

A Novel Method to Treat Progressive Desmoid Tumors Involving Neurovascular Bundles: A Retrospective Cohort Study

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BACKGROUND: More effective therapies are needed to treat progressive desmoid tumors when active surveillance and systemic therapy fail.

OBJECTIVE: To assess the efficacy and safety of sandwich isolation surgery on the local control of progressive desmoid tumors involving neurovascular bundles.

METHODS: A total of 27 patients with progressive desmoid tumors at extremities involving neurovascular bundles who received surgery at our hospital between August 2014 and August 2018 were identified. A total of 13 patients received sandwich isolation surgery, in which R2 resection was performed in neurovasculature-involving regions, and a biomaterial patch was used to envelop involved neurovascular structures and isolate residual tumors. In non-neurovasculature-involving regions, wide resection was performed without isolation. A total of 14 patients received traditional surgery, which included tumor resection without isolation procedure.

RESULTS: In sandwich isolation group, tumor progressions and local recurrences occurred in 3 patients outside the isolated neurovasculature-involving regions. However, no progressions or recurrences occurred in any patients in the isolated neurovasculature-involving regions where R2 resection was performed. Sandwich isolation surgery group and traditional surgery group shared similar baseline clinical characteristics. The estimated 3-yr event-free survival rate was 76.9% after sandwich isolation surgery, and 32.7% after traditional surgery ($P = .025$). Patients who received sandwich isolation surgery were less likely to have local recurrence (hazard ratio: 0.257, $P = .040$). No complications were noted except intermittent mild pain in operative regions (2 cases).

CONCLUSION: Sandwich isolation surgery is effective and safe for local control of desmoid tumors involving neurovascular bundles.

KEY WORDS: Desmoid tumor, Local recurrence, Sandwich isolation surgery

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Desmoid tumors, also known as deep or aggressive fibromatosis, consist of monoclonal myofibroblastic proliferation that may arise from musculo-aponeurotic stromal elements,¹ accounting for <3% of all soft tissue tumors.¹⁻⁴ Despite the lack of metastatic potential, desmoid tumors are locally infiltrative.⁵⁻⁷ Clinical presentation includes severe pain, deformity, swelling, loss of function,

bowel obstruction or perforation, and/or threat to vital organs.⁸ Although active surveillance is replacing surgery as the initial approach after diagnosis, surgical resection is still one of the treatment choices for progressive desmoid tumors during surveillance, especially when systemic therapy fails.^{4,5,9-14}

Traditionally, the surgical strategy for preventing recurrence is to obtain R0 margins by wide resection.^{15,16} However, it is not uncommon for residual tumor to be present around involved neurovascular bundles after surgery (R1/R2 margins). McCarville et al¹⁷ found that patients with desmoid tumors invading adjacent neurovascular bundles were more likely to have recurrence after surgery. The

ABBREVIATIONS: EFS, event-free survival; HR, hazard ratio; IQR, interquartile range

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recurrence rate of desmoid tumors after surgery is high, ranging from 25% to 60% at 5 yr.^{4,7,18} To reduce the recurrence rate and simultaneously preserve neurovascular structures so that extremity function remains intact is challenging when surgically treating desmoid tumors.

In this study, we introduced sandwich isolation surgery, a surgical method to isolate residual tumor around neurovascular bundles from surrounding tissue, aiming to improve the surgical outcome of progressive desmoid tumor patients with R2 resection margin around neurovascular bundles involved. And we reported the effectiveness of the technique for preventing progressions and local recurrences, and its safety.

METHODS

Patients

The records of patients with progressive desmoid tumors who received surgery at the department of orthopedic oncology of our hospital between August 2014 and August 2018 were retrospectively reviewed. Inclusion criteria for the analysis were diagnosis of a desmoid tumor at extremities with adjacent nerves or vessels involved, and progressive enlargement or impairment of function after active surveillance or systemic therapy. Patients who were lost to follow-up or followed-up for less than 12 mo were excluded. A total of 27 patients who met criteria 1 and 2 were identified, and no patients were excluded (Figure 1). Among them, 13 patients received sandwich isolation surgery between August 2014 and August 2018, and 14 patients received traditional surgery between January 2015 and August 2018. Detailed data collections were described in **Text, Supplemental Digital Content**. The study was approved by the medical ethical committee of our hospital, and registered with ChiCTR (Registration number: ChiCTR-ORC-17011154). All patients provided written informed consent for surgical procedures performed.

