

# Progressive Pelvic and Gluteal Desmoid Tumor in a 16-Year-Old Female: A Five-Year Multimodal Imaging Follow-Up

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## Abstract

Desmoid tumors (desmoid-type fibromatosis) are rare, locally aggressive fibroblastic neoplasms that lack metastatic potential but grow infiltratively and follow an unpredictable course; pelvic tumors in adolescents are especially uncommon and difficult to manage.

We report the case of a 16-year-old adolescent female who presented with right gluteal swelling in 2021; biopsy confirmed desmoid-type fibromatosis (Ki-67 index, 5%-10%; APC-negative). Serial MRI, fluorine-18 fluorodeoxyglucose PET/CT (<sup>18</sup>F-FDG PET/CT), and CT performed from 2024 to 2026 documented progressive enlargement of a right gluteal mass and a large presacral pelvic mass despite sequential treatment with imatinib, sorafenib, and off-label nirogacestat. Mass effect caused bilateral hydronephrosis and hematocolpos, which were managed with percutaneous nephrostomies, image-guided drainage, and bladder catheterization; the most recent imaging showed stable disease on nirogacestat.

This case illustrates the refractory behavior of adolescent pelvic desmoid tumors and the central role of longitudinal multimodal imaging in tracking progression and detecting complications amenable to interventional management. It adds to the limited experience with off-label nirogacestat in adolescents, for whom prospective pediatric data remain limited.

**Categories:** Radiology, Oncology, Pediatrics

**Keywords:** adolescent, aggressive fibromatosis, case report, desmoid tumor, magnetic resonance imaging, nirogacestat, pelvic mass, pet/ct

## Introduction

Desmoid tumors, also known as desmoid-type fibromatosis or aggressive fibromatosis, are rare clonal fibroblastic proliferations arising from deep soft tissues. Although they lack metastatic potential, they exhibit infiltrative growth, a high local recurrence rate, and a clinical course that is notoriously difficult to predict [1,2]. The estimated incidence is three to five cases per million per year, with a peak between the second and fourth decades of life and a female predominance [2].

Most sporadic desmoid tumors harbor activating mutations of the catenin beta 1 (*CTNNB1*) gene, while a subset, particularly intra-abdominal lesions, occurs in the context of familial adenomatous polyposis (FAP) and is associated with germline adenomatous polyposis coli (*APC*) mutations [1,2]. Pelvic and gluteal desmoid tumors are anatomically challenging owing to their proximity to neurovascular structures and pelvic viscera; mass effect can produce urinary obstruction, GI compression, neurologic deficits, and substantial pain.

Imaging is central to the diagnosis, characterization, and longitudinal surveillance of desmoid tumors. MRI is the modality of choice owing to its superior soft-tissue contrast and ability to assess extent and adjacent involvement; CT is valuable for the detection of complications such as obstruction or hemorrhage; and fluorine-18 fluorodeoxyglucose PET/CT (<sup>18</sup>F-FDG PET/CT) can complement morphologic assessment in selected cases [3]. Current international consensus advocates active surveillance as first-line management for asymptomatic and slowly progressive disease, reserving systemic therapy or local interventions for progressive, morbid, or symptomatic lesions [1]. The therapeutic armamentarium has recently expanded with the approval of nirogacestat, a gamma-secretase inhibitor, for adults with progressing desmoid tumors [4,5].

We report a 16-year-old female with a progressive pelvic and gluteal desmoid tumor followed for five years through serial MRI, PET/CT, and CT, a rarely documented duration of multimodal follow-up, refractory to multiple lines of systemic therapy and requiring interventional management of obstructive complications.

### How to cite this article

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## Case Presentation

A previously healthy adolescent girl first noticed painless right gluteal swelling in 2021, at the age of 11 years. The lesion was initially evaluated at another institution, where she was followed conservatively. Over the subsequent years, the swelling progressively enlarged, and she developed intermittent lower abdominal discomfort. She was referred to our center for further evaluation in 2024. Personal and family histories were unremarkable. There was no history of abdominal surgery, trauma, or features suggestive of FAP.

