, a long midline incision was insufficient to expose the mass completely, so a crossed incision with additional transverse upper midline incisions were made (Fig. 3). Tumor dissection was performed by peeling off the adjoined bowel, mesentery and retroperitoneum. An ileocectomy with ileocolic anastomosis was conducted due to ileocecal vascular involvement, and the vascular surgical team engaged in dissecting the superior mesenteric vessel and its angioplasty. The total operation time was 311 min and estimated blood loss was 3,000 ml, while intra-operative blood transfusions of 10 units of packed red blood cells, 6 units of PC and 6 units of fresh frozen plasma were performed. After the operation, the patient was admitted to the Surgical Intensive Care Unit, and was moved to the General Surgical Ward 2 days later. Liquid and soft diet was started on postoperative day 6 and 9, respectively, and the patient was discharged at postoperative day 15. The final surgical pathology revealed a 38-cm long desmoid-type fibrosis and a clear resection margin, and the cut surface revealed diffuse myxoid, hemorrhagic and multilocular cysts with yellowish sebaceous fluid (Fig. 4). The IHC staining results were the same as the initial findings: Negative for ALK, S-100, mucin 4, CD34,  $\alpha$ -SMA and SOX10, and positive for  $\beta$ -catenin.

On the first outpatient clinic visit after discharge (post-operative month 1), the patient was planned for maintenance adjuvant chemotherapy (vinblastine 6 mg/m<sup>2</sup> infusion for 1 h plus methotrexate 30 mg/m<sup>2</sup> injection on days 1, 8, 15 and 23) by the pediatric oncologist, due to the large size of the tumor despite *en bloc* resection with an intact tumor capsule. The chemotherapy was started on postoperative day 40 after the skin wound had completely healed and ended on postoperative year 1 (12 cycles total). After the first APCT at 1 month postoperatively, MRIs were performed every 2 cycles of chemotherapy (2 months) for response evaluation, to follow up the subcentimeter T2-hypointense nodular lesion in the upper anterior abdomen, which appeared to be an uncomplicated