Surgical Procedures

The steps of sandwich isolation surgery are summarized in Figure 2. Surgical procedures for both groups were detailed in **Text, Supplemental Digital Content**.

Postoperative Follow-up

Patients were seen in the outpatient clinic and received magnetic resonance imaging (MRI) of the surgical site every 4 mo. During follow-up, data regarding local recurrences and complications were collected.

Statistical Analyses

Categorical variables were presented as number and percentage, and continuous variables as median value and interquartile range (IQR). The chi-square test was employed to compare baseline characteristics between sandwich isolation group and traditional surgery group. Event-free survival (EFS) considered local recurrence, disease progression, and disease-related death. EFS was defined as the time interval from the date of surgery to the date of the occurring events or the date of the last follow-up for patients who had no events. EFS was calculated by the Kaplan-Meier method and examined by the log-rank test (time-to-event data). Univariate Cox regression model was used to investigate the effect of surgery type. A 2-sided value of $P < .05$ was considered to indicate statistical significance. Statistical analysis was performed with SPSS version 22.0 software (IBM Inc, Armonk, New York).

RESULTS

Clinical Characteristics of Patients in Sandwich Isolation Surgery Group

A total of 13 patients who received sandwich isolation surgery were included in the analysis (Table 1), including 7 male and 6 female patients. Their ages ranged from 2 to 59 yr (median: 23 yr). Nine patients (69%) had recurrent desmoid tumors and received at least 1 prior resection surgery before sandwich isolation surgery. The tumors were located at limb girdles (5 cases, 38%), proximal extremities (6 cases, 46%), and distal extremities (2 cases, 15%). Tumor size ranged from 1.8 cm to 24.1 cm. Two patients received chemotherapy: 1 (case 3 from Table 1) received 7 courses of pirarubicin (THP) combined with paclitaxel (PTX) before sandwich isolation surgery, and 4 courses of THP and PTX after surgery; the other (case 8 from Table 1) received 3 courses of THP and ifosfamide before sandwich isolation surgery. Additionally, 2 patients (case 5 and case 7 from Table 1) received postoperative radiotherapy and both received 60 Gy of after-loading radiotherapy.

According to preoperative MRI, the adjacent neurovascular bundles were partly involved in 8 patients (62%) and completely surrounded by tumors in 5 patients (38%). Intraoperative findings matched preoperative MRI findings. During sandwich isolation surgery, R2 resection was performed in the high-risk areas (neurovasculature-involving regions) of 13 patients and then the residual tumors on the neurovascular bundles were isolated by the biomaterial patch. In case 2 from Table 1, the residual tumors on the sciatic nerve were not fully isolated because the biomaterial patch could not enclose the bifurcation of the tibial and common peroneal nerves (Figure 3). In the low-risk areas (non-neurovasculature-involving regions), R0 resection was achieved in 11 patients (85%). For the other 2 patients (15%) who did not receive R0 resection, 1 patient (case 12 from Table 1) received R1 resection because the postoperative pathological report indicated a positive resection margin of the non-neurovasculature-involving region, and the other patient (case 13 from Table 1) received R2 resection because of the involvement of bone tissue.

Follow-up Information of Patients in Sandwich Isolation Surgery Group

The 13 patients were followed up for a median of 31.3 mo (IQR: 17.8-50.1 mo). Tumor progression occurred in 1 patient (case 2 from Table 1) and local recurrences occurred in 2 patients (cases 12 and 13 from Table 1). As for these 3 patients, tumor progressions and local recurrences were found by MRI at 11.5, 9.6, and 9.5 mo after sandwich isolation surgery, respectively, and were located outside the neurovasculature-involving regions isolated by biomaterial patch. However, during the whole follow-up period (range: 15.2-62.4 mo), MRI of the 3 patients showed that no tumor progressions or local recurrences were found in the isolated neurovasculature-involving regions, where R2 resection was performed. In summary, no progressions or local recurrences occurred in any of the 13 patients in the isolated

