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Imaging of Soft Tissue Tumors



Fourth Edition



Springer

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Imaging of Soft Tissue Tumors

Preface to the Fourth Edition

In memoriam Prof. Dr. Arthur M. De Schepper
30 November 1937–04 October 2013



I can vividly remember my first meeting with Professor Arthur De Schepper in Antwerp back in 1988. Being a young resident in radiology educated at the Catholic University of Leuven, I registered for an educational course on ultrasound, organized by Professor De Schepper and his team at Antwerp University Hospital (UZA). In those days, each Flemish university had its own program for (post)graduate radiological education, which was open to all radiologists and trainees. I was immediately impressed by the outstanding educational quality of this meeting and particularly by the open-mindedness of Professor De Schepper, a spirit that he was able to transmit to his whole team of the “Antwerp school of radiology.” I will never forget that he was so easily accessible and that you could talk to him in a very friendly and relaxed atmosphere following the scientific part of the meeting. From that day on, I became a huge fan of the Antwerp school of radiology, and I faithfully attended every single meeting (better known as the “Radiologisch Uur van het UZA”) that was organized in Antwerp.

Later on, after I graduated as a radiologist and I started working at the General Hospital Sint-Maarten Duffel-Mechelen, Professor De Schepper asked me to become a consultant radiologist at Antwerp University Hospital. It was my great privilege to work with him on several projects, such as coediting a book *Medical Imaging of the Spleen* (Springer-Verlag, 2000) and the previous editions of this book *Imaging of Soft Tissue Tumors*, which became his main interest of his very successful scientific career. We developed a close friendship that I will cherish forever.

Arthur De Schepper grew up in Boom, a small industry city south of Antwerp. He completed his medical training at the Catholic University of Leuven and graduated in 1963 as MD. He started his residency of radiology at Stuivenberg General Hospital, under the tutorship of Professor Charles Dochez, who would become a lifelong friend. His main interest in the early stage of his career focused on angiography. He refined his angiography training in the department of Doctor C. Hernandez, at the Hôtel-Dieu Hospital in Paris. After his return to Antwerp, he became chair of radiology at the Stuivenberg General Hospital in 1973. Five years later, he was appointed coordinating chairman of the public assistance hospitals of the city of Antwerp. His special interest in angiography and scientific output in this domain culminated in the successful public defense of his Ph.D. thesis on the ovarian vein syndrome (1976).

In 1978, he was appointed as assistant professor of radiology at the University of Antwerp, and he would become the teacher and mentor of several generations of medical students. At the inauguration of Antwerp University Hospital in 1980, he became chairman of the Department of Radiology. Building from scratch a new imaging department, selecting capable and dedicated collaborators, and developing a structure for teaching residents and students were the challenges he faced. These were exhilarating times, as everything was new and exciting. Arthur De Schepper's success in creating a productive and special ambiance in the Department of Radiology was the result of an intense dedication, true feelings of friendship for his collaborators regardless of their degree or function, critical discussion, and intellectual honesty. He had an uncanny ability to attract students and collaborators of great talent and commitment, and he was able to convey to them his energy and enthusiasm.

Between 1980 and his retirement in 2003, Arthur De Schepper trained more than 80 radiologists. Together with his staff, nurses, technologists, and other collaborators of the department, they constituted his extended family.

His energy and vision resulted in a prolific scientific output. He has authored, or coauthored, more than 350 scientific papers, referenced in PubMed.

In 1986, Antwerp University Hospital was the first academic teaching hospital in Flanders to install a superconducting MRI unit. From then on, Arthur De Schepper developed a keen interest in imaging of bone and soft tissue tumors. He created the "Belgian Soft Tissue Neoplasm Registry" (BSTNR), which provided second opinions to radiologists and other medical specialists from all over the country. His expertise and the BSTNR database led to the publication in 1997 of the first edition of his book *Imaging of Soft Tumors*, which became a huge success. A second edition was published in 2001 followed by a third one in 2006. The book was also translated into Chinese, which reflects the worldwide impact of his work.

After his retirement as chairman of the department in 2003, he remained very active as an expert, and because of his expertise and knowledge, Professor Hans Bloem invited him to become a consultant professor of radiology at Leiden University Medical Center, in the Netherlands. He went on working with unrelenting energy and enthusiasm keeping his body young and his mind alert.