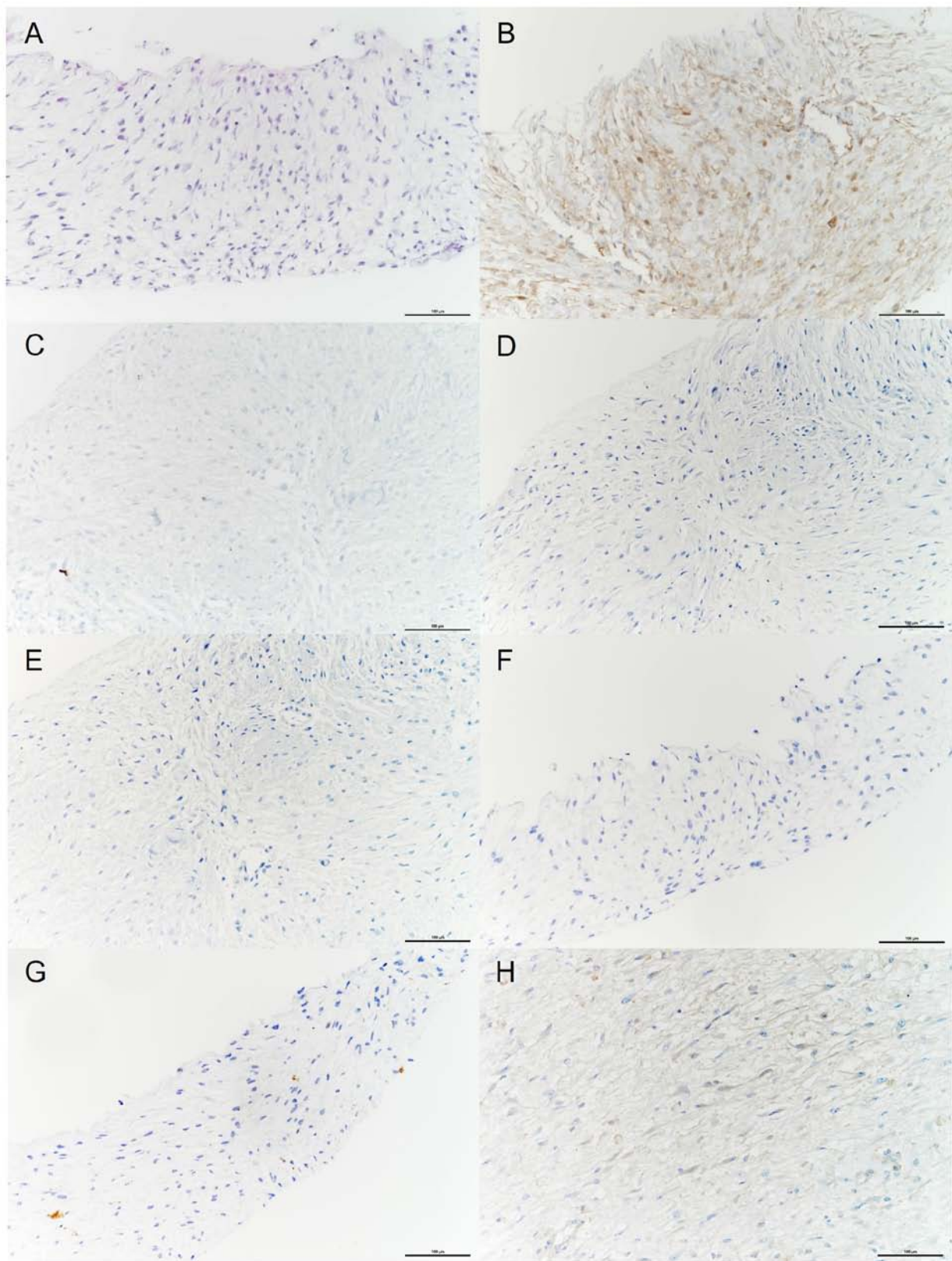


Figure 2. Immunohistochemical staining of the tumor. Scale bars indicate 100  $\mu$ m. Proliferation of spindle cells with minimal atypia in the myxoid stroma was observed with (A) hematoxylin and eosin staining. Immunohistochemical staining was positive for (B)  $\beta$ -catenin, and negative for (C) anaplastic lymphoma kinase, (D) S-100, (E) mucin 4, (F) CD34, (G)  $\alpha$ -smooth muscle actin and (H) SRY-box transcription factor 10.

postoperative change. Rare and occasional upper abdominal discomfort that spontaneously subsided and elevated alanine transaminase (50-80 IU/l) controlled with oral ursodiol

(100 mg three times a day) were noted, and prophylactic oral sulfamethoxazole/trimethoprim (960 mg per day) was maintained during chemotherapy.

Table I. Details of procedures for IHC staining.

