

Editorial

Solitary Fibrous Tumor

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Solitary fibrous tumor (SFT) is an orphan disease of mesenchymal origin. The tumor can occur anywhere in the human body but is most frequently found in the chest. Surgery remains the standard of care in all SFTs. However, even completely resected disease can recur many years after surgery and its clinical behavior is unpredictable [1]. Genetic characterization using NAB2-STAT6 fusion has helped to better define and thus understand the disease for over a decade [2]. In addition, some risk stratification models have been developed and validated to estimate the clinical outcome [3].

Regarding therapy, surgery remains the cornerstone in SFT management but systemic treatment (including antiangiogenic therapy) as well as radiotherapy have been suggested to improve survival and quality of life, especially in advanced stages of the disease [4]. Despite the current advantages of non-surgical SFT therapy, there is still an urgent need to enhance our clinical and biological understanding of this rare malignancy.

The present Special Issue aims to improve—for patients as well as for physicians—the frustrating situation in which an established and effective therapy and follow-up strategies are still lacking, and focuses on providing an overview of the current standard of care by contributing three review articles.

In addition, seven research articles enhance the knowledge regarding risk stratification and novel treatment approaches. On the one hand, a better risk stratification is urgently needed to establish follow-up strategies to treat this orphan disease in a clinically meaningful manner. On the other hand, an effective therapy that could provide an alternative to surgery is of interest for non-resectable patients, as well as in the setting of multimodal therapy before and/or after surgery. We hope that this Special Issue will help to improve the outcome of this rare and challenging disease.



Citation: Ghanim, B. Solitary Fibrous Tumor. *Cancers* **2024**, *16*, 3573. <https://doi.org/10.3390/cancers16213573>

Received: 8 October 2024

Accepted: 15 October 2024

Published: 23 October 2024



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Funding: This Editorial was not supported by funding.

Acknowledgments: As Guest Editor of the Special Issue “Solitary fibrous tumor”, I want to thank all authors who spend their time and effort on exploring an orphan tumor to improve the situation of our patients and thus making this edition possible.

Conflicts of Interest: The author declares no conflicts of interest.

List of Contributions:

1. Bianchi, G.; Lana, D.; Gambarotti, M.; Ferrari, C.; Sbaraglia, M.; Pedrini, E.; Pazzaglia, L.; Sangiorgi, L.; Bartolotti, I.; Dei Tos, A.; et al. Clinical, Histological, and Molecular Features of Solitary Fibrous Tumor of Bone: A Single Institution Retrospective Review. *Cancers* **2021**, *13*, 2470. <https://doi.org/10.3390/cancers13102470>.
2. de Bernardi, A.; Dufresne, A.; Mishellany, F.; Blay, J.; Ray-Coquard, I.; Brahmi, M. Novel Therapeutic Options for Solitary Fibrous Tumor: Antiangiogenic Therapy and Beyond. *Cancers* **2022**, *14*, 1064. <https://doi.org/10.3390/cancers14041064>.
3. Apra, C.; El Arbi, A.; Montero, A.; Parker, F.; Knafo, S. Spinal Solitary Fibrous Tumors: An Original Multicenter Series and Systematic Review of Presentation, Management, and Prognosis. *Cancers* **2022**, *14*, 2839. <https://doi.org/10.3390/cancers14122839>.