His groundbreaking work on soft tissue tumors made him a world-leading authority on this topic. He was invited to present guest lectures at the European Congress of Radiology (ECR), European Society of Skeletal Radiology (ESSR), Radiological Society of North America (RSNA), and many other major international meetings.

In 2006, Professor De Schepper was awarded Honorary Membership of European Society of Skeletal Radiology (ESSR) for his outstanding contribution to musculoskeletal radiology.

While his medical, scientific, and educational achievements were widely known and appreciated, Arthur De Schepper had human and humane qualities only his family and close friends were allowed to fully appreciate. He was the loving husband of Anya Augustynen, the proud father of 12 children, and the “Papie” of 21 grandchildren. His love for them all, the little things of life, the birds in his garden, and his walks in the woods and wanderings through the mysterious little alleys of Antwerp, it all crystallized into hundreds of poems.

In November 2011, Professor De Schepper suddenly found himself at the other side of the divide, when vague abdominal complaints proved to be symptoms of metastatic colon cancer. With admirable courage, he accepted the diagnosis and its consequences. He never tried to dissimulate the seriousness of his condition and at the same time went on working and helping his colleagues as much as physically possible. On 11 September 2013, at a symposium to celebrate the tenth anniversary of his retirement, he gave his last lecture at the University of Antwerp, an overview of his long career and a lucid summary of what had been and what was to come. Less than 4 weeks later, he left us with a last poem:

*to whom it may concern
bury me cautiously
with a bunch of lavender
and pen and paper
one never knows
my soul might write
the timeless poem
I never could alive
forget pomp and circumstance
sprinkle humour in my grave
and a piece of the rainbow
one never knows
my eye might see
a spray of colours
I never could alive
don't bury me loud
put grains of silence in my grave
and an ivory music box
one never knows
there is no heavenly music
but sounds I will hear
I hardly could alive
leave home your funeral wreaths
your elegies and lament
as for an honourable man
put cuddles and caresses in my grave*

*one never knows
our love survives
(Kindly translated by Prof. Dr. Jan L.M. Bosmans in English)*

Professor Arthur De Schepper will be remembered as a greatly admired and internationally respected radiologist, with an almost superhuman dedication to his work.

We are grateful for the support, warmth, and friendship, which Arthur De Schepper bestowed upon us throughout his long and illustrious career. He will live in our hearts and minds as a leader in radiology, but even more so as our friend, “our” professor.

Finally, we would like to express our special thanks to the family of Professor De Schepper and Springer-Verlag for giving us the opportunity to edit the fourth edition of this book, which is coedited with Professor Jan Gielen and Professor Paul M. Parizel, who is the chair of the Department of Radiology at Antwerp University Hospital.

On behalf of my coeditors, Professor Jan Gielen and Professor Paul M. Parizel, we would like to dedicate this fourth edition to the memory of Professor Arthur De Schepper.

Antwerp, Belgium

Filip M. Vanhoenacker

References

1. Parizel PM, Bosmans JLM. <http://www.auntminnieeurope.com/index.aspx?sec=ser&sub=def&pag=dis&ItemID=608826>
2. Parizel PM, Bosmans JLM (2013). In memoriam Prof. Dr. Emeritus Arthur M. De Schepper. JBR–BTR. 96:329–330

Preface of the Third Edition

The Belgian Soft Tissue Neoplasm Registry (BSTNR) is a multiinstitutional database project involving the cooperation of nearly all magnetic resonance imaging (MRI) centers in Belgium. This initiative, which was started in 2001, had two main goals. First, the BSTNR provided a second opinion report (within 48 h) as a professional courtesy toward all cooperating radiologists. Second, the BSTNR served as a scientific data bank of soft tissue tumors, which are rare lesions in daily radiological practice. All cooperating radiologists had access to the data of the register for use in clinical scientific studies. The scientific value of the BSTNR increased with the installation of a peer-review group of pathologists, all of whom shared a large amount of experience in soft tissue tumor pathology. They reviewed the pathological findings of all malignant tumors, all exceptional tumors, and all tumors in which there was a discordance between MRI and histopathological findings. They guarantee that the pathological standard remains “gold.”