Procedures	ALK	S-100	Mucin 4	CD34	SMA	SOX-10	$\beta$ -catenin
Deparaffinization							
Solvent	EZ prep (10x)	EZ prep (10x)	EZ prep (10x)	Clearify Clearing Agent	Clearify Clearing Agent	EZ prep (10x)	EZ prep (10x)
Supplier	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	Roche Tissue Diagnostics	Roche Tissue Diagnostics
Temperature, °C	75	75	75	25	25	75	75
Duration, min	4	4	4	1	1	4	4
Antigen retrieval							
Reagent	Cell conditioning solution-1	Cell conditioning solution-1	Cell conditioning solution-1	EnV FLEX TRS High pH (50x)	EnV FLEX TRS High pH (50x)	Cell conditioning solution-1	Cell conditioning solution-1
Supplier	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	Roche Tissue Diagnostics	Roche Tissue Diagnostics
Temperature, °C	100	100	100	97	97	100	100
Duration, min	64	38	38	30	30	38	38
Primary antibody reaction							
Dilution	Prediluted	1:2,000	1:1,000	1:100	1:1,000	1:100	1:400
Catalogue no.	IS641	Z0311	MABT395	M7165	M0851	383R-14	224M-15
Supplier	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	MilliporeSigma	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	MilliporeSigma	MilliporeSigma
Temperature, °C	37	37	37	37	32	37	37
Duration, min	32	32	32	20	20	32	32
Secondary antibody reaction <sup>a</sup>							
Catalogue no.	760-700	05269806001 (760-500)	05269806001 (760-500)	GV800, GV823, GV900	GV800, GV823, GV900	05269806001 (760-500)	05269806001 (760-500)
Product	Optiview HQ universal linker	ultraView Universal DAB	ultraView Universal DAB	EnVision FLEX HRP	EnVision FLEX HRP	ultraView Universal DAB	ultraView Universal DAB
Supplier	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	Roche Tissue Diagnostics	Roche Tissue Diagnostics
Temperature, °C	37	37	37	37	37	37	37
Duration, min	8	8	8	20	20	8	8
Counterstain							
Counterstain	Hematoxylin	Hematoxylin	Hematoxylin	Hematoxylin	Hematoxylin	Hematoxylin	Hematoxylin
Temperature, °C	37	37	37	37	37	37	37
Duration, min	8	8	8	4	4	8	8



Table I. Continued.

Procedures	ALK	S-100	Mucin 4	CD34	SMA	SOX-10	$\beta$ -catenin
Automated IHC staining system							
Instrument	Ventana benchmark XT	Ventana benchmark XT	Ventana benchmark XT	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	Ventana benchmark XT	Ventana benchmark XT
Supplier	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Roche Tissue Diagnostics	Dako; Agilent Technologies, Inc.	Dako; Agilent Technologies, Inc.	Roche Tissue Diagnostics	Roche Tissue Diagnostics

<sup>a</sup>All secondary antibodies were purchased prediluted and horseradish peroxidase was the conjugate for all secondary antibodies. ALK, anaplastic lymphoma kinase; SMA,  $\alpha$ -smooth muscle actin; SOX-10, SRY-box transcription factor 10; IHC, immunohistochemical.

After completion of chemotherapy, CT and MRI were alternately performed every 3 months until 2 years after surgery and every 6 months thereafter. The patient was admitted to hospital 1 year after the surgery due to small bowel obstruction lasting for 5 days. The patient has been followed up for 2.5 years and is recurrence-free. The patient will be followed up for 5 years, but the follow-up period may be extended due to the young age. It is assumed that the tumor will not recur, but if it does, the multidisciplinary team will discuss the possibility of surgical resection or enroll the patient in a clinical trial.

## Discussion

A desmoid tumor is defined as ‘clonal fibroblastic proliferations that arise in the deep soft tissues and is characterized by infiltrative growth and a tendency toward local recurrence but an inability to metastasize’ by the World Health Organization (10). While 10-15% of desmoid tumors derive from germline mutations of the *APC* gene and present as FAP, the majority of desmoid tumors are sporadic and are associated with somatic mutations of the *CTNNT1* gene (7). The mutation of either gene leads to alterations in the Wnt/ $\beta$ -catenin signaling pathway and eventually to uncontrolled fibroblastic proliferations (11). The *APC* and *CTNNT1* genes are known to be mutually exclusive, and mutational analysis of the desmoid tumor biopsied specimen is recommended in the initial diagnostic phase (10). The etiologies of desmoid tumors, other than hereditary syndromes, include a history of previous trauma or the hormonal effects of estrogen during pregnancy (12,13). In the present case study, the patient did not have any medical or traumatic history, and mutational analysis was not performed, as relieving the symptomatic tumor was the treatment priority at the time. In the future, a post hoc mutational analysis is planned since, although the surgery has already been performed, detecting the triggering genetic mutation may be a requirement as the patient gets older.

