

# *Manejo de la pieza quirúrgica. Márgenes*



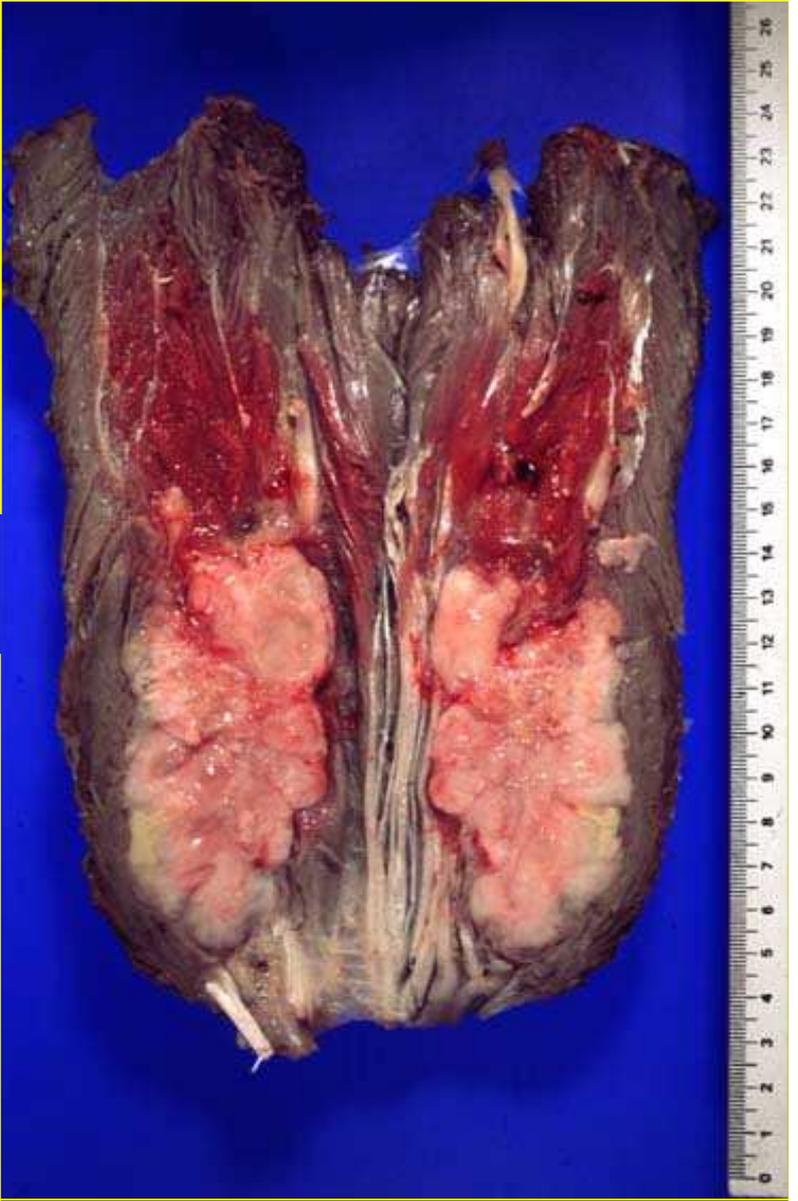
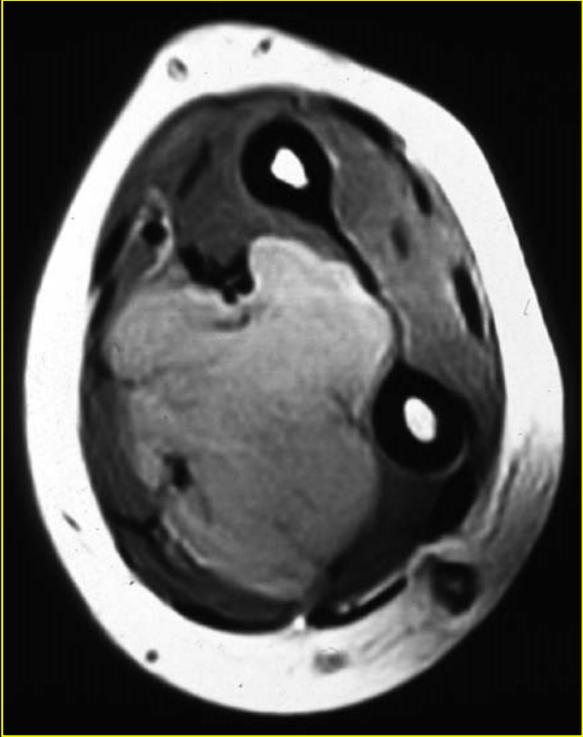
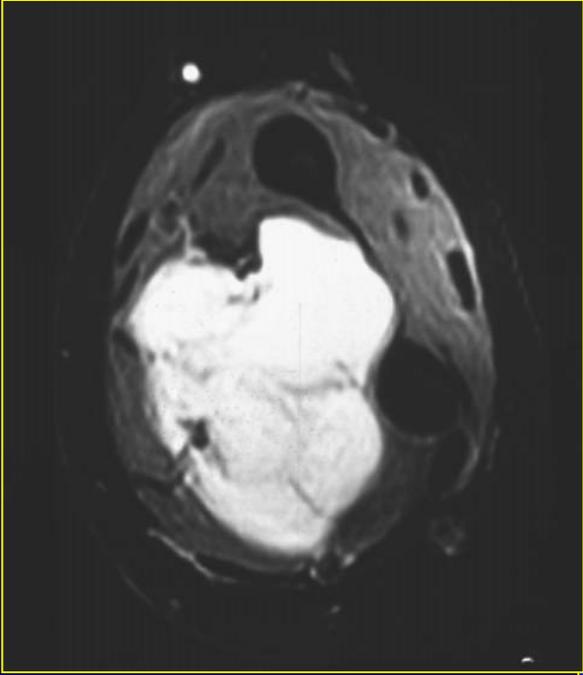
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# **Gradación histológica** es considerado el factor pronóstico mas importante en Sarcomas de partes blandas