Image-guided core-needle biopsy of the right gluteal lesion confirmed desmoid-type fibromatosis with a Ki-67 proliferation index of 5%-10% (Figure 1). Genetic screening was negative for pathogenic *APC* mutations, supporting a sporadic presentation. Routine laboratory investigations were within normal limits.

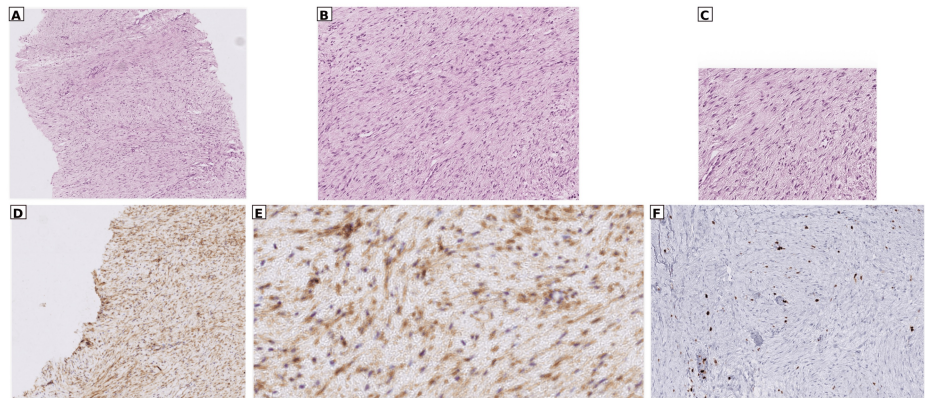


**FIGURE 1: Histopathology and immunohistochemistry of the right gluteal core-needle biopsy, October 2024.**

(A) HES,  $\times 5$ : overview of the biopsy core showing a mildly cellular spindle-cell proliferation within an abundant collagenous stroma, with the characteristic architecture of desmoid-type fibromatosis. (B) HES,  $\times 10$ : sweeping fascicles of bland fibroblastic spindle cells embedded in a dense collagenous background; no significant atypia or mitotic activity is identified. (C) HES,  $\times 20$ : cellular detail confirming the absence of cytologic atypia and mitoses. (D) Beta-catenin immunohistochemistry,  $\times 5$ : overview of the biopsy core showing beta-catenin labeling in spindle cells. (E) Beta-catenin,  $\times 20$ : brown beta-catenin immunolabeling in spindle cells with hematoxylin counterstain, supporting the diagnosis of desmoid-type fibromatosis in the appropriate histologic context. (F) Ki-67,  $\times 10$ : rare Ki-67-positive nuclei (brown) dispersed among a majority of negative cells, corresponding to a low proliferation index of 5%-10%.

HES: Hematoxylin-eosin-saffron.

## Therapeutic course

Given documented radiologic progression at presentation, watch-and-wait management was deemed inappropriate. Systemic therapy was initiated and adjusted sequentially in a multidisciplinary setting. Imatinib (Glivec) was started in December 2024. Sorafenib (Nexavar) was introduced in September 2025 after documented progression on imatinib. Nirogacestat (Ogsiveo) was initiated off-label in January 2026 following further progression on sorafenib, with multidisciplinary tumor board approval and informed parental consent. At the most recent follow-up, the patient remained on nirogacestat with stable disease on imaging. A chronological summary of the patient's imaging findings, therapies, and key clinical milestones is provided in Table 1.

