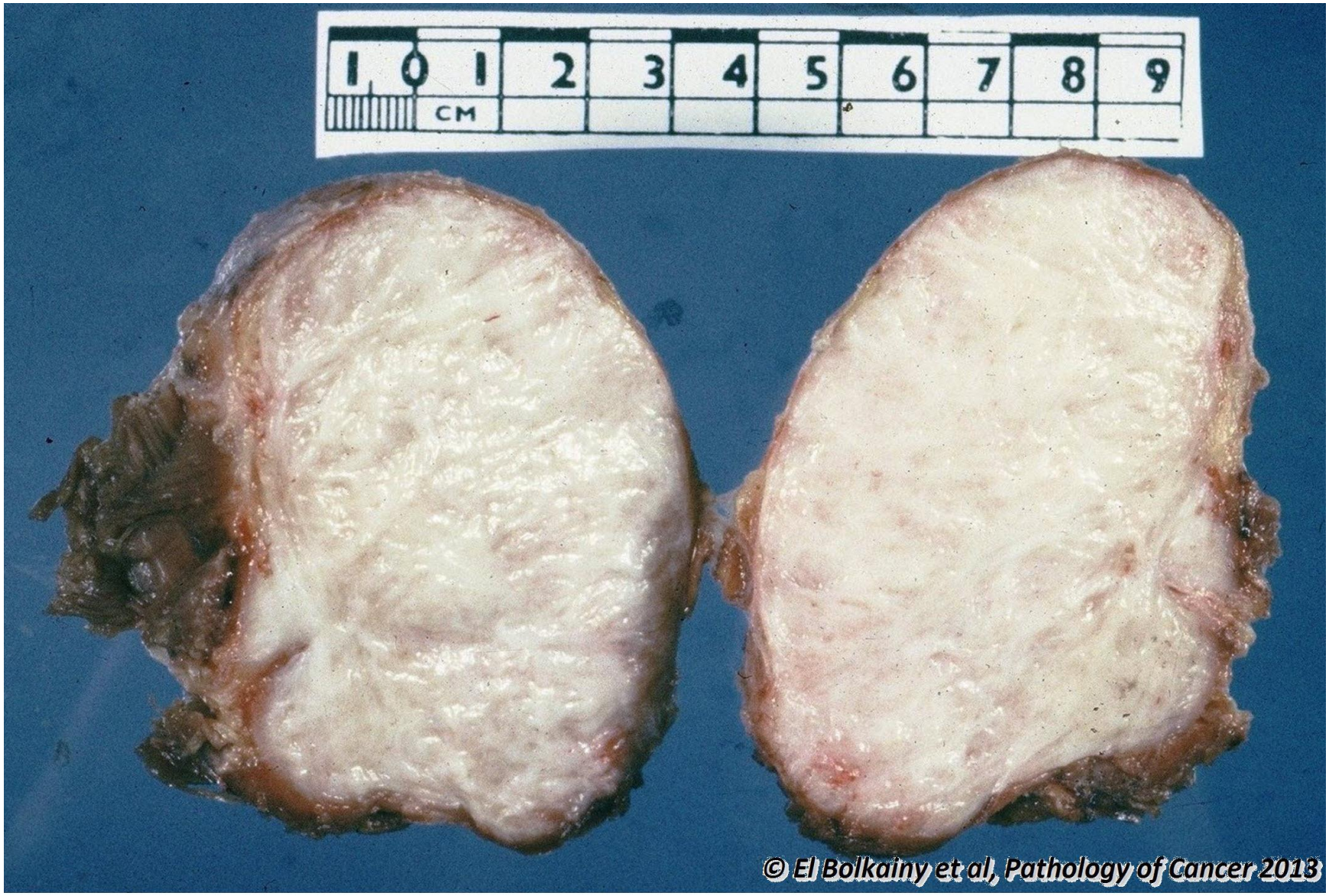
Chapter 21

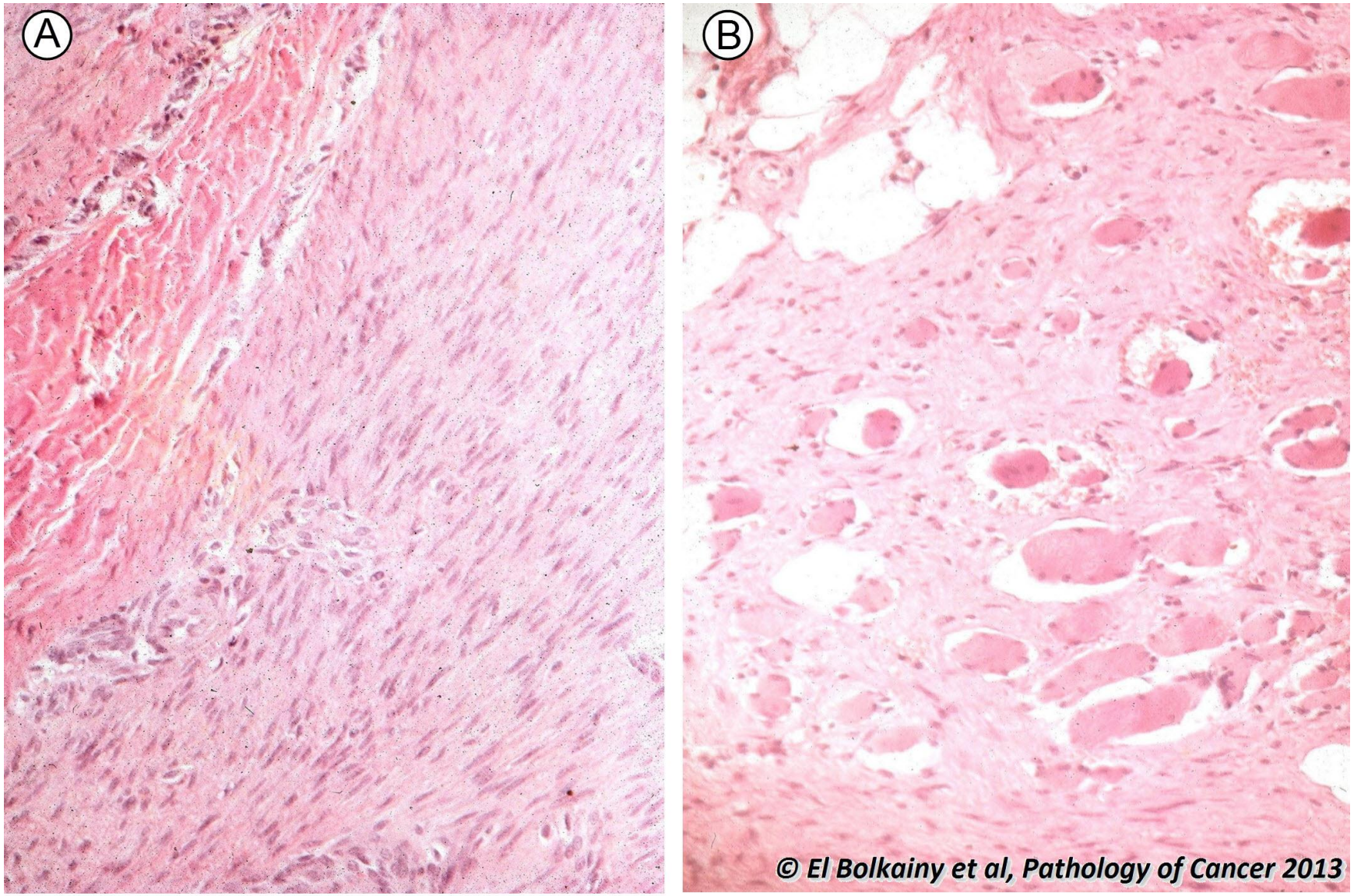
Soft tissue and peripheral neu-roectodermal tumors,