- Russell et al . Cancer 1977;40:156-70
- Costa et al. Cancer 1984;53:530-41
- Trojani et al. Cancer 1984;33:37-43
- Lack et al. J Surg Oncol 1989;41:63-73
- Coindre et al. J Clin Oncol 1997;15:350-63

## Debe ser efectuada en :

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- Solo sarcomas (no pseudosarcomas)
- Sarcomas no tratados con neoadyuvancia.
- Material representativo y bien procesado.
- No reemplaza tipo histológico.
- No tiene valor en MPNST, angiosarcoma, condrosarcoma mixoide, sarcoma alveolar de partes blandas, sarcoma de células claras, sarcoma epitelioides.

# Parámetros de gradación histológica

(tiene que ser práctica y reproducible, confiable y certera)

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- **Indice mitótico**
- **Necrosis**
- **Diferenciación tumoral**
- **Celularidad**
- **Grado de atipía nuclear**
- **Permeación vascular**

# Factores que potencialmente pueden afectar la determinación del grado

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- Tiempo de fijación del tejido
- Espesor del corte histológico
- Muestreo inadecuado (tejido reactivo periférico, pseudo cápsula, tejido necrótico)
- Variación en la arquitectura y citología en un mismo tumor.
- Número de campos evaluados
- Variabilidad intratumoral de la actividad mitótica.
- Heterogeneidad de la densidad celular intratumoral.

El grado de un tumor es la variable histológica que marca la tendencia a producir MTS.

Las lesiones que se caracterizan por la marcada atipía celular, elevado índice mitótico, extensa necrosis, marcada vascularización y pobre diferenciación presentan mayor probabilidad de metástasis que los tumores que no presentan estos rasgos histológicos.

