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Pediatric Soft Tissue Tumors

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1. Introduction

The literature on cancer, of the Children Hospital "JM de los Ríos", allowed us to distinguish two series of solid tumors in children. The first serie (1, 2, 3), from 1937 to 1976, with 581 cases of malignant solid tumors, 75 of which were sarcomas (Table N° 1) and a second serie (4, 5), from 1985 to 2001, with 1.796 cases of solid tumors, with 280 cases of soft tissue tumors (Table N° 2). In both series, soft tissue tumors ranked the third in frequency with the 12.90% and 15.96% of cases, respectively. We refer to these statistics, since they are the base of our discussion.

	1937-1962 (1)	1963-1971 (2)	1972-1976 (3)	1937-1976
Lymphoma / Hodgkin	41(13)	104(21)	39(12)	184(46) (31.66%)
Central nervous system tumors	20	56	19	95 (16.35%)
Wilms' tumor.	35	40	11	86 (14.80%)
Sarcomas	27 (soft tissue sarcomas 20, osteosarcomas 7), (fibrosarcomas of soft tissue 6, bone fibrosarcomas 2)	30	18 (soft tissue sarcomas 12, osteosarcomas 6)	75 (12.90%)
Neuroblastoma	10	21	12	43 (7.40%)
Bone tumors				
Retinoblastoma	16	19	2	37 (6.36%)

Teratomas	17	11	12	40 (6.88%)
Carcinoma	8	10	2	20 (3.44%)
Ovarian dysgerminoma			1	1 (0.17%)
Liver tumors				
Other tumors				
Total	174	291	116	581

Table 1. Solid tumors in the Children´s Hospital "JM de los Ríos" 1937-1976 (1, 2, 3)

	1985-1995 (4)	1996-2001	1985-2001 (5)
Lymphoma / Hodgkin	304/135	86/7	390/142 (21.71%)
Central nervous system tumors	199	189	388 (21.60%)
Renal Tumors / Wilms' tumor.	115/98	80/71	195/169 (10.85%)
Soft tissue tumors /Rhabdomyosarcomas	178/114	102/54	280/168 (15.59%)
Neuroblastoma	79	62	141 (7.85%)
Bone tumors	58	67	125 (6.95%)
Retinoblastoma	35	19	54 (3.00%)
Germ cell tumors Germ	111	16	127 (7.07%)
Carcinoma	31	15	46 (2.56%)
Ovarian dysgerminoma			
Liver tumors	25	18	43 (2.39%)
Other tumors	12	-5	7 (0.38%)
Total	1147	649	1796

Table 2. Solid tumors in the Children´s Hospital "JM de los Ríos" 1985-2001

1.1 Incidence

Solid tumors are approximately the 70% of cases of cancer in the children, and the other 30% are leukemias. In the group of solid tumors, soft tissue tumors rank third in frequency with 15.59%, only surpassed by lymphomas 21.71% and central nervous system tumors 21.60% (5) Table N° 2.

Incidence of soft tissue tumors in children and adolescents is not uniform across all ages. It is greater in children younger than 10 years with 73.92% and, children less of 5 years old, account for nearly half (48.57%) the cases of soft tissue tumors (Table N° 3).

Age groups	Porcentaje
Under 1 year of age	27 (9.64%)
1 to 4 years	109 (38.92%)
5 to 9 years	71 (25.35%)
10 to 15 years	69 (24.64%)
16 to 18 years	4 (1.42%)

Table 3. Soft tissue tumors. Distribution by age groups. Children's Hospital "J.M. de los Ríos" 1985-2001 (5)

1.2 Definition

Soft tissue have been defined as nonepithelial extraskeletal tissue of the body, exclusive the reticuloendothelial system, glia and supporting tissue of various parenchymal organs (6); it is represented by voluntary muscle, fat, fibrous tissue and vessels serving these tissues. The soft tissue tumors are a heterogeneous group of tumors that derived from embryonic mesenchymal cells; they are histopathológically classified, according to adult tissue that resembles, and may be benign or malignant. Malignant soft tissue tumors are called sarcomas, and there are three main groups: - rhabdomyosarcoma, - non-rhabdomyosarcoma soft tissue sarcoma, and - Ewing's sarcoma.