21.1 Fibromatosis, gross features of abdominal desmoid tumor.



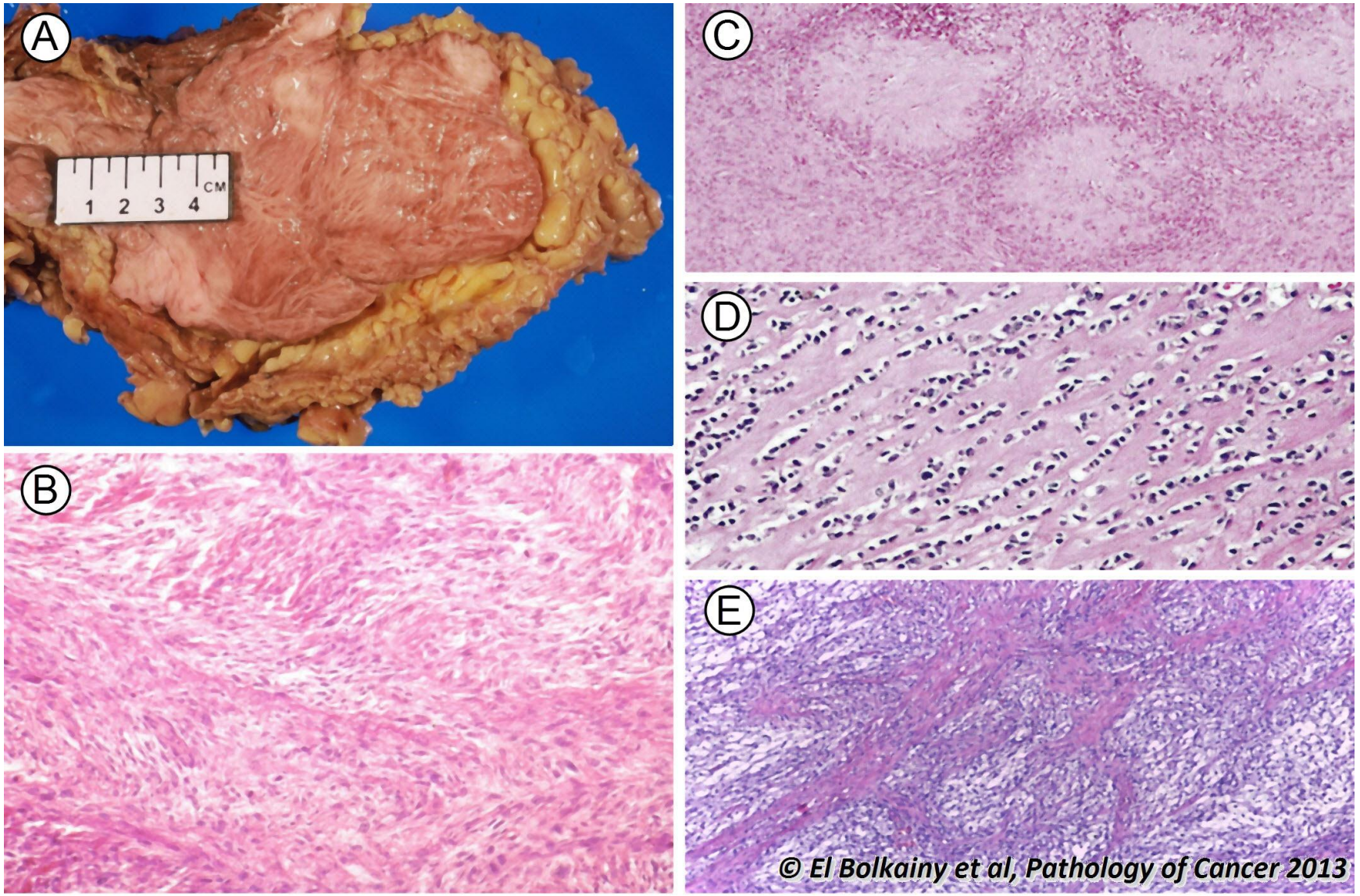
Picture 21-1 **Fibromatosis, gross features of abdominal desmoid tumor.** A whitish fibrotic trabeculated tumor with ill-defined margins.