Until now we have included more than 1500 histologically proven soft tissue tumors. This exceptional material constitutes the foundation of this third edition. We are grateful to all the coinvestigators of the BSTNR for their long-term contribution. We asked all coauthors to update their chapters with pertinent new data and images. We also asked them to respect the new World Health Organization classification of soft tissue tumors, which changed considerably in 2002, taking into account the usefulness of the classification for the radiologist. This implies that tumors have moved from one chapter to another according to their tissue of origin and their malignancy grade, e.g., the formerly named malignant fibrous histiocytoma, the synovial cell sarcoma, the hemangiopericytoma, and the solitary fibrous tumor. We also asked our coauthors to include at the end of their chapters a shortlist of striking features and a concise message to take home. The content of many chapters has changed substantially, e.g., the chapter on tumors of connective tissue, on pseudotumors, on biopsy of soft tissue tumors, and on posttreatment follow-up. The chapter on imaging strategy is tuned according to evolution of the MR technique and sequences. In the chapter on MRI, we omit the general principles of the method and focus on the sequences that are currently used in the study of soft tissue tumors. The index at the end of the book is better organized

and more comprehensive. Finally we have added two new chapters, one on pathology and a second on molecular biology and genetics. We asked both authors to focus on those features that are most important to radiologists, who will be the main readers of this book. We are grateful to Springer-Verlag for giving us the opportunity to produce a third edition of a book on a radiological subject, which is a rather exceptional event.

Antwerp, Belgium
March 2005

Arthur M. De Schepper

Preface of the Second Edition

At the time of writing, our group has had more than 10 years' experience in the imaging of soft tissue tumors. We are now,—more than ever,—convinced that a multidisciplinary dialogue between orthopedic surgeons, oncologists, pathologists and radiologists is imperative for the medical management of these lesions. The common goals of all specialists dealing with soft tissue tumors should be: early detection, minimally invasive staging and grading procedures, specific diagnosis (or suitably ordered differential diagnosis), guided percutaneous biopsies, and the most suitable therapy. This approach will guarantee the patient the optimal chances of survival with the best possible quality of life. To help us achieve these goals, we have established a Commission for Bone and Soft Tissue Tumors at the University Hospital in Antwerp, which convenes every 2 weeks. This multidisciplinary group formulates opinions and recommendations on diagnosis, prognosis, treatment and follow-up, and is highly valued by referring physicians. In addition, we are organizing a Belgian Registry of Soft Tissue Tumors with the cooperation of all Belgian centers in which MRI equipment is available and intend to invite students and investigators from all over the world to share our scientific interest in this fascinating field of medical imaging.

The main objective of this second edition of “Imaging of Soft Tissue Tumors” is to provide radiologists with an updated and easy-to-read reference work. This second edition includes new literature references and illustrations. Older illustrations have been replaced with higher quality images, generated by newer equipment and/or MRI pulse sequences. New tables organizing information into summaries have been included and the subject index has been updated. Most importantly, the text contains newer insights (for instance about fibrohistiocytic tumors), and reflects our own experience of increasing understanding of soft tissue tumors and their imaging. The chapter about magnetic resonance imaging has been shortened, and now focuses mainly on principles, pulse sequences and applications that are directly related to the examination of soft tissues and soft tissue tumors. We have included new chapters on “Soft Tissue Tumors in Pediatric Patients” and “Soft Tissue Lymphoma”, and also a chapter on the controversial subject of (percutaneous) biopsy.

The readers and the reviewers of our book will judge whether we have succeeded in our objectives.

Finally, we would like to thank our editor and Mrs. Mennecke-Bühler at Springer-Verlag for sharing in the challenge of editing a second edition of this book on a rare pathology.

Antwerp, Belgium
July 2001

Arthur M. De Schepper

Preface of the First Edition

Although the soft tissues constitute a large part of the human body, soft tissue tumors are rare, accounting for less than 1 % of all neoplasms. The annual incidence of benign soft tissue tumors in a hospital population is 300 per 100,000. Moreover, benign lesions outnumber their malignant counterparts by about 100–1. The clinical and biochemical findings of soft tissue tumors are frequently nonspecific. The first sign is usually a soft tissue swelling or a palpable mass with or without pain or tenderness. Laboratory results are frequently normal or show minimal nonspecific changes.

Until a few decades ago, detection of soft tissue tumors usually did not take place until late in the course of disease. This resulted from their low incidence and nonspecific clinical findings and from the poor sensitivity of conventional radiography, which was the only imaging technique available. Soft tissue tumors and soft tissue disorders in general were practically unknown to radiologists until the introduction of ultrasound and computed tomography (CT). Unfortunately, these methods suffered from inherent drawbacks, such as the poor specificity of ultrasound and the poor contrast resolution of CT.