	Signos de buen pronóstico	Signos de mal pronóstico
Tamaño	Pequeño < de 5cm.	Grande > de 5cm
localización	superficial	profunda
Grado histológico	I-II	II-III

SLEM a los 5 años 90%	SLEM a los 5 años 50-70%	SLEM a los 5 años 35%
< de 5cm.	> de 5cm.	> de 5cm
superficiales	Profundos o superficiales	profunda
<b>Grado histológico I- II</b>	<b>Grado histológico II- III</b>	<b>Grado histológico III</b>

## Histological classification of soft tissue tumors

- I. Fibrous tumors**
- A. Benign tumors**
- Nodular fasciitis (including intravascular and cranial types)
  - Proiferative fasciitis and myositis
  - Aypical decubital fibroplasia (ischemic fasciitis)
  - Fibroma (dermal, tendon sheath, nuchal)
  - Keloid
  - Elastofibroma
  - Calcifying aponeurotic fibroma
  - Fibrous hamartoma of infancy
  - Fibromatosis coli
  - Infantile digital fibromatosis
  - Myofibromatosis (solitary, multicentric)
  - Hyalin fibromatosis
  - Calcifying fibrous pseudotumor
- B. Fibromatoses**
- Superficial fibromatoses
    - Palmar and plantar fibromatosis
    - Penile (Peyronie's) fibromatosis
    - Knuckle pads
  - Deep fibromatoses (desmoid tumor)
    - Abdominal fibromatosis (abdominal desmoid)
    - Extraabdominal fibromatosis (extraabdominal desmoid)
    - Intraabdominal fibromatosis (intraabdominal desmoid)
    - Mesenteric fibromatosis (including Gardner's syndrome)
    - Infantile (desmoid-type) fibromatosis
- C. Malignant tumors**
- Fibrosarcoma
    - Adult fibrosarcoma
    - Congenital or infantile fibrosarcoma
    - Inflammatory fibrosarcoma (inflammatory myofibroblastic tumor)
- II. Fibrohistiocytic tumors**
- A. Benign tumors**
- Fibrous histiocytoma
    - Cutaneous fibrous histiocytoma (dermatofibroma)
      - Deep fibrous histiocytoma
      - Juvenile xanthogranuloma
      - Recurvifasciitis
      - Xanthoma
    - Intermediate tumors
      - Aypical fibroxanthoma
      - Dermatofibrosarcoma protuberans (including pigmented form, Bednar tumor)
      - Giant cell fibroblastoma
      - Plexiform fibrohistiocytic tumor
      - Angiomatoid fibrous histiocytoma
- B. Malignant tumors**
- Leiomyoma (cutaneous, deep and pleomorphic)
  - Angiomyoma (vascular leiomyoma)
  - Epithelioid leiomyoma
  - Intravenous leiomyomatosis
  - Leiomyomatosis peritonealis disseminata
- III. Lipomatous tumors**
- A. Benign tumors**
- Lipoma
    - Cutaneous lipoma
    - Deep lipoma
      - Intramuscular lipoma
      - Tendon sheath lipoma
      - Lumbosacral lipoma
      - Intraaxonal and perineural fibrolipoma
    - Multiple lipomas
  - Angiolipoma
  - Spindle cell or pleomorphic lipoma
  - Myolipoma
  - Angiomyolipoma
  - Myelolipoma
  - Chondroid lipoma
  - Hibernoma
  - Lipoblastoma or lipoblastomatosis
  - Lipomatosis
    - Diffuse lipomatosis
    - Cervical, symmetrical lipomatosis (Madelung's disease)
  - Atypical lipoma
- B. Malignant tumors**
- Liposarcoma
    - Well-differentiated liposarcoma
      - Lipoma-like liposarcoma
      - Sclerosing liposarcoma
      - Inflammatory liposarcoma
    - Myxoid liposarcoma
    - Round cell (poorly differentiated myxoid) liposarcoma
    - Pleomorphic liposarcoma
    - Dedifferentiated liposarcoma
- IV. Smooth muscle tumors**
- A. Benign tumors**