21.2 Fibromatosis, histology.



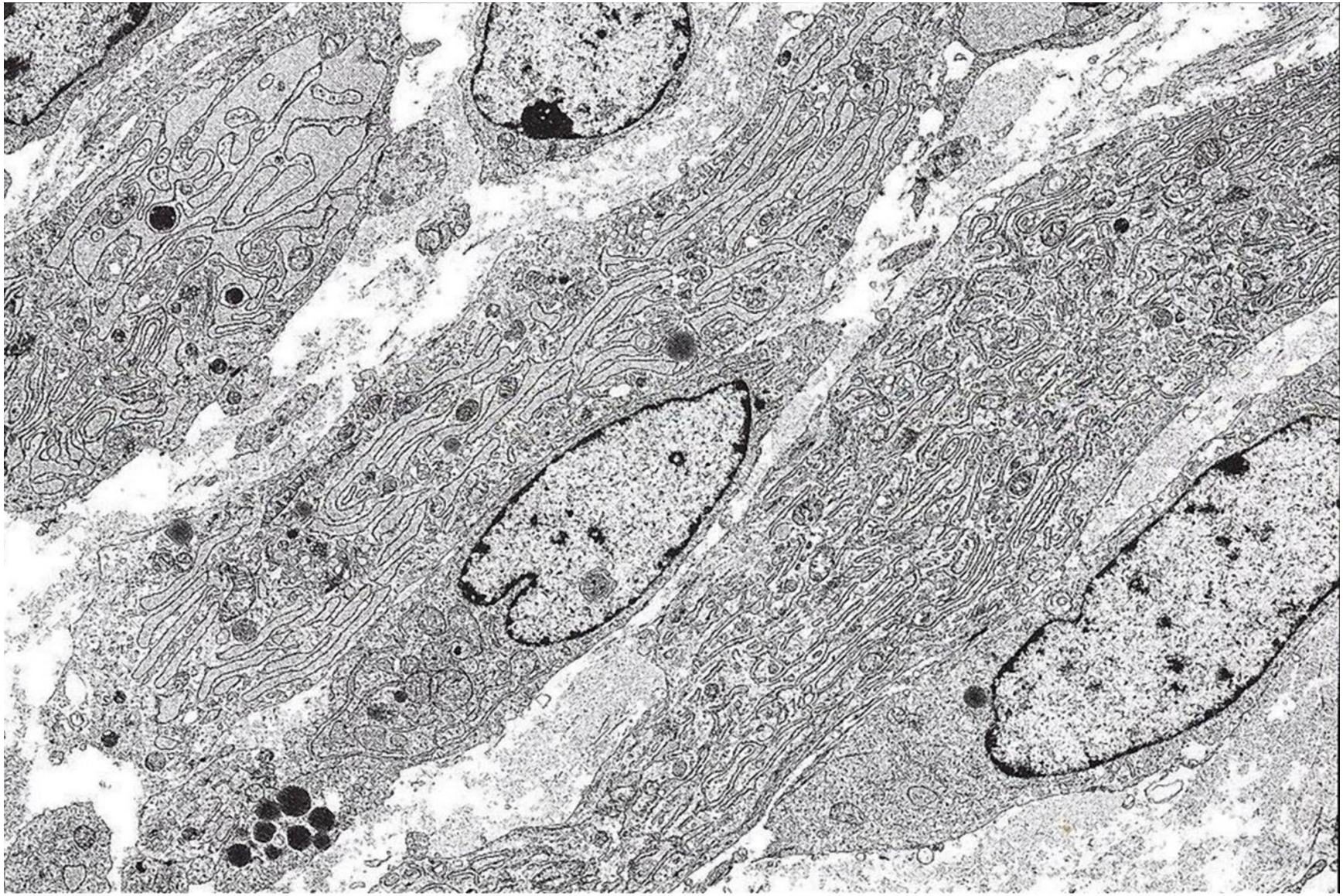
Picture 21-2 **Fibromatosis, histology. A** The tumor is hypocellular (stromal collagen more than myofibroblasts), lacks mitotic figures. The cells have ill-defined cytoplasm. Rarely, metaplastic osteoid component may be evident (osteofibromatosis). Nuclear immunoreactivity to ß-catenin is confirmatory. **B** Tumor margin showing muscle invasion.