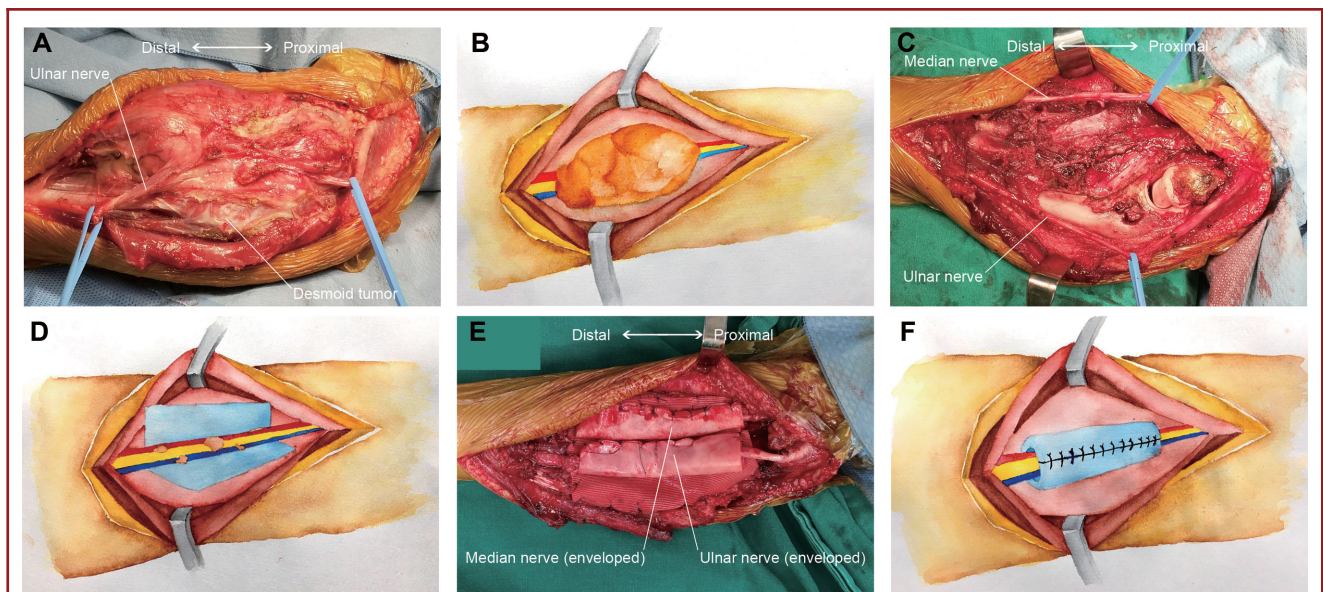
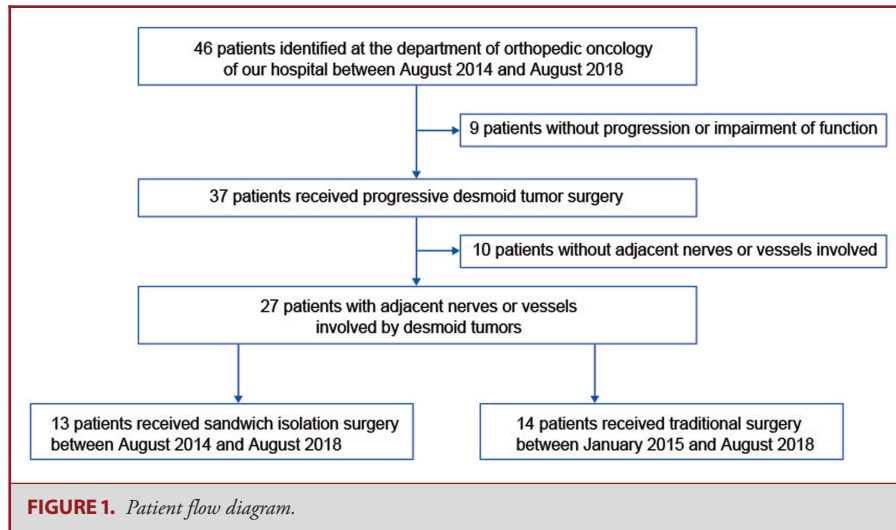