2. Rhadomyosarcoma (RMS)

2.1 Definition e incidence

Rhabdomyosarcoma is the most common malignant soft tissue tumor in childhood and adolescense; and represents 60% of cases in under 18 years of age. It is originate from embryonic mesenchymal cells, with potential to differentiate into skeletal muscle cells, and is characterized by a tendency to exhibit histologic and molecular features of skeletal myogenesis. These tumors may arise anywhere in the body, even in sites where skeletal muscle is not normally found (7), and at diagnosis, the more frequent location of the tumor was: head and neck, genitourinary and limbs (8).

2.2 Age distribution

The incidence of rhabdomyosarcoma in childhood and adolescence is 60%, and is higher in the first decade of life with 82.73%. Approximately, 50 percent of the cases of rhabdomyosarcoma (48.80%) are diagnosed in children under 5 years old (1-4 years old) (5), and, represent 75% of soft tissue tumors in this age group tumors. (Table No. 4).

Age groups	soft tissue tumors	Rhabdomyosarcomas
Under 1 year of age	27	12 (44.44%)
1 to 4 years	109	82 (75.22%)
5 to 9 years	71	45 (63.38%)
10 to 15 years	69	27 (39.13%)
16 to 18 years	4	2 (50%)
Total	280	168 (60%)

Table 4. Relation rhabdomyosarcoma/soft tissue tumor. Distribution by age groups. Children's Hospital "J.M. de los Ríos" 1985-2001 (5)

2.3 Histopathologic classification

According to the histopathological and prognosis features, the rhadomyosarcomas are classified in the following varieties (8). Table N°5:

Histopathologic varieties	Prognosis
Botryoid embryonal rhabdomyosarcoma Spindle cell rhabdomyosarcoma	Superior prognosis
Embryonal rhabdomyosarcoma	Intermediate prognosis
Alveolar rhabdomyosarcoma Undifferentiated sarcoma Rhabdomyosarcoma with diffuse anaplasia	Poor prognosis

Table 5. International Prognostic Classification of Pediatric Rhabdomyosarcoma

The incidence of the different varieties of rhabdomyosarcoma, in descending order, is the following (9): Embryonic (64%), Alveolar (21%), undifferenciated (8%), botryoides (6%), pleomorphic (1%).

2.4 Etiology, pathogenesis and cytogenetics

Most rhabdomyosarcomas occur sporadically without predisposing factors, and only one third of patients have recognizable genetic anomalies (7, 9). The cause of the rhabdomyosarcomas remains unknown; but now we know, that certain genetic alterations are associated with the development of this tumor. Alveolar RMS has a characteristic translocation between the long arm of chromosome 2 and the long arm of chromosome 13. This translocation has been cloned molecularly, and shown to involve the juxtaposition of the PAX3 gene, which thought to regulate transcription during early neuromuscular development (7). The embryonal rhabdomyosarcoma, has loss of heterozygosity (LOH) at the 11p15 locus (10).

The Li-Fraumeni syndrome, a well-defined family cancer, that includes rhabdomyosarcoma and other soft tissue sarcomas has been associated with germline mutations of the p53 gene. Rhabdomyosarcoma has been observed in association with Beckwith-Wiedemann syndrome, a fetal overgrowth syndrome associated with abnormalities on 11p15.

The history of cancer in the family is an important, reported in 45.76% of cases. (11).

2.5 Histopathological varieties

There are 4 types of rhabdomyosarcoma in children, clearly defined by Imbach (9), which we reproduce it textually.