- Leiomyoma (cutaneous, deep and pleomorphic)
  - Angiomyoma (vascular leiomyoma)
  - Epithelioid leiomyoma
  - Intravenous leiomyomatosis
  - Leiomyomatosis peritonealis disseminata
- B. Malignant tumors**
- Leiomyosarcoma
  - Angiosarcoma
  - Epithelioid leiomyosarcoma
  - Myxoid leiomyosarcoma
  - Round cell leiomyosarcoma
  - Pleomorphic leiomyosarcoma
  - Dedifferentiated leiomyosarcoma
- V. Skeletal muscle tumors**
- A. Benign tumors**
- Adult rhabdomyoma
  - Genital rhabdomyoma
  - Fetal rhabdomyoma
  - Intermediate (cellular) rhabdomyoma
- B. Malignant tumors**
- Rhabdomyosarcoma
    - Embryonal rhabdomyosarcoma
    - Alveolar rhabdomyosarcoma
    - Spindle cell rhabdomyosarcoma
    - Atypical rhabdomyosarcoma
  - Pleomorphic rhabdomyosarcoma
  - Rhabdomyosarcoma with ganglionic differentiation (extenstomyoma)
- VI. Tumors of blood and lymph vessels**
- A. Benign tumors**
- Papillary endothelial hyperplasia
  - Hemangioma
    - Cavernary (including juvenile) hemangioma
    - Cavernous hemangioma
    - Venous hemangioma
    - Epithelioid hemangioma (angiolymphoid hyperplasia, histiocytoid hemangioma)
    - Granulation type hemangioma (pyogenic granuloma)
    - Traumatic hemangioma
  - Deep hemangioma (intramuscular, synovial, perineural)
  - Lymphangioma
  - Lymphangiomatosis and lymphangio-matosis
  - Angiomatosis
  - Lymphangiosarcoma
- B. Malignant tumors**
- Angiosarcoma
  - Hemangioendothelioma
  - Epithelioid hemangioendothelioma
  - Endovascular papillary angioendothelioma (Kaposi's tumor)
  - Spindle cell hemangioendothelioma
  - Malignant tumors
    - Angiosarcoma and lymphangiosarcoma
    - Kaposi's sarcoma
- VII. Perivascular tumors**
- A. Benign tumors**
- Giant tumor
- B. Malignant tumors**
- Storiform-pleomorphic fibrous histiocytoma
  - Myxoid fibrous histiocytoma
  - Giant cell fibrous histiocytoma (malignant giant cell tumor of soft parts)
  - Xanthomatous (inflammatory type) fibrous histiocytoma
- VIII. Synovial tumors**
- A. Benign tumors**
- Tenosynovial giant cell tumor
    - Localized tenosynovial giant cell tumor
    - Diffuse tenosynovial giant cell tumor (extraarticular pigmented villonodular synovitis, fibrod tenosynovitis)
- B. Malignant tumors**
- Synovial sarcoma
    - Biphasic (fibrous and epithelial) synovial sarcoma
    - Monophasic (fibrous or epithelial) synovial sarcoma
  - Malignant giant cell tumor of tendon sheath
- IX. Mesothelial tumors**
- A. Benign tumors**
- Benign mesothelioma
    - Lowest tumor of pleura and peritoneum (localized fibrous mesothelioma)
  - Multicystic mesothelioma
  - Adenomatoid tumor
  - Well-differentiated papillary mesothelioma
- B. Malignant tumors**
- Malignant solitary fibrous tumor of pleura
  - Diffuse mesothelioma
    - Epithelial diffuse mesothelioma
    - Fibrous (spindled, sarcomatoid) diffuse mesothelioma
    - Biphasic diffuse mesothelioma
- X. Neural tumors**
- A. Benign tumors**
- Neuroma
    - Morrie's neuroma
    - Multiple mucosal neuromas
    - Neuromuscular hamartoma (benign Triton tumor)
  - Nerve sheath glioma
  - Schwannoma (neurileioma)
    - Cystic schwannoma
    - Plexiform schwannoma
    - Degenerated (ancient) schwannoma
  - Schwannomatosis
  - Neurofiblioma (nerve sheath myxoma)