FIGURE 2. Images illustrating sandwich isolation surgery. **A** and **B**, a desmoid tumor invading adjacent nerves and vessels. **C** and **D**, resect the tumor as completely as possible by the wide resection of peripheral muscles in non-neurovasculature-involving regions (R0 resection) and perform R2 resection in neurovasculature-involving regions to preserve adjacent blood vessels and nerves and then envelop involved neurovascular structures with a polyester cardiovascular patch. **E** and **F**, polyester cardiovascular patch is sutured in place to isolate the involved neurovascular bundle from normal tissue.

neurovasculature-involving regions during follow-up. There were no amputations or deaths during follow-up.

Comparison of Sandwich Isolation Surgery and Traditional Surgery

A total of 14 patients who received traditional surgery during the same period were followed-up for a median of 36.3 mo (IQR: 27.1-50.1 mo), and local recurrence occurred in 10 of 14 patients (71.4%). No statistically significant differences were found in age, sex, primary vs recurrent disease, limb localization, tumor size, the percentage of patients with nerves or

vessels surrounded or partly involved by desmoid tumor, the percentage that received chemotherapy or radiotherapy, margin status of neurovasculature-involving region, and margin status of non-neurovasculature-involving region between the 2 groups (Table 2). The estimated 3-yr EFS rate was 76.9% (95% CI: 54.0-99.8%) for patients who received sandwich isolation surgery, and 32.7% (95% CI: 7.0-58.4%) for patients who received traditional surgery ($P = .025$, Figure 4). Patients who received sandwich isolation surgery were less likely to have local recurrence (hazard ratio [HR]: 0.257; 95% CI: 0.070-0.940, $P = .040$).

TABLE 1. Clinical Characteristics and Outcomes of the Patients Who Received Sandwich Isolation Surgery

Case	Age (year)	Sex	Disease	Number of previous surgeries	Time between previous surgeries and recurrences (months)	Tumor site	Involved nerves and vessels, and relation with tumor	Radiotherapy or chemotherapy before surgery	Radiotherapy or chemotherapy after surgery	Follow-up (months)	Progression or local recurrence after sandwich isolation surgery
1	10	Female	Recurrent	2	29, 6	Right thigh	Sciatic nerve, surrounded	None	None	63.6	No
2	23	Female	Recurrent	1	5	Left thigh	Sciatic nerve, surrounded	None	None	62.4	Progression, 11.5 mo after surgery
3	9	Male	Recurrent	3	6, 12, 29	Left upper arm	Axillary vessels and nerves, partly involved	Chemotherapy times 7	Chemotherapy times 4	48.5	No
4	17	Male	Recurrent	1	6	Left gluteal region	Sciatic nerve, partly involved	None	None	51.6	No
5	27	Male	Recurrent	1	9	Right forearm	Interosseous vessels and nerves, ulnar vessels, and nerves, partly involved	None	Radiotherapy	32.2	No
6	51	Male	Primary	0	None	Right shoulder	Axillary vessels and nerves, partly involved	None	None	31.3	No
7	22	Female	Primary	0	None	Right shoulder	Axillary vessels and nerves, partly involved	None	Radiotherapy	37.4	No
8	2	Male	Recurrent	1	2	Right upper arm	Axillary vessels and nerves, partly involved	Chemotherapy times 3	None	24.8	No
9	39	Female	Recurrent	1	24	Right gluteal region	Sciatic nerve, surrounded	None	None	22.5	No
10	55	Male	Primary	0	None	Right gluteal region	Sciatic nerve, partly involved	None	None	16.7	No
11	59	Male	Primary	0	None	Left leg	Common peroneal nerve, surrounded	None	None	19.0	No
12	16	Female	Recurrent	1	12	Left thigh	Sciatic nerve, surrounded	None	None	15.8	Local recurrence, 9.6 mo after surgery
13	47	Female	Recurrent	1	24	Left thigh	Popliteal vessels and sciatic nerve, partly involved	None	None	15.2	Local recurrence, 9.5 mo after surgery

Complications of Sandwich Isolation Surgery

During follow-up, 2 patients (cases 6 and 9 from Table 1) receiving sandwich isolation surgery reported intermittent mild pain in the operative regions. No analgesics were needed, and their life quality was not affected. No numbness or limb paralysis, or disturbances of blood circulation were observed during follow-up in any of the 13 patients who received sandwich isolation surgery. Additionally, no mesh-related complications were reported, including mesh infection, local fibrosis, and mesh migration.