2.5.1 Embryonal

- Frequency: 53–64% of all rhabdomyosarcomas in childhood
- Location: orbit, head and neck, abdomen, genitourinary tract
- Microscopically resemblance to embryonic muscle tissue; mainly primitive round cells, some spindle cells with central nucleus and eosinophilic cytoplasm; cross striations characteristic of skeletal muscle in about 30% of cases.
- **Subtype: Sarcoma botryoides** (6% of all rhabdomyosarcomas in children); in vagina, bladder, uterus; microscopically as embryonal type with polypoid mass and presence of a dense subepithelial cell layer.

- **Subtype Spindle cell rhabdomyosarcoma**, is a variety of embryonal rhabdomyosarcoma composed of tight bundles of spindle cells and resembling smooth muscle and fibrous neoplasms (8). It is usually located paratesticular or head and neck, and is difficult to differentiate from congenital fibrosarcoma. Compared to other rhabdomyosarcomas, this has a good prognosis. (spindle cell subtype does not appear in the classification of Imbach (9), so we add it)

2.5.2 Alveolar

- Frequency: 21% of all rhabdomyosarcomas in children
- Location: mainly extremities
- Histology: round cells with eosinophilic cytoplasm, occasionally with vacuoles; multinucleated giant cells; rarely cross-striations; groups of tumor cells separated by fibrotic septation (alveolar structure)

2.5.3 Pleomorphic

- Frequency: 1% of all rhabdomyosarcoma in children
- Occurrence: mainly in adulthood
- Histology: undifferentiated muscle tissue; spindle cells with variable eosinophilic cytoplasm and pleomorphic nuclei, frequently mitotic cells, often cross-striations, structured in rows and bundles.

2.5.4 Undifferentiated subtype

- Frequency: 8% without muscle-specific gene proteins.

The histopathological types found in order of decreasing frequency, in Children's Hospital "J.M. de los Ríos", are shown in Table N°6 (10):

histopathological types	number of cases (%)
Embryonal rhabdomyosarcoma	39 (66.10%)
Embryonal rhabdomyosarcoma botryoides subtype	6 (10.16%)
Embryonal rhabdomyosarcoma spindle cell subtype	1 (1.69%)
Alveolar rhabdomyosarcoma	11 (18.64%)
Non typeable	2 (3.38%)
Total	59 (100%)

Table 6. Rhabdomyosarcoma, Rhabdomyosarcoma histopathological types, Children's Hospital "J.M. de los Ríos" 1997-2005 (10)

The histologic variants embryonal and alveolar are the two more common.

2.6 Location

The most common sites of primary tumor are: head and neck, including the orbit; genitourinary tract including the prostate, testis, vulva, cérvix and uterus; and extremities. (Table N° 7); and less frequent are: trunk, retroperitoneum, perianal and anal (7, 9, 10, 12).

Location	number of cases (%)
Head and neck	20 (33.89%)
Genitourinary	14 (23.72%)
Extremities	13 (22.03%)
Pelvic floor	7 (11.86%)
Trunk	4 (6.77%)
Perianal and anal	1 (1.69%)

Table 7. Rhabdomyosarcoma, Location of primary tumor, Children's Hospital "J.M. de los Ríos" 1997-2005 (10)

The age and location of the tumor are associated with histological varieties certain of rhabdomyosarcoma. Head and neck tumors are more common in children younger than eight years of age and when arising in the orbit are almost always of the embryonal variety, while extremity tumors are more common in adolescents and are typically of the alveolar subtype. The variant botryoides of bladder or vagina, occurs almost exclusively in infants (7).

2.7 Clinical presentation

The clinical manifestations of rhabdomyosarcoma depends on the age at diagnosis, location of primary tumor, and the presence or absence of metastasis.

