Volume 6, No. 1 January, 2025
p ISSN 2723-6927-e ISSN 2723-4339

Desmoid Tumor Patients Profile in Dr. Soetomo Surabaya Hospital: A Rare Event

Beatta Meidini Rahmat1*, Edwin Danardono[2], Asdi Wihandono[3]
Airlangga University/RSUD Dr. Soetomo, 60286, Surabaya, Indonesia
Email: meidinibeatta@gmail.com

Abstract
Desmoid tumor (DT) is a fibroblast soft tissue tumor that deep seated connective tissue malignancy developing in musculoaponeurtic tissues. Desmoid tumor is a rare tumor, reported 5-6 cases per million population and account 0.03% of all neoplasm yet the etiopathogenesis of this tumor remains unclear. DT most commonly develops between the age of 15-60 years and common in females, although it rarely metastasized but locally aggressive and affect the adjacent structures. Knowing the profile of desmoid tumor patients at RSUD Dr. Soetomo Surabaya for the period January 2020 – December 2024. Including demographic patients, risk factors, tumor sites, and patients' managements. All patients between January 2020-December 2024 whose primary DTs were observed. Datas were obtained and observational descriptive study conducted from medical files to record patient's age, sex, tumor site, and therapeutic managements. All 16 patients were obtained during 5 years with female predominantly (1:4) and adult was more common 19-44 years. Risk factors including in this study were history of trauma, history on pregnancy, and hormonal contraceptive. Location of tumors were found mostly in abdominal wall and the therapy conducted were surgery followed by chemotherapy, and combination of hormonal therapy, TKIs, and radiotherapy. DT is a rare fibroblastic neoplasm that shows locally aggressive behavior with very low incidence annually with female, history of trauma, history of pregnancy, and hormonal contraceptive as a major risk factors. The therapeutic management includes multidisciplinary approach.
Keywords: Desmoid tumor, Risk factors, Therapeutic Management

Introduction
Desmoid tumor, or also known as desmoid fibromatosis, is a benign tumor of fibroblastic tissue that originates from visceral soft tissue and is aggressive but rarely metastasizes.(Mastoraki et al., 2022)This tumor has infiltrative growth characteristics and can affect adjacent organs, causing clinical symptoms that can worsening the patient's quality of life.(Antasouras et al., 2023) This causes morbidity for patients because it results in late diagnosis and because the tumor is so aggressive, it can affect quality of life (Alemany et al., 2021)

The incidence of desmoid tumors is 5-6 cases per million people per year with a peak age in the third decade and fourth decade and there are no studies that mention this tumor occurring at ages under 11 years (Mangla et al., 2024). In other studies, it is even stated that the incidence is very low, around 3-5 cases per million people per year, this is due to low awareness of this disease.(Soerjomataram & Bray, 2021) Desmoid tumors are dominated by women with a ratio of 2:1 to men.(Testa et al., 2022) It is known from various studies that desmoid tumors are associated with familial adenomatous polyposis (FAP) (Bektas et al., 2023) Patients with desmoid tumors generally have symptoms such as chronic pain (63%), sleep disturbances (73%), lethargy (46%), and anxiety to depression (15%). Signs that are often associated with desmoid tumors are pain, impaired function and mobility, fatigue, muscle weakness, and swelling around the tumor (Bektas et al., 2023). Desmoid tumors have a tendency to recur, rarely metastasize, and can grow in any part of the body, extra-abdominal including the head, neck, thorax, breast, extremities or intra-

abdominal, or on the abdominal wall.(Figueredo & Schiano, 2023) The incidence of intra-abdominal desmoid tumors is more common in women, especially during or after pregnancy, while those in the abdominal wall and mesentery are more common in patients with FAP (2) (3). (Zubor et al., 2024)Only one cohort study reported a 5-year mortality rate of 7 per 1,000 with a hazard ratio of death of 1.3. This death was mostly found in patients with FAP, even though this was only 7% of the study population (Mona et al., 2022)