## Histological classification of soft tissue tumors—cont'd

- B. Malignant tumors**
- Epithelioid sarcoma
  - Epithelial fibrosarcoma
- B. Malignant tumors**
- Malignant glomus tumor
  - Malignant hemangiopericytoma
- VIII. Synovial tumors**
- A. Benign tumors**
- Tenosynovial giant cell tumor
    - Localized tenosynovial giant cell tumor
    - Diffuse tenosynovial giant cell tumor (extraarticular pigmented villonodular synovitis, fibrod tenosynovitis)
- B. Malignant tumors**
- Synovial sarcoma
    - Biphasic (fibrous and epithelial) synovial sarcoma
    - Monophasic (fibrous or epithelial) synovial sarcoma
  - Malignant giant cell tumor of tendon sheath
- IX. Mesothelial tumors**
- A. Benign tumors**
- Benign mesothelioma
    - Lowest tumor of pleura and peritoneum (localized fibrous mesothelioma)
  - Multicystic mesothelioma
  - Adenomatoid tumor
  - Well-differentiated papillary mesothelioma
- B. Malignant tumors**
- Malignant solitary fibrous tumor of pleura
  - Diffuse mesothelioma
    - Epithelial diffuse mesothelioma
    - Fibrous (spindled, sarcomatoid) diffuse mesothelioma
    - Biphasic diffuse mesothelioma
- X. Neural tumors**
- A. Benign tumors**
- Neuroma
    - Morrie's neuroma
    - Multiple mucosal neuromas
    - Neuromuscular hamartoma (benign Triton tumor)
  - Nerve sheath glioma
  - Schwannoma (neurileioma)
    - Cystic schwannoma
    - Plexiform schwannoma
    - Degenerated (ancient) schwannoma
  - Schwannomatosis
  - Neurofiblioma (nerve sheath myxoma)

## Histological classification of soft tissue tumors—cont'd

- 8. Neurofibroma
    - a. Diffuse neurofibroma
    - b. Plexiform neurofibroma
    - c. Pacinian neurofibroma
    - d. Epithelioid neurofibroma
  - 9. Granular cell tumor
  - 10. Melanocytic schwannoma
  - 11. Ectopic meningioma
  - 12. Ectopic ependymoma
  - 13. Ganglioneuroma
  - 14. Pigmented neuroectodermal tumor of infancy (retinal anlage tumor, melanotic progenoma)
  - B. Malignant tumors
    - 1. Malignant peripheral nerve sheath tumor (MPNST) (malignant schwannoma, neurofibrosarcoma)
      - a. Malignant Triton tumor (MPNST with rhabdomyosarcoma)
      - b. Glandular MPNST (malignant glandular schwannoma)
      - c. Epithelioid MPNST (malignant epithelioid schwannoma)
    - 2. Malignant granular cell tumor
    - 3. Clear cell sarcoma (malignant melanoma of soft parts)
    - 4. Malignant melanocytic schwannoma
    - 5. Gastrointestinal autonomic nerve tumor (plexosarcoma)
      - a. Neuroblastoma
      - b. Ganglioneuroblastoma
      - c. Neuroepithelioma (peripheral neuroectodermal tumor)
      - d. Extraskelatal Ewing's sarcoma
- XI. Paraganglionic tumors
  - A. Benign tumors
    - 1. Paraganglioma
  - B. Malignant tumors
    - 1. Malignant paraganglioma
- XII. Extraskelatal cartilaginous and osseous tumors
  - A. Benign tumors
    - 1. Panniculitis ossificans and myositis ossificans
    - 2. Fibroosseous pseudotumor of the digits
    - 3. Fibrodysplasia (myositis) ossificans progressiva
    - 4. Extraskelatal chondroma or osteochondroma
    - 5. Extraskelatal osteoma
  - B. Malignant tumors
    - 1. Extraskelatal chondrosarcoma
      - a. Well-differentiated chondrosarcoma
      - b. Myxoid chondrosarcoma
      - c. Mesenchymal chondrosarcoma
    - 2. Extraskelatal osteosarcoma
- XIII. Pluripotential mesenchymal tumors
  - A. Benign tumors
    - 1. Mesenchymoma
  - B. Malignant tumors
    - 1. Malignant mesenchymoma
- XIV. Miscellaneous tumors
  - A. Benign tumors
    - 1. Congenital granular cell tumor
    - 2. Tumoral calcinosis
    - 3. Myxoma
      - a. Cutaneous myxoma
      - b. Intramuscular myxoma
      - c. Juxtaarticular myxoma
    - 4. Angiomyxoma
    - 5. Amyloid tumor
    - 6. Parachordoma
    - 7. Ossifying and nonossifying fibromyxoid tumors
    - 8. Palisaded myofibroblastoma of lymph node
  - B. Malignant tumors
    - 1. Alveolar soft part sarcoma
    - 2. Epithelioid sarcoma
    - 3. Malignant extrarenal rhabdoid tumor
    - 4. Desmoplastic small cell tumor
- XV. Unclassified tumors