| Time point            | Imaging/clinical findings                                                                                                                                                     | Management                                                                            |
|-----------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------|
| 2021 (age 11)         | Painless right gluteal swelling noted                                                                                                                                         | Conservative follow-up at an outside institution                                      |
| 2024                  | Referral; core-needle biopsy confirmed desmoid-type fibromatosis (Ki-67, 5%-10%; negative for pathogenic APC mutations)                                                       | -                                                                                     |
| October 2024          | Baseline pelvic MRI: right gluteal mass (~55 × 39 × 33 mm) and presacral pelvic mass (~95 × 87 × 66 mm), displacing the bladder, uterus, and rectum                           | -                                                                                     |
| November 2024         | <sup>18</sup> F-FDG PET/CT: moderate uptake; no distant FDG-avid disease                                                                                                      | -                                                                                     |
| December 2024         | -                                                                                                                                                                             | Imatinib initiated                                                                    |
| June 2025             | Follow-up MRI: marked progression, with pelvic mass measuring ~130 × 95 mm                                                                                                    | -                                                                                     |
| September 2025        | -                                                                                                                                                                             | Sorafenib initiated after progression on imatinib                                     |
| January 2026          | -                                                                                                                                                                             | Nirogacestat initiated off-label after progression on sorafenib                       |
| February 2026         | Contrast-enhanced CT: masses stable; new hematoocolpos, left pelvic fluid collection, bilateral hydroureteronephrosis secondary to ureteric compression; no bowel obstruction | Bilateral percutaneous nephrostomies; hematoocolpos drainage; bladder catheterization |
| Most recent follow-up | Stable disease                                                                                                                                                                | Ongoing nirogacestat; clinical and imaging surveillance                               |

TABLE 1: Timeline of imaging findings, therapies, and clinical milestones.

<sup>18</sup>F-FDG PET/CT: Fluorine-18 fluorodeoxyglucose PET/CT; APC: Adenomatous polyposis coli.

Imaging findings and longitudinal evolution

Baseline pelvic MRI performed in October 2024 identified a lobulated soft-tissue mass measuring approximately 55 × 39 × 33 mm in the right gluteal region. A larger presacral pelvic mass measuring approximately 95 × 87 × 66 mm was also seen, displacing the bladder, uterus, and rectum without overt invasion. Both lesions showed intermediate signal intensity on T1-weighted images and heterogeneous, predominantly intermediate-to-high signal intensity on T2-weighted and proton density fat-suppressed (Dixon) sequences, with internal hypointense bands consistent with the dense fibrocollagenous matrix typical of desmoid tumors (Figure 2).

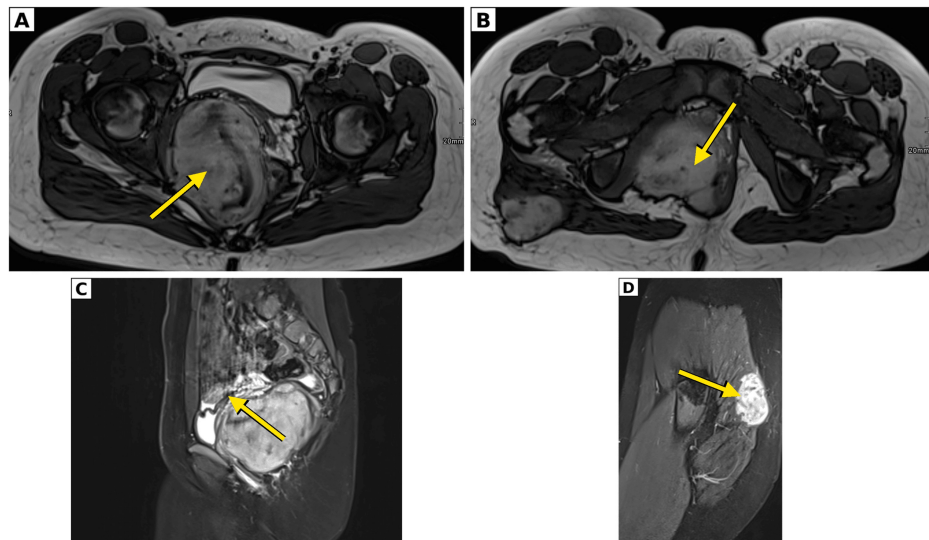


**FIGURE 2: Baseline MRI of the pelvis, October 2024.**

(A, B) Axial proton density-weighted TSE Dixon opposed-phase images showing the presacral pelvic mass and right gluteal lesion, both demonstrating intermediate signal intensity with heterogeneous internal signal and hypointense fibrocollagenous bands. (C) Sagittal proton density-weighted TSE Dixon water-only fat-suppressed image showing the pelvic mass displacing the bladder, uterus, and rectum. (D) Sagittal proton density-weighted TSE Dixon water-only fat-suppressed image of the right gluteal lesion.