It is suspected that the mechanism of desmoid tumors is through mutations in the Wnt/□-catenin (CTNNB1)/APC gene pathway and the Notch receptor pathway (Riedel & Agulnik, 2022) The potential for crosstalk between the Notch pathway and the Wnt pathway that causes Notch activation and Wnt signaling dysregulation is thought to be involved in the pathogenesis of desmoid tumors (Federman, 2022). The presence of mutations in CTNNB1 influences the risk of desmoid tumor recurrence after initial therapy. Several meta-analyses show that desmoid tumor patients with CTNNB1 mutations have a higher risk of recurrence after surgery and respond less well to meloxicam and imatinib (Spolverato et al., 2022)

Up until some time ago surgery was still considered the main choice that must be taken in dealing with desmoid tumors. Various therapeutic modalities that can be given include active surveillance, surgery, radiotherapy, locoregional therapy with cryoablation, high-intensity-focused ultrasound, radiofrequency ablation, and systemic therapy with chemotherapy. Handling desmoid tumors actually requires multidisciplinary management that can relieve the patient's symptoms in addition to providing therapy that is most appropriate to the patient's condition. The current paradigm has shifted from surgery as the primary therapy, to active surveillance for some patients (Liu et al., 2021)

In our center, treating desmoid tumors is still a challenge starting from establishing diagnostics which causes delays in starting therapy in patients and misdiagnosis.(Hendricks et al., 2021) This of course reduces the patient's quality of life. To date, surgery remains the primary modality applied and no data have been collected for this case.(Sun et al., 2021)

Research Methods

A retrospective observational study that describes the profile of patients with desmoid tumors at Dr. Soetomo General Hospital Surabaya for the period January 2020 – December 2024. Characteristic of the samples including demography, risk factors of desmoid tumor, tumor sites, and therapeutic management.

The study included patients whose initial desmoid tumor confirmed by histopathology and patients whose already been treated by surgery and/or systemic therapy in surgical ward. Data was conducted from medical record from January 2020 to December 2024. The exclusion criteria was incomplete medical records. A total of 16 patients met the inclusion criteria and then analyzed.

For the selected criteria, the following characteristics were recorded: gender, age, taking into considerations their location, tumor were divided into abdominal wall, intraabdominal, and extra abdominal. The data analysis and interpretation were performed using the SPSS version 26. The collected data is then classified into two groups, namely quantitative data presented in the form of frequencies and percentages while qualitative data is presented in words or symbols.

Results and Discussion

This study has 16 patients of desmoid tumor treated in Dr. Soetomo General Hospital Surabaya. From the total subjects, 1 patient had histopathological results dermatofibrosarcoma protuberans so it was excluded from this study. Thus, the total sample that met the research inclusion and exclusion criteria was 16 subjects. Sociodemographic data collected was in the form of gender and age group. Table 5.1 below describes the characteristics of sociodemographic data of patients with desmoid tumors in the surgical division

Table 1. The Gender and Age Distributions

	Frequency (N=16)	**Percentage (%)**
Gender		
Men	3	18,7%
Women	13	81,25%
Age		
Adolescence (10-18 years)	0	0,00%
Adult (19-44 years)	1	1,56%
Elderly (≥60 years)	5	7,81%

The study revealed that the proportion of women with desmoid tumors was predominat (81.2%) than men (18.7%). The youngest age obtained in this study was 17 years and the oldest age was 69 years. Based on age group, it was found that the adult group (19-44 years) was the largest age group with a percentage of 75%. To determine the characteristics of risk factors for this tumor, we collect data from medical records. The characteristics were history of previous trauma, history of pregnancy, and history of using oral hormonal contraceptives. History of trauma in the form of injuries due to collisions or other causes and a history of previous surgeries. The last two risk factors were only taken from female subjects.

Table 2. History of Trauma

	Frequency (N=16)	**Percentage (%)**	**Frequency (N=16)**	**Percentage (%)**
	Yes		**No**	
History of trauma	8	50%	8	50%

It is known that there was a history of trauma in 8 subjects (50%). The trauma consisted of 3 previous cesarean sections, 4 history of collisions and injuries, and 1 history of previous tumor excision surgery in the neck region. There were 8 subjects (50%) who had no history of trauma.