Parameter	Score
<b>Degree of tumor differentiation</b>	
Close resemblance to normal adult tissue (e.g., well-differentiated liposarcoma)	1
Tumor type clearly recognizable (e.g., alveolar soft part sarcoma)	2
Tumor type uncertain (e.g., undifferentiated sarcoma)	3
<b>Tumor necrosis</b>	
No tumor necrosis on any slide	0
Less than 50% tumor necrosis	1
More than 50% tumor necrosis	2
<b>Mitotic count</b>	
0-9 /per 10 HPF	1
10-19 /per 10 HPF	2
20+ / per 10 HPF	3
<b>Histological grade</b>	<b>Total score</b>
Grade 1	2, 3
Grade 2	4, 5
Grade 3	6, 7, 8

Modified from Coindre JM et al: *Cancer* 58:306, 1986.

# Factores pronósticos no morfológicos

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- Contenido aneuploide de DNA
- Marcadores proliferativos
- Organizadores nucleolares (AgNORs)
- Ki67, factores de crecimiento (PDGF-B)
- Bcl-2

**La estadificación es el proceso de clasificación de un tumor, con respecto a su grado de diferenciación, extensión local y a distancia a fin de establecer un pronóstico para el paciente.**

**AJC staging of soft tissue sarcomas:  
definitions of TNMG**

**T: Primary tumor**

T1 Tumor less than 5 cm

T2 Tumor 5 cm or greater

**N: Regional lymph nodes**

N0 No histologically verified metastasis to regional lymph nodes

N1 Histologically verified regional lymph node metastasis

**M: Distant metastasis**

M0 No distant metastasis

M1 Distant metastasis

**G: Histological grade of malignancy**

G1 Low (well-differentiated)