2.7.1 Head and neck

The primary tumor is usually located in: orbit, head and neck superficial, and parameningeal. Clinical symptoms will depend on the location of the tumor and usually present with a painless, enlarging mass that can obstruct a sinus, grow into the nasal cavity, cause proptosis, or simulate chronic otitis media (12) and clinical symptoms include nasal discharge or obstruction of the airways, otorrhea, and rapid proptosis. The more deep-seated tumors, signs and symptoms may result from compression of nerves, blocked vessels, or both; cranial nerve palsy or other neurological deficits indicates the extent of the tumor at the base of the skull or the central nervous system. (13). The parameningeal localization is the most frequent and a poorer prognosis; usually located in pterygoid infratemporal fossa, nasopharyngeal cavity, paranasal sinuses and middle ear and mastoid, and these four locations include the 91.52% of cases (14).

2.7.2 Genitourinary tract

The embryonal type is the commonest in this región, and arise in the bladder, prostate, vagina, uterus, vulva, paratesticular regions. Children with bladder rhadomyosarcoma are usually under 4 years of age, and may present with hematuria, urinary obstruction and rarely extrusion of tumor tissue. The bladder tumors usually grow intra-luminally, in the region of the trigone and have a polypoidal appearance on gross or endoscopic examination. Prostatic tumors can occur in relatively older children and usually present as large pelvic masses with or without urethral strangury and/ or constipation. Within the category of genitourinary rhabdomyosarcoma, tumors located in the vulva, vagina and paratesticular are a good prognosis; whereas those located in the bladder and prostate have the worst prognosis.

2.7.3 Extremities

Rhabdomyosarcomas are located in the extremities present clinically as painless masses.
As soon as the suspected tumor, this diagnosis should be confirmed through an MRI, and to establish the histopathological diagnosis of the lesion should be performed an open biopsy, or with needle. If frozen sections suggest that the lesion is malignant, tissue samples must be sent for chromosome analysis.

2.8 Staging classification, treatment and prognosis

To plan appropriate treatment, it is necessary to determine the degree of progression of the disease. The clinical classification grouping (15), classifies the extent of the disease into four groups.

IRS clinical grouping classification (stage)

Group I: Localized disease, completely resected, no microscopic residual

Regional nodes not involved—lymph node biopsy or dissection is required except for head and neck lesions

A. Confined to muscle or organ of origin, completed resected
B. Infiltrating beyond site of origin, completely resected

Notation: This includes both gross inspection and microscopic confirmation of complete resection. Any nodes that may be inadvertently taken with the specimen must be negative. If the latter should be involved microscopically, then the patient is placed in group IIB or group IIC (see below).

Group II: Total gross resection with evidence of regional spread, completely resected

A. Grossly resected tumor with microscopic residual disease

Surgeon believes that all the tumor has been removed, but the pathologist finds tumor at the margin of resection, and additional resection to achieve a clean margin is not feasible. No evidence of gross residual tumor; no evidence of regional node involvement; once radiotherapy and/or chemotherapy have been started, re-exploration and removal of the area of microscopic residual does not change the patient's group.

B. Regional disease with involved nodes, completely resected with no microscopic residual

Notation: Complete resection with microscopic confirmation of no residual disease makes this different from group IIA and group IIC. Additionally, in contrast to group IIA, regional nodes (which are completely resected, however) are involved, but the most distal node is histologically negative.

C. Regional disease with involved nodes, grossly resected, but with evidence of microscopic residual and/or histologic involvement of the most distal regional node (from the primary site) in the dissection

Notation: The presence of microscopic residual disease makes this group different from group IIB, and nodal involvement makes this group different from group IIA.

Group III: Incomplete resection with gross residual disease

A. After biopsy only
B. After gross or major resection of the primary (>50%)

Group IV: Distant metastatic disease present at onset

Lung, liver, bones, bone marrow, brain, and distant muscle and nodes.

Notation: The above excludes regional nodes and adjacent organ infiltration, which places the patient in a more favorablegrouping (as noted above under group II).

The presence of positive cytology in the cerebrospinal fluid, pleural or abdominal fluids, as well as implants on pleural or peritoneal surfaces are regarded as indications for placing the patient in group IV.