PD: Proton density; TSE: Turbo spin-echo.

$^{18}\text{F}$ -FDG PET/CT performed in November 2024 showed moderate metabolic activity in both the gluteal and pelvic lesions. No regional or distant FDG-avid disease was identified. The findings were consistent with metabolically active fibromatosis; uptake in desmoid tumors is variable and may overlap with that of malignant soft-tissue lesions, depending on cellularity and collagen content (Figure 3).

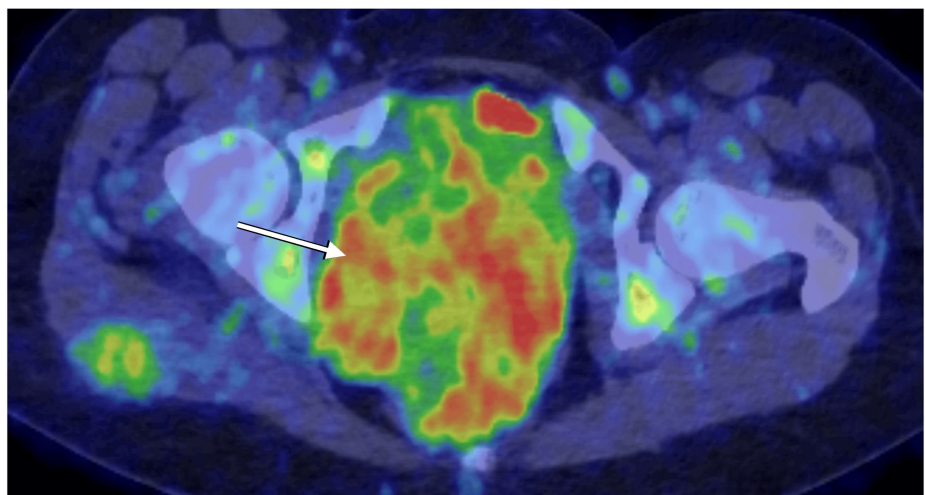


**FIGURE 3: Initial  $^{18}\text{F}$ -FDG PET/CT, November 2024.**

Axial fused PET/CT image demonstrating moderate FDG uptake within the pelvic mass, consistent with metabolically active desmoid-type fibromatosis. No distant FDG-avid disease was identified.

$^{18}\text{F}$ -FDG: Fluorine-18 fluorodeoxyglucose.

Follow-up pelvic MRI performed in June 2025 demonstrated marked interval enlargement of both lesions, with the pelvic component reaching approximately 130 × 95 mm and the gluteal lesion also progressing in size. T2 signal remained heterogeneous, with intermediate-to-high intensity relative to muscle and persistent hypointense fibrotic bands, in keeping with aggressive but biologically active fibromatosis (Figure

4).

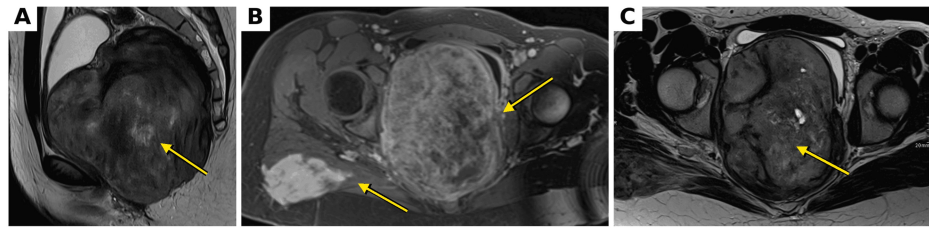


**FIGURE 4: Follow-up pelvic MRI, June 2025.**

(A) Sagittal T2-weighted TSE image demonstrating the enlarged presacral pelvic mass with mass effect and displacement of the bladder, uterus, and rectum (arrows). (B) Axial T1-weighted volumetric interpolated breath-hold examination (VIBE) image with fat saturation showing marked interval enlargement of the pelvic mass and persistence of the right gluteal lesion (arrows). (C) Axial T2-weighted TSE image showing heterogeneous intermediate-to-high T2 signal intensity with internal hypointense fibrotic bands, typical of a desmoid tumor (arrow).