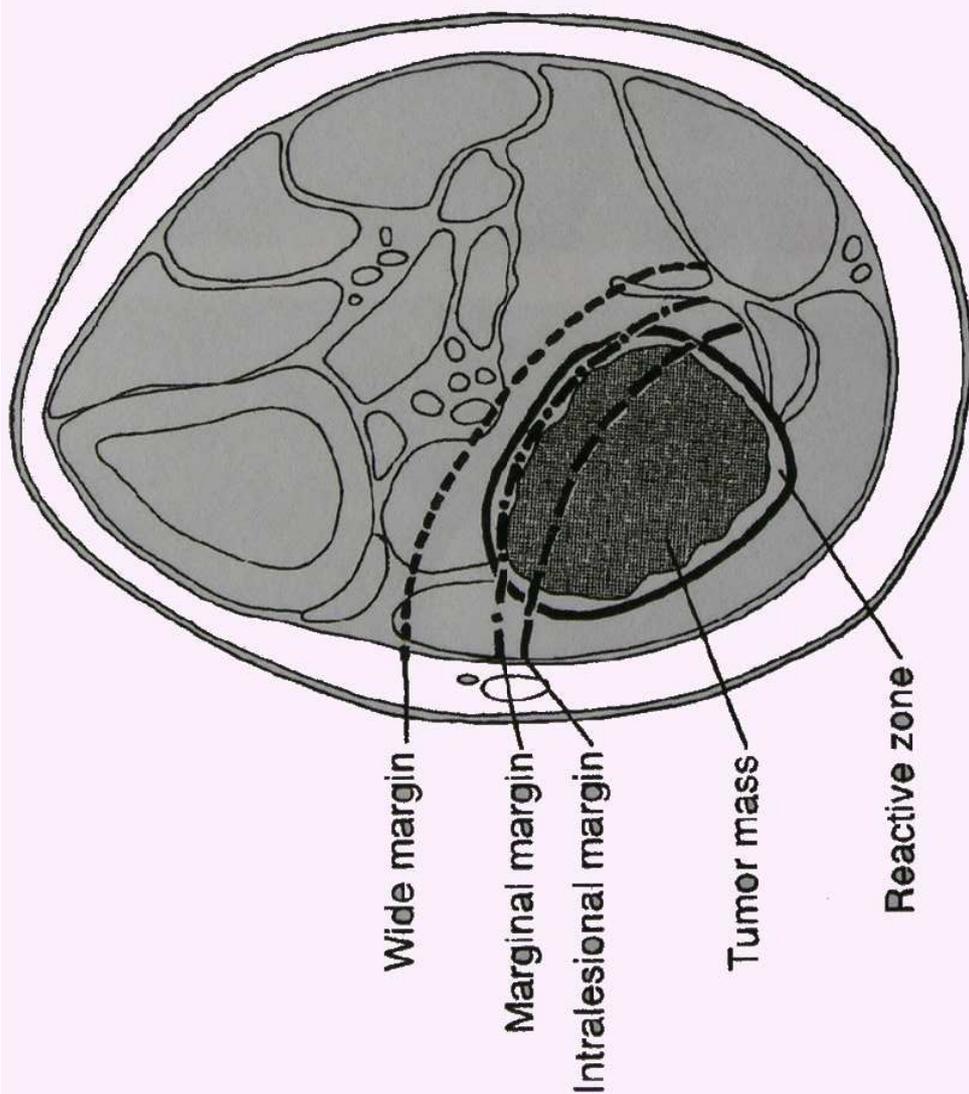
G2 Moderate (moderately well-differentiated)

G3 High (poorly differentiated)