DISCUSSION

For progressive desmoid tumors of the extremities, treatment options include radiotherapy, isolated limb perfusion, systemic therapy, and surgery. Radiotherapy is one of the primary options for inoperable tumors, but its role in the local control of desmoid tumors remains controversial because of late adverse effects

and second malignant neoplasms.^{1,4} Isolated limb perfusion is considered as an effective alternative to radiation. Disease control was seen in 90% of the 25 patients who received isolated limb perfusion treatment, but 40% developed disease progression and procedure-related toxicity led to substantial side effects which necessitated surgical interventions.¹⁹ As for systemic chemotherapy, there is no standard agent because of the lack of high-quality clinical evidence with respect to effectiveness.^{4,12,20} Surgery can provide rapid relief of symptoms but the local recurrence rate after surgery is high. It requires more effective treatment modalities to prevent local recurrences of progressive desmoid tumors.

Challenges of Surgical Removal of Desmoid Tumors

Desmoid tumors, also known as deep fibromatosis, often originate from deep tissues with greater hardness than many other soft tissue tumors. Although desmoid tumors seldom metastasize, deep-infiltrating growth pattern and the lack of

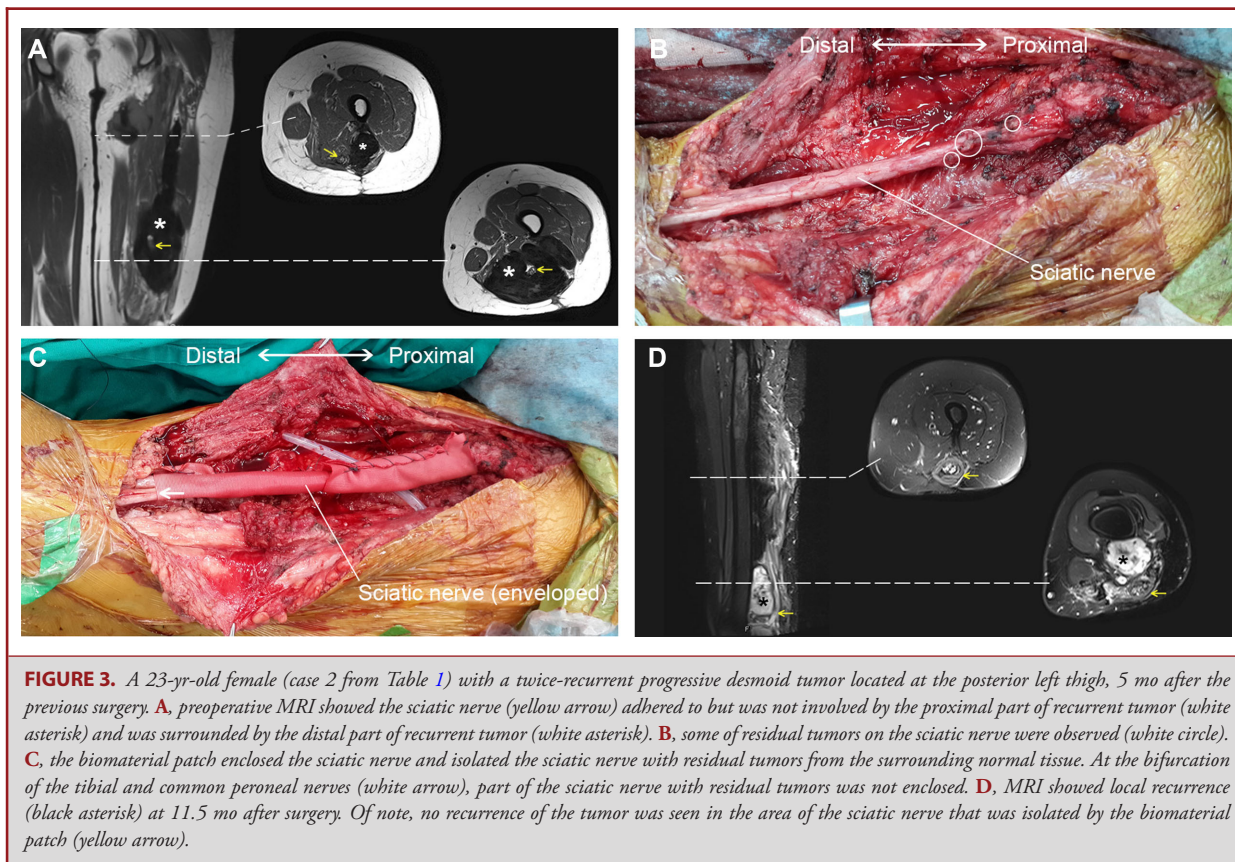


FIGURE 3. A 23-yr-old female (case 2 from Table 1) with a twice-recurrent progressive desmoid tumor located at the posterior left thigh, 5 mo after the previous surgery. **A**, preoperative MRI showed the sciatic nerve (yellow arrow) adhered to but was not involved by the proximal part of recurrent tumor (white asterisk) and was surrounded by the distal part of recurrent tumor (white asterisk). **B**, some of residual tumors on the sciatic nerve were observed (white circle). **C**, the biomaterial patch enclosed the sciatic nerve and isolated the sciatic nerve with residual tumors from the surrounding normal tissue. At the bifurcation of the tibial and common peroneal nerves (white arrow), part of the sciatic nerve with residual tumors was not enclosed. **D**, MRI showed local recurrence (black asterisk) at 11.5 mo after surgery. Of note, no recurrence of the tumor was seen in the area of the sciatic nerve that was isolated by the biomaterial patch (yellow arrow).