Despite their high tendency for local recurrence, desmoid tumors are likely to stay dormant or to spontaneously regress in 20-30% of patients (14,15). Penel *et al* (16) demonstrated that the 2-year event-free survival rate was not significantly different between a surgically and non-surgically treated group in all tumor locations (53 vs. 58%, respectively;  $P=0.415$ ) and in locations including the abdominal wall, intra-abdominal, breast, digestive viscera and lower limb (70 vs. 63%, respectively;  $P=0.413$ ). Burtenshaw *et al* (17) demonstrated that among 109 patients with intra-abdominal and abdominal wall desmoid tumors who were initially observed, 55 (50.5%) patients underwent medical or surgical treatments either due to disease progression or symptom escalation, while 31 (28.4%) and 23 (21.1%) patients exhibited stable disease (SD) and SR, respectively. Accordingly, the initial management of desmoid tumors has evolved into observation and active surveillance with repeated CT or MRI scans.

However, symptomatic and progressive tumors may be indicated for surgical or medical intervention. In particular, FAP-associated desmoid tumors are typically managed more actively due to their more aggressive tumor course (10). Medical management includes antihormonal therapy, administration of tyrosine kinase inhibitors (TKI) and chemotherapy.

Table II. Details of procedures for hematoxylin and eosin staining.

Procedure	Reagent	Catalogue no.	Supplier	Temperature, °C	Duration, sec
Deparaffinization	Organic solution	07095163001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	150
Rehydration	Transfer fluid	06544380001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	20
Hematoxylin staining	Hematoxylin	07024282001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	60
Differentiation of the nucleus	Differentiation solution	06544339001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	120
Bluing	Bluing	06544347001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	30
Eosin staining	Eosin	06544304001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	300
Dehydration	Transfer fluid	06544380001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	20
Clearing	Organic solution	07095163001	Roche Tissue Diagnostics; Roche Diagnostics, Ltd.	40±3	10

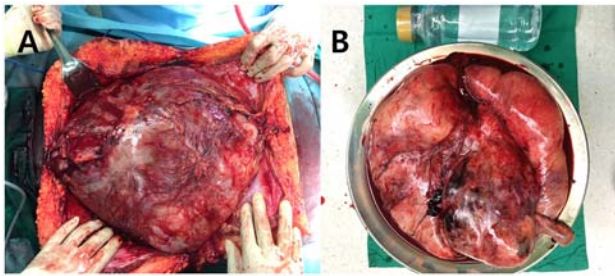


Figure 3. Gross finding of the tumor. The appearance of the tumor (A) before and (B) after the resection.

The only TKIs that have been demonstrated to be effective in randomized trials are sorafenib and pazopanib. Sorafenib demonstrated a significant higher overall response rate of SR compared with that of the placebo group (33 vs. 20%, respectively) and a longer 2-year progression-free survival (PFS) rate compared with that of the placebo group (81 vs. 36%, respectively; HR 0.13;  $P < 0.001$ ) (18). Available chemotherapy regimens include methotrexate plus vinblastine or vinorelbine, oral vinorelbine, doxorubicin plus dacarbazine, methotrexate with vinca alkaloids, anthracycline-based regimens and pegylated liposomal doxorubicin (7,9). Furthermore, the methotrexate with vinblastine regimen was investigated in two phase II trials; Azzarelli *et al* (19) demonstrated a 67% 5-year PFS and Skapek *et al* (20) reported that 69% of patients had SD or SR. In June 2018, a meeting for an evidence-based joint global consensus on the management of desmoid tumors was held by experts from the association of European Reference Network on Rare Adult Solid Cancers, the European Organization for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group, Sarcoma Patients EuroNet and The Desmoid Tumor Research Foundation, in Milan, Italy. The published papers that followed summarized the consensus

that, as mesenteric desmoid tumors are close to vital organs, clinicians should make an earlier decision towards their active treatment (8,9,21).

The assessment of the treatment effects and tumor follow-up examinations are typically implemented with radiological assessments using CT or MRI. There are currently no standardized and validated response criteria for desmoid tumors, but the majority of previous studies used the Response Evaluation Criteria in Solid Tumors (10).