4. Ghanim, B.; Baier, D.; Pirker, C.; Müllauer, L.; Sinn, K.; Lang, G.; Hoetzenecker, K.; Berger, W. Trabectedin Is Active against Two Novel, Patient-Derived Solitary Fibrous Pleural Tumor Cell Lines and Synergizes with Ponatinib. *Cancers* **2022**, *14*, 5602. <https://doi.org/10.3390/cancers14225602>.
5. Lottin, M.; Escande, A.; Bauchet, L.; Albert-Thananayagam, M.; Barthoulot, M.; Peyre, M.; Boone, M.; Zouaoui, S.; Guyotat, J.; Penchet, G.; et al. Intracranial Solitary Fibrous Tumour Management: A French Multicentre Retrospective Study. *Cancers* **2023**, *15*, 704. <https://doi.org/10.3390/cancers15030704>.
6. Kinslow, C.; Rae, A.; Kumar, P.; McKhann, G.; Sisti, M.; Bruce, J.; Yu, J.; Cheng, S.; Wang, T. Risk Stratification for Management of Solitary Fibrous Tumor/Hemangiopericytoma of the Central Nervous System. *Cancers* **2023**, *15*, 876. <https://doi.org/10.3390/cancers15030876>.
7. Hassani, M.; Jung, S.; Ghodsi, E.; Seddigh, L.; Kooner, P.; Aoude, A.; Turcotte, R. Value of Cellular Components and Focal Dedifferentiation to Predict the Risk of Metastasis in a Benign-Appearing Extra-Meningeal Solitary Fibrous Tumor: An Original Series from a Tertiary Sarcoma Center. *Cancers* **2023**, *15*, 1441. <https://doi.org/10.3390/cancers15051441>.
8. Li, Y.; Nguyen, J.; Ammanamanchi, M.; Zhou, Z.; Harbut, E.; Mondaza-Hernandez, J.; Meyer, C.; Moura, D.; Martin-Broto, J.; Hayenga, H.; et al. Reduction of Tumor Growth with RNA-Targeting Treatment of the NAB2-STAT6 Fusion Transcript in Solitary Fibrous Tumor Models. *Cancers* **2023**, *15*, 3127. <https://doi.org/10.3390/cancers15123127>.
9. Bertoglio, P.; Querzoli, G.; Kestenholz, P.; Scarci, M.; La Porta, M.; Solli, P.; Minervini, F. Surgery for Solitary Fibrous Tumors of the Pleura: A Review of the Available Evidence. *Cancers* **2023**, *15*, 4166. <https://doi.org/10.3390/cancers15164166>.
10. Piccinelli, M.; Law, K.; Incesu, R.; Tappero, S.; Cano Garcia, C.; Barletta, F.; Morra, S.; Scheipner, L.; Baudo, A.; Tian, Z.; et al. Demographic and Clinical Characteristics of Malignant Solitary Fibrous Tumors: A SEER Database Analysis. *Cancers* **2024**, *16*, 3331. <https://doi.org/10.3390/cancers16193331>.

References

1. Thway, K.; Ng, W.; Noujaim, J.; Jones, R.L.; Fisher, C. The Current Status of Solitary Fibrous Tumor: Diagnostic Features, Variants, and Genetics. *Int. J. Surg. Pathol.* **2016**, *24*, 281–292. [[CrossRef](#)] [[PubMed](#)]
2. Robinson, D.R.; Wu, Y.M.; Kalyana-Sundaram, S.; Cao, X.; Lonigro, R.J.; Sung, Y.S.; Chen, C.L.; Zhang, L.; Wang, R.; Su, F.; et al. Identification of recurrent NAB2-STAT6 gene fusions in solitary fibrous tumor by integrative sequencing. *Nat. Genet.* **2013**, *45*, 180–185. [[CrossRef](#)] [[PubMed](#)]
3. Demicco, E.G.; Park, M.S.; Araujo, D.M.; Fox, P.S.; Bassett, R.L.; Pollock, R.E.; Lazar, A.J.; Wang, W.L. Solitary fibrous tumor: A clinicopathological study of 110 cases and proposed risk assessment model. *Mod. Pathol.* **2012**, *25*, 1298–1306. [[CrossRef](#)] [[PubMed](#)]
4. Ren, C.; D’Amato, G.; Hornicek, F.J.; Tao, H.; Duan, Z. Advances in the Molecular Biology of the Solitary Fibrous Tumor and Potential Impact on Clinical Applications. *Cancer Metastasis Rev.* **2024**; *Online ahead of print*. [[CrossRef](#)]

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