Table 3. Pregnancy and Hormonal Contraceptive

	Frequency (N=13)	Percentage (%)	Frequency (N=13)	Percentage (%)
	Yes		No	
Pregnancy	7	53,8%	6	46,1%
Hormonal contraceptive	8	61,5%	5	38,4%

Based on the table above, 8 subjects (61.5%) used oral hormonal contraception before being diagnosed with desmoid tumors and 5 subjects (38.4%) did not use hormonal contraception.

Table 4 shows tumor sites in this study, it was found that the abdominal wall was the most frequently found (31.2%), followed by intra-abdominal (25%), flank (18.7%), and posterior neck, hemithorax, brachii, and femur (6.2%).

Table 4. Location of Desmoid Tumor

Tumor Site	Frequency (N=16)	Percentage (%)
Posterior neck	1	6,2%
Flank	3	18,7%
Abdominal wall	5	31,2%
Intra abdominal	4	25%
Chest wall	1	6,2%
Brachii	1	6,2%
Femur	1	6,2%

Descriptive analysis was performed on all patients with desmoid tumors. The patients were treated by locoregional control i. e. surgery and radiotherapy as well as systemic therapy, hormonal therapy, TKIs, and/or chemotherapy.

Table 5. Location of Desmoid Tumor

Therapy	Frequency (N=16)	Percentage (%)
Surgery	10	62,5%
Chemotherapy	2	12,5%

Therapy	Frequency (N=16)	Percentage (%)
Chemotherapy + hormonal	1	6,2%
CHemotherapy + TKIs	1	6,2%
Surgery + hormonal	1	6,2%
Surgery + chemoradiation	1	6,2%

The most common treatment was only surgery (62.5%) followed by only chemotherapy (18.7%), namely intra-abdominal and thoracic wall desmoid tumors that was treated by regimen of vincristine, doxorubicin and cyclophosphamide. Then methotrexate vinorelbine chemotherapy with a combination of anti-hormonal tamoxifen (6.2%) for intra-abdominal desmoid tumors, followed by a combination of chemotherapy vincristine, doxorubicin, cyclophosphamide and imatinib (6.2%) and surgery followed by anti-hormonal tamoxifen (6.2%) and surgery followed by chemotherapy with a doxorubicin dacarbazine regimen and radiation in a case of intra-abdominal desmoid tumors (6.2%)

Conclusion

During the period January 2020–December 2024, 16 cases of desmoid tumors were recorded at Dr. Soetomo General Hospital Surabaya, with women being the most affected compared to men and adults being the most common age group. The study identified several risk factors frequently associated with desmoid tumors, including a history of previous trauma or surgery, pregnancy, and the use of oral hormonal contraceptives. The primary treatment modality in this study was surgery, which remained the main therapeutic approach for resectable tumors. However, for unresectable cases, treatment options included chemotherapy combined with hormonal therapy or radiation. Additionally, one patient received tyrosine kinase inhibitors (TKIs) followed by chemotherapy as part of their treatment strategy.

These findings emphasize the importance of early detection and individualized treatment strategies for desmoid tumors, particularly in patients with known risk factors. Given the limited number of cases and the variability in treatment responses, future research should focus on long-term outcomes of different treatment modalities, the role of targeted therapies such as TKIs, and the effectiveness of combination therapies in unresectable cases. Additionally, molecular and genetic studies could provide deeper insights into the pathogenesis of desmoid tumors, potentially leading to the development of novel therapeutic approaches.

Reference

Alemany, M., Velasco, R., Simó, M., & Bruna, J. (2021). Late effects of cancer treatment: consequences for long-term brain cancer survivors. *Neuro-Oncology Practice*, *8*(1), 18–30.