pseudocapsule make the boundary between desmoid tumors and normal tissues indistinguishable,²¹ which poses a challenge to the surgical removal of desmoid tumors. When nerves, vessels, or vital organs are involved, wide excision and en bloc resection are usually not acceptable. Even if microsurgical techniques are adopted, adjacent nerves and vessels are prone to injury. In order to avoid functional sacrifice, it is difficult to achieve R0 resection around neurovascular bundles.

The residual tumor on neurovascular bundles may be the source of local recurrence. It has been reported that the recurrence rate for patients with tumors invading adjacent neurovascular bundles is higher than that for those without invasion,¹⁷ and that recurrences most commonly occur between 14 and 17 mo after surgery.²² Extremity tumors have a higher local recurrence rate than abdominal wall tumors,¹⁶ which indirectly supports the correlation between neurovascular involvement and local recurrence. The involved nerve trunks and great vessels of the extremities are preserved during surgery, such that function remains intact, but the residual tumor on neurovascular bundles may be the source of local recurrence. On the other hand, nerves and vessels in the abdominal wall are less important, and thus are typically resected to avoid residual tumor if they are involved. Therefore, neurovasculature-involving regions are the high-risk

areas for functional impairments and local recurrences, and non-neurovasculature-involving regions are low-risk areas. Different strategies should be adopted in the two regions. R0 resection could be obtained in non-neurovasculature-involving regions by wide resection. But in neurovasculature-involving regions, it is important to preserve involved nerves or vessels and minimize the risk of local recurrence from residual tumors.

Theoretical Basis of Sandwich Isolation Surgery

According to the concept of curative margin, intrinsic barriers such as epineurium, vascular sheaths, cartilage, pleura, and peritoneum provide resistance against tumor invasion.²³ Even though it is common for desmoid tumors involving in adjacent nerves or vessels, we observed that desmoid tumors seldom penetrate into neurovascular bundles, suggesting that the neural sheath and vascular sheath act as intrinsic barriers against tumor invasion.