VIBE: Volumetric interpolated breath-hold examination; TSE: Turbo spin-echo.

Contrast-enhanced abdominopelvic CT performed in February 2026 showed stable size of the dominant masses compared with the June 2025 MRI. However, new complications had developed: a large intravaginal hematoma (hematocolpos), a left pelvic fluid collection, and bilateral hydroureteronephrosis secondary to extrinsic ureteric compression by the pelvic mass. There was no evidence of bowel obstruction (Figure 5).



**FIGURE 5: Contrast-enhanced CT of the abdomen and pelvis, portal venous phase, February 2026.**

(A) Axial image showing the bulky pelvic mass with mass effect on adjacent pelvic viscera. (B) Coronal image demonstrating bilateral hydroureteronephrosis secondary to extrinsic ureteric compression by the pelvic desmoid tumor.

## Procedural interventions

Owing to obstructive complications and symptomatic hematocolpos, three procedures were performed: bilateral percutaneous nephrostomies for relief of hydroureteronephrosis, image-guided drainage of the hematocolpos, and indwelling urinary bladder catheterization for ongoing urinary management. The patient tolerated the procedures well, with prompt resolution of obstructive uropathy and improvement of pelvic symptoms. She continues on nirogacestat under close clinical and radiologic surveillance.

## Discussion

Desmoid tumors are rare, locally aggressive neoplasms whose biological behavior ranges from spontaneous stabilization or regression to relentless local progression [1,2]. The pelvic and gluteal localization observed in our patient is uncommon in adolescents and is associated with substantial morbidity owing to mass effect on the urinary tract, genital tract, and rectum. This case illustrates several teaching points relevant to radiologists and clinicians involved in the care of these patients.

## Role of imaging in desmoid tumor assessment

MRI remains the cornerstone of desmoid tumor assessment. Lesions typically appear as infiltrative soft-tissue masses with intermediate signal intensity on T1-weighted images and variable, often heterogeneous, signal intensity on T2-weighted sequences, with hypointense bands corresponding to dense collagenous stroma [3]. A reduction in T2 signal intensity over time is often interpreted as a marker of decreased cellularity and favorable response to therapy, although this correlation is imperfect. CT remains useful for the detection of secondary complications, particularly obstructive uropathy and intrapelvic collections, as illustrated in our patient. The role of <sup>18</sup>F-FDG PET/CT is more limited and less standardized; uptake is variable and may overlap with that of malignant soft-tissue lesions, but PET/CT can be helpful for whole-body staging in selected cases and for characterizing metabolically active lesions [3].

### **Toward an individualized, multidisciplinary strategy**

International consensus has shifted away from upfront surgery toward an individualized, multidisciplinary strategy [1]. Active surveillance with serial MRI every three to six months is now recommended as first-line management for asymptomatic and slowly progressive lesions, given the high local recurrence rate after resection and the possibility of spontaneous stabilization [1,6,7]. Active treatment is indicated for tumors that are symptomatic, morbid, or radiologically progressing. Available systemic options include nonsteroidal anti-inflammatory drugs, hormonal therapy, conventional cytotoxic chemotherapy, most often low-dose methotrexate-vinblastine in younger patients, tyrosine kinase inhibitors such as imatinib, sorafenib, and pazopanib, and, more recently, the gamma-secretase inhibitor nirogacestat [4,5,7,8].