In the present case study, the tumor grew 20% in diameter in 6 weeks after applying two cycles of doxorubicin and dacarbazine. Doxorubicin and dacarbazine have been administered for fibrous tumors and FAP-related intra-abdominal desmoid tumors, and in one study (22), implementation of the regimen caused significant tumor regression in all 7 patients with intra-abdominal mesenteric desmoid tumors. Although the evidence is weak due to the small number of cases and, unlike in the present case, the patients in the aforementioned study had FAP-related tumors, the finding that the tumor grew rapidly in the present study despite the chemotherapy treatment suggests that it may have been due to the nature of the tumor rather than the effect of the chemotherapy.

Adjuvant chemotherapy with a regimen of vinblastine and methotrexate was performed for the patient in the present report to reduce recurrence. Postoperative use of this regimen has rarely been reported but it has been applied to recurrent or progressive desmoid tumors. Garbay *et al* (23) reported that among 27 patients who were treated with vinblastine (6 mg/m<sup>2</sup>) and methotrexate (30 mg/m<sup>2</sup>) on days 1, 8, 15 and 23 for recurrent or progressive desmoid tumors, 4 (15%), 14 (52%) and 9 (33%) patients exhibited partial response, SD and progression, respectively. The number of chemotherapy cycles was not specified in the study. In a study reporting pediatric patients (24), vinblastine (6 mg/m<sup>2</sup>) and methotrexate (30 mg/m<sup>2</sup>) given for 10 months to a 10-year-old patient with

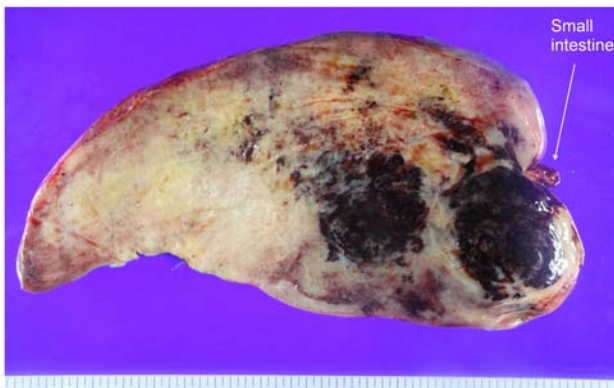


Figure 4. Cross section of the tumor. The distance between long bars is 1 cm, and the distance between long and short bars is 0.5 cm.

an incompletely resected tumor and for 2 years to a 14-year-old patient with a recurrent tumor resulted in complete remission and SD, respectively. A phase-II randomized clinical trial compared pazopanib with vinblastine plus methotrexate in patients with progressive desmoid tumors and reported that the 6-month non-progression rate was 83.7 and 45%, respectively (25). However, the 2-year PFS was 67.2 and 79%, respectively. The present study is one of the few to describe adjuvant therapy with vinblastine and methotrexate after complete tumor removal.

Radiation therapy (RTx) before or after the surgery was not implemented in the present study. Preoperative RTx was not considered, as it was assumed that the tumor was resectable. Currently, adjuvant RTx is rarely employed, and when it is, it is used for retroperitoneal or recurrent tumors (9,21). Furthermore, as the tumor in the present study was extensively adjacent to the bowel, it was considered that postoperative RTx may be complicated by enteritis and that the patient would be followed up well regardless, as CT and MRI scans are easily available in South Korea. Additionally, the angiographic embolization that was performed 1 day before the surgery was conducted to help the surgeons control the bleeding more easily during the surgery, rather than to decrease the size of the tumor. Unfortunately, the procedure did not prevent intra-operative major bleeding, since various minor vessels caused oozing bleeding.