The size of the primary tumor is a prognostic factor. Tumors less than or equal to 5 cm are classified in the subgroup a, and tumors larger than 5 cm are classified in subgroup b (16). However, this tumor size has a different meaning according to the body surface. A tumor of 5 cm in a children or an adolescent have not the same meaning a 5 cm tumor in a neonate or an infant, what is proposed, relating the size of the primary tumor with the patient's body surface (17).

Rhabdomyosarcoma is a systemic disease, with high probability of spread to lymph nodes, bone marrow, bone, soft tissue distance, and pleural or peritoneal spaces adjacent to the primary site; and have propensity to spread to the lung parenchyma. Therefore, the diagnostic study to determine the extent of the disease includes: obtaining a chest radiograph and lateral, computed tomography (CT) scan of the chest, bone marrow aspiration, biopsy, total body bone scan and cerebrospinal fluid samples from patients with orbital tumors parameningeal or other (eg, tumors of the nasopharynx, paranasal sinuses, pterygoid fossa / infratemporal, and the region middle-ear/mastoid). Patients with rhabdomyosarcomas are classified on the basis de their low, intermediate, or high risk for treatment failure (18). Treatment is then tailored to the appropriate risk level. It is standard practice to repeat imaging studies at 2- to 4-month intervals during and after therapy and to obtain blood samples and chemistries before each course of multiple-agent chemotherapy. (12).

At the present time, more than 70% of children and adolescents with rhabdomyosarcoma are cured with combined modality treatment (chemotherapy, radiation and surgery) (19), but the results will be different depending on the clinical group, histological type, anatomic location and age at presentation of disease, factors that determine the prognosis of the disease and levels of risk of treatment failure.

- The low-risk patients are those with rhabdomyosarcomas of the embryonal variety, in any anatomical location, which have been surgically resected (stages 1-3), or irresectables but in favorable anatomical sites. Favorable anatomic locations are considered nonparameningeal head and neck sites (oropharynx, scalp, parotid, neck, larynx, cheeks, eyelids, hypopharynx), and genitourinary system excluding the bladder and prostate. Their survival rate is over 90% when treated with vincristine and dactinomycin or vincristine, dactinomycin and cyclophosphamide with or without radiotherapy.
- The intermédiate-risk patients are those with unresectable tumors of the embrional variety, in unfavorable locations, ó that are metastatic at the time of diagnosis in patients younger than 10 years old; and all those with non metastatic rhabdomyosarcomas of the alveolar variety. The survival of this group is about 50%-75% and investigated the effectiveness of new drugs such as topotecan. Rhabdomyosarcomas of the alveolar variety, stages I-III, require complementary treatment with radiotherapy
- The high-risk patients are those with metastatic at the time of diagnosis and the survival of this group is only 25%

3. Nonrhabdomyosarcomas soft tissue tumors

3.1 Definition e incidence

Nonrhabdomyosarcomas soft tissue tumors are a heterogeneous group of mesenchymal cell neoplasms, most of which are typified with the named for the mature tissue that the tumor

most resembles and represent 4.23% of solid tumors (5) and 27.14% of soft tissue tumors in patients under 18 years Fig Nº 1

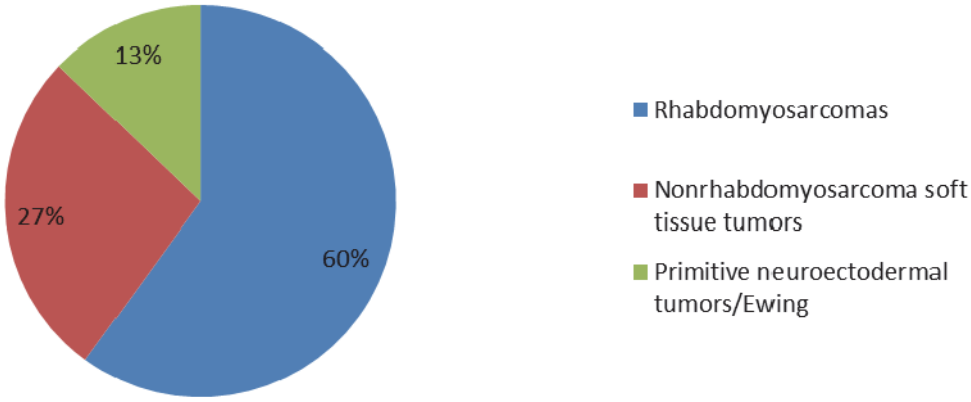

Fig. 1. Soft tissue tumors 280 cases, Children Hospital "JM de los Rios" 1985-2001.