Many of these problems were solved by the introduction of magnetic resonance imaging (MRI). Thanks to its high contrast tissue resolution and its multiplanar imaging capability, new horizons were opened for imaging soft tissues. Today, a correct assessment of disorders of bones, joints, or soft tissues is unimaginable without MRI.

In view of recent developments in surgery, radiation therapy, systemic chemotherapy, and regional perfusion techniques, the imaging of soft tissue tumors is gaining in importance. Correct diagnosis includes the detection, characterization, and staging of the lesions. The inadequate diagnosis and therapy of soft tissue sarcomas frequently results in tumor recurrence, necessitating major therapeutic “aggression.” MRI is the optimal imaging technique for avoiding inadequate assessment.

Despite the interest of many groups of radiologists in the subject and despite the considerable number of overview articles that have been published in the radiologic literature, soft tissue tumors receive only minimal attention in modern state-of-the-art books on musculoskeletal imaging. Nevertheless, since all radiologists involved in the fascinating field of MRI are now confronted with tumoral pathology of soft tissues, there is a need for an illustrated radiologic guide on the subject.

From the beginning of our experience using MRI, back in 1985, we have been interested in soft tissue tumors. Our initial findings were discussed at an international congress in 1992. Conflicting findings in the literature concerning the sensitivity and specificity of MRI, which were mainly caused by the limited number of patients in published series, prompted us to start a multicenter European study. At the European Congress of Radiology 1993 in Vienna, 29 co-investigators from all over Europe agreed to participate (see the list 'Investigators of Multicentric European Study on Magnetic Resonance Imaging of Soft Tissue Tumors'). More than 1000 cases were collected, which constitute the basis of the radiologic work we prepared.

It was not our intention to write the 'all you ever wanted to know' book on soft tissue tumors. This objective has already been achieved for the pathology of soft tissue tumors by Enzinger and Weiss. Although their famous textbook contains a brief discussion of modern medical imaging, you will find it rarely on the office desk of radiologists. This present book is intended to serve as a reference guide for practising radiologists and clinicians seeking the optimal imaging approach for their patients with a soft tissue tumor.

The book is divided into four sections. In the first section we discuss the different imaging modalities and their respective contribution to the diagnosis of soft tissue tumors. As MRI is generally accepted to be the method of choice, there is a detailed theoretical description of this technique combined with a short discussion of imaging sequences. We also included a chapter on scintigraphy of soft tissue tumors, in which the current literature on the subject is summarized because scintigraphy was hardly used in our own patient material.

The second part deals with staging and characterization of soft tissue tumors and is concluded by a chapter on general imaging strategy. Tumor-specific imaging strategy is, where needed, added at the end of the tumor-specific chapters, which are collected in Part III. These chapters include a short description of epidemiology, clinical and pathological presentation, and a detailed discussion of imaging findings. For this Part, we used the classification of E.B.Chung (Current classification of soft tissue tumors. In: Fletcher CD, McKee PH (eds) *Pathobiology of soft tissue tumors*, 1st edn. Churchill Livingstone, Edinburgh, 1990, pp 43–81), which is an updated version of the most comprehensive system of classification, that of the World Health Organization. Because the illustrations originate from different institutions using different MR systems and pulse sequences, the figure legends only mention the plane of imaging (sagittal, axial, coronal), the kind of sequence (SE, TSE, GRE, ...), and the weighting (T1, T2).

The fourth part consists of only one chapter dealing with post-treatment imaging findings.

I would like to thank my co-editors Dr. Paul Parizel, Dr. Frank Ramon, Dr. Luc De Beuckeleer, and Dr. Jan Vandevenne, and all the coauthors for the tremendous job they have done. From this work I learned that writing a good book requires a sabbatical leave, which good fortune I did not have.

As previously mentioned, it has been possible to include many of the illustrations shown in the book only because of the cooperation of the 29 European investigators, to whom I owe my gratitude. We gratefully acknowledge the

support of Prof. Eric Van Marck, pathologist at our institution, for reviewing the manuscript, and of Ingrid Van der Heyden (secretary) for her aid in preparing so many chapters. Finally, I wish to express my gratitude to Springer-Verlag and to Dr. Ute Heilmann for sharing the challenge of preparing this book with us.

Antwerp, Belgium
June 1996

Arthur M. De Schepper

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