There are only a small number of case reports describing patients with large sporadic intra-abdominal mesentery-derived desmoid tumors  $\geq 19$  cm in diameter, with no family history of cancer and no operative or traumatic history (Table III) (26-31). All patients in the aforementioned reports were male, with a mean age of 33 years old. It is unclear whether the reason these tumors appear to occur more frequently in men is due to a biological cause or a delay in disease recognition. In addition, the majority of the tumors reported originated from the ileal mesentery. The largest sporadic tumor was reported by Williams *et al* (26), in which the patient exhibited a 45-cm long tumor. The majority of patients underwent upfront surgery (5 out of 6 patients) and genetic analysis was not performed in the majority of cases (5 out of 6 patients).

A limitation of the present study was that it lacked genetic screening of the patient for FAP, Gardner syndrome and mutation of the *CTNNT1* gene. However, to the best of

Table III. Case reports of patients with a sporadic intra-abdominal mesentery-derived desmoid tumor  $\geq 19$  cm in diameter.

First author/s, year	Country	Age, years	Sex	Duration of symptoms	Location	Tumor, diameter cm	Genetic analysis	Treatment	F/U, months	Recurrence	(Refs.)
Williams <i>et al</i> , 2016	USA	33	Male	Few months	Ileocolic mesentery <sup>a</sup>	45	NA	Neoadj therapy <sup>b</sup> and surgery	NA	NA	(26)
Mizuta and Tsunemi, 2018	Japan	17	Male	6 months	Transverse mesocolon	30	NA	Surgery	NA	NA	(27)
Sioda <i>et al</i> , 2020	USA	24	Male	4 weeks	Ileocolic mesentery	31	APC mutation (-)	Surgery	NA	NA	(28)
El-Helou <i>et al</i> , 2020	Lebanon	34	Male	NA	Ileocolic mesentery <sup>a</sup>	23	NA	Surgery	12	None	(29)
Kuwabara <i>et al</i> , 2021	Japan	51	Male	Acute	Proximal ileal	19	NA	Surgery	36	None	(30)
Elhaddad <i>et al</i> , 2022	USA	38	Male	3 months	Ileocolic mesentery	40	NA	Surgery	5	None	(31)

<sup>a</sup>Exact origin is not specified, but it appears to be of ileocolic mesenteric origin due to tumor involvement. <sup>b</sup>Neoadjuvant imatinib was administered as a gastrointestinal stromal tumor was first suspected. F/U, follow-up; Neoadj, neoadjuvant; NA, not available.

our knowledge, the present study reported the third largest sporadic intra-abdominal mesentery-derived desmoid tumor with rapid growth. Considering the rarity of the disease and the fact that the treatment for mesentery-derived oversized desmoid tumors has not been fully established, the present case study may provide direction for the treatment of similar conditions in the future.

In conclusion, the present study reported the case of a patient with a 38-cm intra-abdominal desmoid tumor, who was treated with chemotherapy followed by surgical resection due to non-responsiveness and progression of symptoms, then with maintenance adjuvant chemotherapy to prevent recurrence due to the large size of the tumor. The patient has been recurrence-free for 2 years, and further follow-up is planned. In the future, prompt surgical resection may be required for rapidly growing symptomatic intra-abdominal desmoid tumors of mesenteric origin.

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### Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

### Authors' contributions

SJK contributed to medical record review, patient data analysis, literature research, and writing. JWH contributed to conceptualization and review of this paper. TY contributed to medical record review and obtaining medical images. HC contributed to specimen collection and interpretation. YDH contributed to conceptualization, review and supervision of this paper. SJK and YDH confirm the authenticity of all the raw data. All authors read and approved the final version of the manuscript.

### Ethics approval and consent to participate

The study was conducted according to the guidelines of the Declaration of Helsinki and approved by The Ethics Committee of Severance Hospital of Yonsei University College of Medicine (IRB no. 4-2023-0487; Seoul, South Korea). Written informed consent was obtained from the patient.

### Patient consent for publication

Written informed consent was obtained from the patient for publication of the data and images in this case report.

### Competing interests

The authors declare that they have no competing interests.

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