Based on aforementioned theories and findings, we developed a surgical method called sandwich isolation surgery to treat progressive desmoid tumor at extremities with adjacent nerves or vessels involved. A biomaterial nonabsorbable patch was used to envelop involved blood vessels and nerves after R2 resection of the primary tumor (Figure 2). The patch acts as an artificial barrier

TABLE 2. Characteristics of Patients Who Received Sandwich Isolation and Traditional Surgery

	Sandwich isolation surgery	Traditional surgery	χ^2	P
Age (year)				
<25	7 (54)	11 (61)	1.854	.236
≥25	6 (46)	3 (39)		
Sex				
Male	7 (54)	3 (21)	3.038	.120
Female	6 (46)	11 (79)		
Disease				
Primary	4 (31)	3 (21)	0.306	.678
Recurrent	9 (69)	11 (79)		
Limb localization				
Girdle	5 (39)	6 (43)	0.345	1.000
Proximal	6 (46)	5 (36)		
Distal	2 (15)	3 (21)		
Tumor size				
≤10 cm	7 (54)	5 (36)	0.898	.449
>10 cm	6 (46)	9 (64)		
Nerves or vessels involved				
Surrounded	5 (38)	4 (29)	0.297	.695
Partly involved	8 (62)	10 (71)		
Received chemotherapy				
Yes	2 (15)	1 (7)	0.464	.596
No	11 (85)	13 (93)		
Received radiotherapy				
Yes	2 (15)	3 (21)	0.163	1.000
No	11 (85)	11 (79)		
Neurovasculature margin				
R0	0 (0)	2 (14)	3.602	.098
R1	0 (0)	2 (14)		
R2	13 (100)	10 (71)		
Non-neurovasculature margin				
R0	11 (85)	10 (71)	1.194	.789
R1	1 (8)	3 (21)		
R2	1 (8)	1 (7)		

Data are presented as number (percentage).

between normal tissues and the residual tumor or tumor bed, and the neurovascular sheath functions as an intrinsic barrier.²³ Thus a “biomaterial patch-tumor/tumor bed-neurovascular sheath” sandwich structure is formed that has the potential of preventing the infiltration and progression of desmoid tumors.

Effectiveness of Sandwich Isolation Surgery

Of the 13 patients receiving sandwich isolation surgery, 9 patients had recurrent tumors and received at least 1 prior resection surgery before sandwich isolation surgery. Because of neurovascular involvement, the 13 patients were at a high risk of local recurrence.¹⁷ However, only 3 patients (23.1%) had tumor progressions and local recurrences after sandwich isolation surgery during follow-up. MRI of the 3 patients showed that tumor progressions and local recurrences were located outside the isolated neurovasculature-involving regions and that no tumor progressions or local recurrences were found within the sandwich

sleeves (the isolated neurovasculature-involving regions) during follow-up. Residual tumors outside the biomaterial patch may be the source of progressions and recurrences in the 3 patients (Table 3). The 3 cases substantiated the necessity of R0 resection in non-neurovasculature-involving regions and adequate isolation of neurovasculature-involving regions.

To further examine the effectiveness of sandwich isolation surgery, the local recurrence rate was compared between patients who received sandwich isolation surgery and those who received traditional surgery. The 2 groups had similar baseline clinical characteristics (Table 2), and their surgeries were performed in the same department during similar period, so they were considered comparable. The estimated 3-yr EFS rate of the patients who received sandwich isolation surgery was 76.9%, which was higher than that of the patients who received traditional surgery ($P = .025$, Figure 4), with a median duration of follow-up of approximately 34 mo (IQR: 22.5–48.8 mo). This follow-up duration covers the period during which most local