### **Nirogacestat in adolescents: an evolving evidence base**

Nirogacestat received approval from the US FDA in November 2023 for adults with progressing desmoid tumors requiring systemic therapy, on the basis of the placebo-controlled DeFi trial, which demonstrated a significant progression-free survival benefit (hazard ratio for progression or death, 0.29; 95% CI, 0.15–0.55) and an objective response rate of 41% versus 8% with placebo [4,5]. The National Comprehensive Cancer Network (NCCN) guidelines list nirogacestat as a category 1 preferred systemic therapy in adults with desmoid tumors [5]. Importantly, nirogacestat is not currently approved in patients younger than 18 years; an ongoing Children's Oncology Group phase II trial (NCT04195399) is evaluating its safety, pharmacokinetics, and efficacy in children and adolescents with progressive, surgically unresectable disease [9]. In our case, nirogacestat was offered off-label after failure of two prior systemic lines, with multidisciplinary tumor board approval and informed parental consent. To date, the disease has remained stable on treatment. As desmoid tumors are typically slow-growing, the median progression-free survival in the DeFi placebo arm was approximately 15 months, the present follow-up remains short, and the observed stability should be regarded as a hypothesis-generating observation rather than evidence of efficacy in this age group. As pediatric data accumulate, the role of nirogacestat in adolescents with refractory desmoid tumors will need to be further defined.

### **Pelvic complications and the role of interventional radiology**

Pelvic desmoid tumors can produce significant obstructive complications, even when the tumor itself is biologically stable. In our patient, bilateral hydronephrosis and hematoocolpos developed despite stable tumor dimensions on the most recent CT, illustrating that mass effect, rather than tumor growth alone, can drive clinical deterioration. Image-guided interventions, including percutaneous nephrostomies and drainage of pelvic collections, played a key role in symptom control and preservation of renal function, in line with reports of pelvic and abdominal-wall desmoid tumors involving the urinary tract [10]. This experience reinforces the importance of an integrated radiology and interventional radiology pathway for these patients.

### **Comparison with the published literature**

Published reports on desmoid tumors emphasize both the heterogeneity of presentations and the difficulty of management. Series of pelvic and abdominal-wall lesions document substantial morbidity from mass effect on the urinary and genital tracts and a need for multidisciplinary input [10]. Reports of unusual locations, including the posterior cruciate ligament, the subscapularis muscle, the hand, and the neck, illustrate the wide anatomic spectrum of the disease and the need for tailored management plans [11–14]. Tyrosine kinase inhibitor therapy may be associated with lymphadenopathy that can mimic progression and must be recognized on follow-up imaging [15]. Recent specialist sarcoma-unit experience confirms that conservative, individualized strategies yield favorable long-term outcomes for many patients [16]. To our knowledge, this is one of few reports describing a five-year multimodal imaging follow-up of an adolescent pelvic-gluteal desmoid tumor treated sequentially with imatinib, sorafenib, and off-label nirogacestat, highlighting both the imaging evolution and the management of obstructive complications.

### **Limitations**

This is a single-patient case report, and the findings cannot be generalized. Quantitative response evaluation using volumetric or diffusion-weighted MRI metrics was not systematically performed across all time points. Long-term outcome data on nirogacestat in adolescents remain limited.

## Conclusions

Adolescent pelvic and gluteal desmoid tumors represent a diagnostic and therapeutic challenge. This case demonstrates the value of longitudinal multimodal imaging in characterizing tumor behavior, guiding the choice and sequencing of systemic therapies, and detecting complications that may require interventional management. It also illustrates the off-label use of nirogacestat in an adolescent patient with refractory disease, an area in which prospective pediatric data are still awaited. A multidisciplinary, patient-centered approach combining surveillance, systemic therapy, and image-guided interventions is essential to optimize outcomes in this rare and unpredictable disease.

## Additional Information

### Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

**Concept and design:** Tarek Ghamraoui, Nicolas Naccarella

**Acquisition, analysis, or interpretation of data:** Tarek Ghamraoui, Nicolas Naccarella

**Drafting of the manuscript:** Tarek Ghamraoui

**Critical review of the manuscript for important intellectual content:** Nicolas Naccarella

**Supervision:** Nicolas Naccarella

### Disclosures

**Human subjects:** Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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