There are differences in soft tissue sarcomas in children and adults in terms of histological types and frequency; differences which have therapeutic implications (20). In Children are more frequent the malignant tumors of the peripheral nerve sheath and fibrosarcoma (5), while adults are more frequent Kaposi's sarcoma, leiomyosarcoma and malignant fibrous histiocytoma (20).

3.2 Age distribution

The distribution of the different varieties of norhabdomyosarcomas soft tissue tumors varies with age; myofibromas and fibrosarcoma are more common in infants, whereas the synovial sarcoma and malignant peripheral nerve sheath tumor is more common in older children and adolescents (12). It is necessary to highlight the characteristics of soft tissue tumors in the first year of life. Approximately 20% of soft tissue tumors that occur in children under 20 years are presented in the first year of age, and of these just over half presented in the first three months of life. 85% of soft tissue tumors present in the first year of life are classified as benign or borderline lesions and the remaining 15% are malignant. The benign and borderline lesions most common are: infantile hemangioendothelioma, lymphangiomas, myofibromas, fibrous histiocytoma, and congenital or infantile fibrosarcoma and represent 68.71% of this group, while embryonal rhabdomyosarcoma and primitive neuroectodermal tumor are the most common malignant lesions, and represent for 62.96% of this group (21).

3.3 Histopathologic classification

Soft tissue tumors are grouped for classification according to cell type that most resembles. Table Nº 8.

Cell Type	Benign Tumor	Malignant Tumor
Fibroblast, including myofibroblast	Fibroma, myxoma	Fibrosarcoma, malignant fibrous histiocytoma
Adipocyte	Lipoma	Liposarcoma
Smooth muscle cell	Leiomyoma	Leiomyosarcoma
Skeletal muscle cell	Rhabdomyoma	Rhabdomyosarcoma
Endothelial cell	Hemangioma	Angiosarocoma, Kaposi sarcoma
Schwann cell	Schwannoma, neurofibroma	Some malignant peripheral nerve sheath tumors
Cartilage cell	Chondroma	Chondrosarcoma
Interstitial cell of Cajal of intestines		Gastrointestinal stromal tumors, a spectrum from benign to malignant
Histiocyte	Juvenile xanthogranuloma Rosai-Dorfman disease?	Histiocytic sarcoma (True histiocytic lymphoma)
Lymphoid cells	Benign lymphoid hyperplasia	Extranodal lymphomas in soft tissues
No known normal cell or benign counterparts		Ewing family tumors Synovial sarcoma Epithelioid sarcoma Alveolar soft part sarcoma

[a]Intermediate categories between benign and malignant tumors are excluded for simplicity.

Table 8. Simplified Chart of the Major Types of Primary Soft Tissue Tumors Grouped According to the Cell Types that They Resemblea (22)

3.4 Etiology, pathogenesis and cytogenetics

The cause of the soft tissue sarcomas remains unknown, but in nonrhabdomyosarcomas soft tissue tumors have identified genetic alterations. In the fibrosarcoma, a characteristic translocation t (12; 15) (p13; q25) with an ETV6-NTRK3 gene fusion, and gains of chromosomes 8, 11, 17 and 20 (23). Synovial sarcoma has and a specific chromosomal translocation t(X;18)(p11;q11) (23).