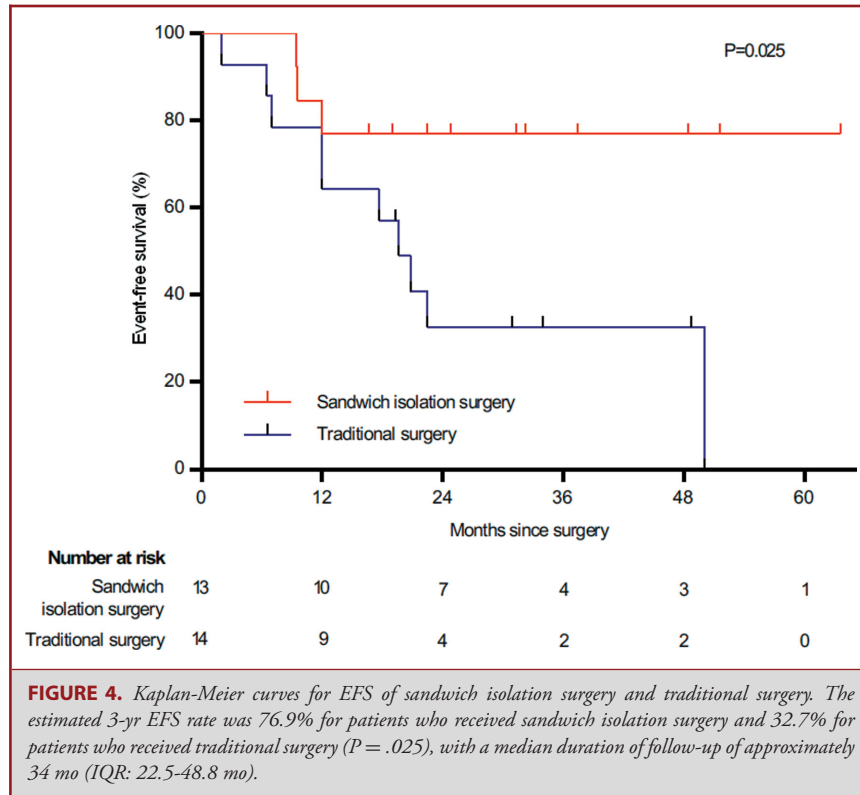


TABLE 3. Analysis of Progression and Local Recurrence after Sandwich Isolation Surgery

Case ^a	Margin status of neurovasculature-involving region	Margin status of non-neurovasculature-involving region	Location of progression or local recurrence	Progression or local recurrence in isolated neurovasculature-involving region	EFS (months)
2	R2, not fully isolated	R0	Non-isolated neurovasculature-involving region	None	11.5
12	R2	R1	Non-neurovasculature involving region	None	9.6
13	R2	R2	Non-neurovasculature involving region	None	9.5

^aCase numbers in concordance with Table 1.

recurrences occur.²² Sandwich isolation surgery was found to be a statistically significant protective factor against recurrence (HR: 0.257, $P = .040$).

The reduction of local recurrence rate suggested that sandwich isolation surgery can be an effective treatment option for patients with progressive extremity desmoid tumor and neurovascular involvement in which R0 resection cannot be achieved. It makes surgery possible for them because the involved nerves and vessels are preserved during tumor resection and surgical outcome of

R2 resection seems similar to that of R0 resection. Furthermore, for recurrent patients who received sandwich isolation surgery, subsequent surgery is less likely to result in important functional sacrifice, because biomaterial patch also isolates the neurovascular bundles from the recurrent tumors. Given the 3 cases with tumor progressions and local recurrences, sandwich isolation surgery is applicable when R0 resection can be obtained in non-neurovasculature-involving regions. If not, subsequent radiotherapy or systemic therapy should be considered. Only 2 patients

reported intermittent mild pain in the operative regions and no other complications were noted during follow-up, which suggested the safety of sandwich isolation surgery.

Limitations

There are some limitations to this study that should be considered. It is a retrospective study with all the limitations thereof. Additionally, the number of patients was relatively small because of the rare nature of desmoid tumors, limiting the power of statistical analysis. The determination of the exact mechanism requires future pathological studies. A multicenter study of the technique with a larger number of patients is needed to confirm the results.

CONCLUSION

Sandwich isolation surgery using an artificial barrier to isolate residual tumor reduces the progression and local recurrence rate of progressive desmoid tumors involving neurovascular bundles, especially when R0 resection can be obtained in non-neurovasculature-involving regions. The technique is simple to perform, and is not associated with any severe complications or functional impairments. By reducing the recurrence rate, the potential of functional impairment caused by repeated operations is avoided.

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Disclosures

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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Supplemental Digital Content. Text. Methods: details of data collections and surgical procedures.