

# Preface

Desmoid tumors are currently amongst the rarest of rare tumors that afflict patients. The incidence of these tumors is not as low as is currently believed, however. Misdiagnosed by treating physicians and oncologists alike, especially in cases which remain stable or even regress over time, they may be labeled inaccurately or overlooked entirely. Indeed there are several different pathologic terms for desmoid tumors which confuse the diagnosis. Despite progress in molecular genetic profiling that would aid in precise identification, once designated as benign further efforts at identification are often abandoned.

Over the past decade, at major sarcoma centers, at high esteemed research institutions and at professional meetings such as the prestigious annual CTOS (Connective Tissue Oncology Society) meeting, the importance of understanding desmoid tumors has become increasingly more evident. More research projects were performed and publications submitted in the last 5 years than in the preceding 20 years. Much of this increasing awareness can be credited to the advent of vocal grass-root advocacy groups. Patient education has been heightened through contacts made online and powerful alliances forged between researchers, resulting in shared resources and improved outcomes. However, the majority of patients do not receive their care at dedicated sarcoma centers and many oncologists remain unfamiliar with the identification of currently recommended treatments for desmoid tumors. This book will serve as the first comprehensive publication on the desmoid tumor. Although it may not answer all the questions, as most of these answers have not yet been found, it will introduce the reader, be he a scientist, physician or patient, to what a desmoid is and to the current important players who are leading the quest to find a cure.

Chapter 1 summarizes the increased recognition of the need to identify and treat desmoid tumors; Chap. 2 describes the clinical presentation and epidemiology of desmoid tumors; Chap. 3 discusses the pathology of desmoids; Chap. 4 describes the role of the APC gene and  $\beta$ -catenin in the genesis of desmoid tumors; Chap. 5 reviews the preferred imaging techniques to diagnose and monitor the disease; Chap. 6 outlines the surgical options; Chap. 7 describes current systemic therapy; Chap. 8 and 9 discuss the roles of traditional and interventional radiotherapy in the treatment of desmoid tumors; Chap. 10 describes desmoid tumors in the context of Familial Adenomatous Polyposis; Chap. 11 addresses the unique features and chal-

lenges in treating children and adolescents with desmoid tumor; Chap. 12 details the role of microarrays in studying and distinguishing between desmoids and scar tissue and offers a glimpse into the new techniques of high-throughput sequencing; Chap. 13 outlines the difficulty in categorizing desmoids as benign or malignant and the implications of assigning either label; Chap. 14 examines the role of advocacy groups in promoting better recognition, patient-physician liaisons, researcher interest, desperately needed research funding and emerging patient support systems. Each of these chapters is followed by an extensive list of key references.

I would like to thank all the distinguished authors who enthusiastically agreed to contribute to this book and who without exception are working collaboratively to elucidate the etiology of and advance the search for a cure for this debilitating disorder.

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