3.5 Location

The anatomical location of primary tumor in descending order of frequency is: head and neck, trunk and extremities. However this varies depending on the histologic type of tumor. The most frequent primary tumor site was: in fibrosarcoma, the limbs in 66% (24); in myofibromas, head, neck, and trunk in 69% (25); in sinovial sarcoma, the limbs nears joints and tendons in the 77.96% (26).

3.6 Clinical presentation

The most common clinical presentation is a painless mass, although the involvement of adjacent structures can cause pain and other symptoms (27)

3.7 Staging classification, treatment and prognosis

Non rhabdomiosarcomas soft tissue tumors can be benign or malignant and malignant are called sarcomas. The histopathologic features determine the prognosis and treatment follow (28, 29) and hence the classification of degrees. Table Nº 9

Grade 1
Myxoid and well-differentiated liposarcoma
Well-differentiated or infantile (≤4 years old) fibrosarcoma
Well-differentiated or infantile (≤4 years old) hemangiopericytoma
Well-differentiated malignant peripheral nerve sheath tumor
Angiomatoid malignant fibrous histiocytomab
Deep-seated dermatofibrosarcoma protuberans
Myxoid chondrosarcoma
Grade 2
≤15% of the surface area shows necrosis
The mitotic count is <5 mitotic figures per 10 high-power fields using a 340 objective
Nuclear atypia is not marked
The tumor is not markedly cellular
Grade 3
Pleomorphic or round-cell liposarcoma
Mesenchymal chondrosarcoma
Extraskeletal osteogenic sarcoma
Malignant triton tumor
Alveolar soft part sarcoma
Any other sarcoma not in grade 1 with >15% necrosis and/or ‡5 mitotic figures per10 high-power fields using a 340 objective

Table 9. The Pediatric Oncology Group Grading System for Nonrhabdomyosarcomatous Soft Tissue Sarcomas of Children

The proper classification of non rhabdomyosarcomas soft tissue tumors is not easy and raises diagnostic problems that require additional methods such as immunohistochemistry, genetic studies, and consultation by experts (30). Myofibromas are benign lesions with difficulty diagnostic because they may be mistaken for malign lesions with hemangiopericytoma-like findings (31).

The evalucación diagnosed, requires magnetic resonance imaging and computed tomography to determine the extent of the disease and a plan for surgery (32).

Treatment and prognosis depends on the extent of disease and histological type of injury, poor prognostic factors are: high histologic grade, intra-abdominal primary tumor, and microscopic residual disease after initial resection (32, 33). Radiation therapy is useful in tumors whose location does not allow excision and all tumors larger than 5 cm. or that could not be completely resected. Sarcomas in children are more sensitive to chemotherapy report answer 40 to 60% using multiple drugs, hence its use in all tumors larger than 5 cm, of axial location, with histological high grade, or with metastatic disease.

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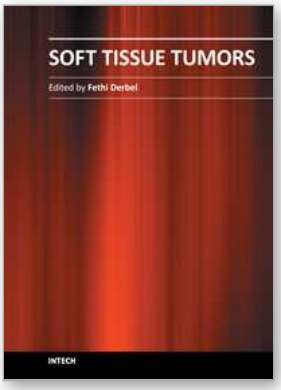

Soft Tissue Tumors
Edited by Prof. Fethi Derbel

ISBN 978-953-307-862-5
Hard cover, 270 pages
Publisher InTech
Published online 16, November, 2011
Published in print edition November, 2011

Soft tissue tumors include a heterogeneous group of diagnostic entities, most of them benign in nature and behavior. Malignant entities, soft tissue sarcomas, are rare tumors that account for1% of all malignancies. These are predominantly tumors of adults, but 15% arise in children and adolescents. The wide biological diversity of soft tissue tumors, combined with their high incidence and potential morbidity and mortality represent challenges to contemporary researches, both at the level of basic and clinical science. Determining whether a soft tissue mass is benign or malignant is vital for appropriate management. This book is the result of collaboration between several authors, experts in their fields; they succeeded in translating the complexity of soft tissue tumors and the diversity in the diagnosis and management of these tumors.

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