Antasouras, G., Papadopoulou, S. K., Tolia, M., Pandi, A.-L., Spanoudaki, M., Tsoukalas, N., Tsourouflis, G., Psara, E., Mentzelou, M., & Giaginis, C. (2023). May nutritional status positively affect disease progression and prognosis in patients with esophageal and pharyngeal cancers? A scoping review of the current clinical studies. *Medical Sciences*, *11*(4), 64.

Bektas, M., Bell, T., Khan, S., Tumminello, B., Fernandez, M. M., Heyes, C., & Oton, A. B. (2023). Desmoid tumors: a comprehensive review. *Advances in Therapy*, *40*(9), 3697–3722.

Federman, N. (2022). Molecular pathogenesis of desmoid tumor and the role of γ-secretase inhibition. *NPJ Precision Oncology*, *6*(1), 62.

Figueredo, C., & Schiano, T. (2023). A Review of the Clinical Presentation, Outcomes, and Treatments of Patients Having Desmoid Tumors. *Gastro Hep Advances*, *2*(4), 588–600.

Hendricks, A., Boerner, K., Germer, C.-T., & Wiegering, A. (2021). Desmoplastic small round cell tumors: a review with focus on clinical management and therapeutic options. *Cancer Treatment Reviews*, *93*, 102140.

Liu, J. L., Patel, H. D., Haney, N. M., Epstein, J. I., & Partin, A. W. (2021). Advances in the selection of patients with prostate cancer for active surveillance. *Nature Reviews Urology*, *18*(4), 197–208.

Mangla, A., Agarwal, N., & Schwartz, G. (2024). Desmoid tumors: current perspective and treatment. *Current Treatment Options in Oncology*, *25*(2), 161–175.

Mastoraki, A., Schizas, D., Vassiliu, S., Saliaris, K., Giagkos, G.-C., Theochari, M., Vergadis, C., Tolia, M., Vassiliu, P., & Felekouras, E. (2022). Evaluation of diagnostic algorithm and therapeutic interventions for intra-abdominal desmoid tumors. *Surgical Oncology*, *41*, 101724.

Mona, C. E., Benz, M. R., Hikmat, F., Grogan, T. R., Lueckerath, K., Razmaria, A., Riahi, R., Slavik, R., Girgis, M. D., & Carlucci, G. (2022). Correlation of 68Ga-FAPi-46 PET biodistribution with FAP expression by immunohistochemistry in patients with solid cancers: interim analysis of a prospective translational exploratory study. *Journal of Nuclear Medicine*, *63*(7), 1021–1026.

Riedel, R. F., & Agulnik, M. (2022). Evolving strategies for management of desmoid tumor. *Cancer*, *128*(16), 3027–3040.

Soerjomataram, I., & Bray, F. (2021). Planning for tomorrow: global cancer incidence and the role of prevention 2020–2070. *Nature Reviews Clinical Oncology*, *18*(10), 663–672.

Spolverato, G., Capelli, G., Kasper, B., & Gounder, M. (2022). Management of Desmoid tumors. *Surgical Oncology Clinics*, *31*(3), 447–458.

Sun, Y., Huang, Q., Cui, S., Wang, M., Zhang, N., Zhang, S., Yang, B., Qiu, E., Huang, Z., & Zhou, B. (2021). Outcomes and Quality-of-Life Measures after Endoscopic Endonasal Resection of Kadish Stage C Olfactory Neuroblastomas. *World Neurosurgery*, *151*, e58–e67.

Testa, S., Bui, N. Q., Charville, G. W., Avedian, R. S., Steffner, R., Ghanouni, P., Mohler, D. G., & Ganjoo, K. N. (2022). Management of patients with newly diagnosed desmoid tumors in a first-line setting. *Cancers*, *14*(16), 3907.

Zubor, P., Henriksen, C. M., Økstad, M. E., Cerskuviene, E., Visnovsky, J., Kajo, K., Valkov, A., & Lind, K. O. (2024). Desmoid Fibromatosis of the Anterior Abdominal Wall in Pregnancy: A Case Report and Review of the Literature. *Diseases*, *12*(1), 27.