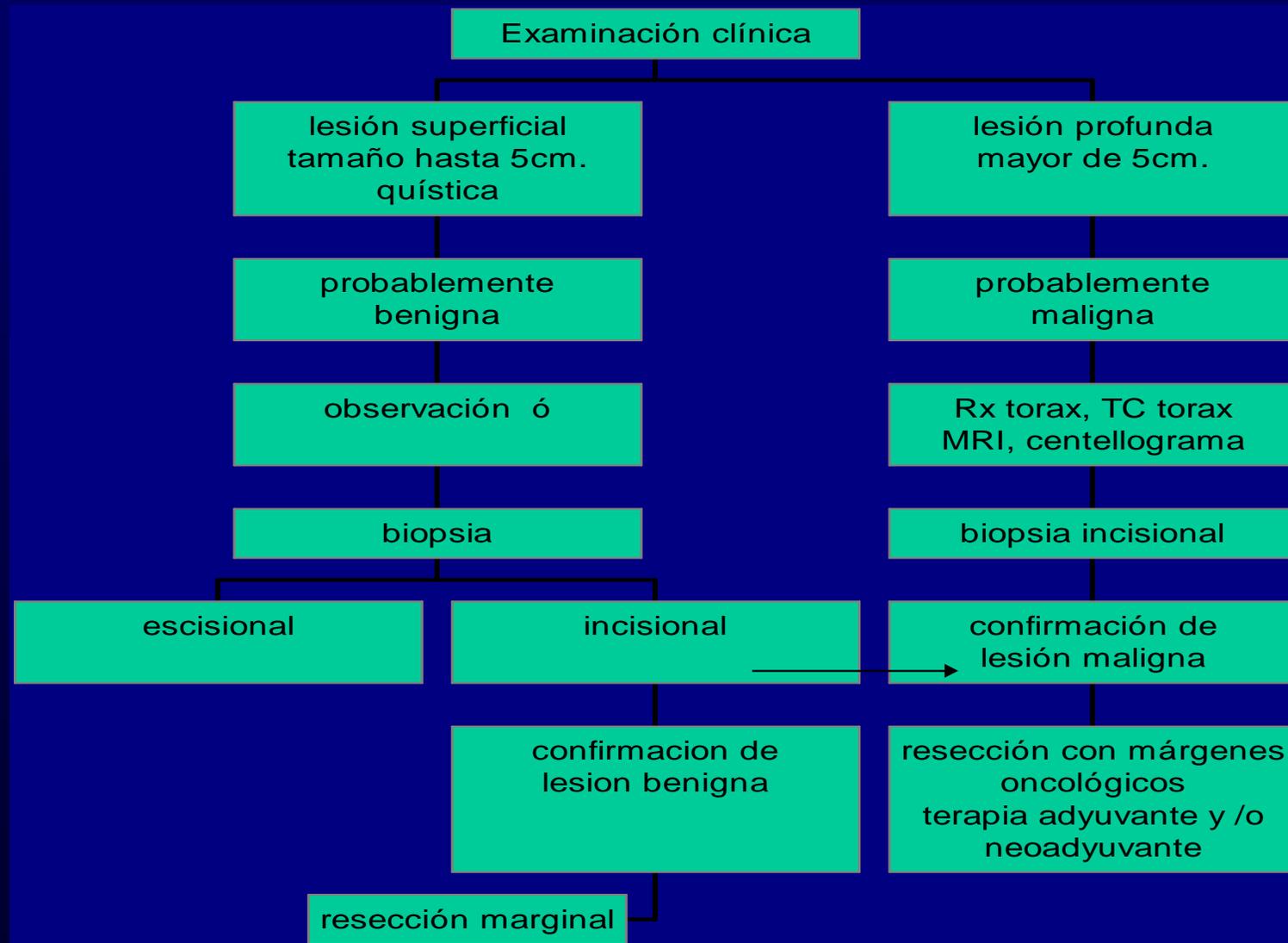
G4 Undifferentiated

From Beahrs OH et al: American Joint Committee on Cancer, 1992.

**Fig. 2** Diagram of types of surgical margins. An intralesional line of resection enters the substance of the tumor. A marginal line of resection travels through the reactive zone of the tumor. A wide surgical margin removes the tumor with a cuff of normal tissue.



# Estrategia en la evaluación, estadificación y tratamiento



## **En los informes anatómo-patológicos debe constar los siguientes datos**

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- Tamaño del tumor (diámetro mayor)
- Tejidos aledaños al tumor, espesor de los mismos,
- Relación del tumor y los límites de resección teniendo como referencia las marcas efectuadas por el cirujano.
- Consistencia, áreas de necrosis y cambios mixoides.
- Tipo histológico.
- Gradación histológica.

## Potencial biológico (OMS 2003)

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- **Benigno:** no recidivan o lo hacen en forma no agresiva (FH cutáneo, lipoma)
- **Intermedio:** localmente agresivo, recidiva local. Crecimiento infiltrativo. Sin potencial metastásico. Amplia resección (desmoide).
- **Intermedio:** potencialmente metastásico, localmente agresivo. Metástasis a distancia <2% no predecible en base a H-P. (FH angiomatoide, FH plexiforme).
- **Maligno:** Localmente destructivo. Recidiva. Metástasis a distancia >20%