21.3 Fibrosarcoma and its variants.



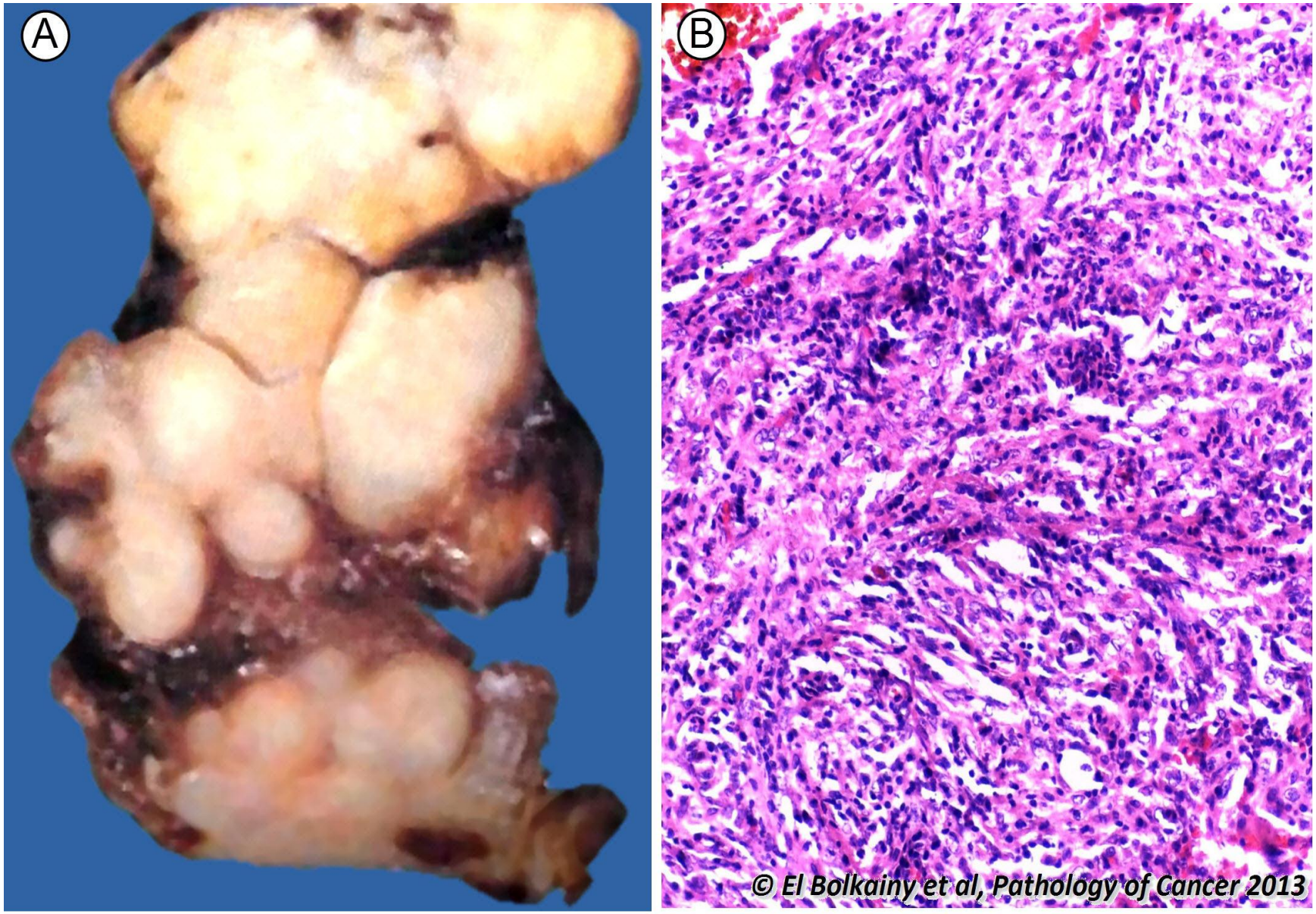
Picture 21-3 **Fibrosarcoma and its variants. A** Grossly, cut section shows trabeculated appearance. **B** Conventional type, histology. Hypercellular tumor (fibroblasts more than collagen), short bundles crossing at an angle (Herring-bone), well-defined cytoplasm, pointed nuclei and mitosis (**>** 2/10HPF). **C** Low-grade myxohyaline sarcoma, with giant pseudorosettes. **D** Sclerosing epithelioid sarcoma. **E** Myxofibrosarcoma, myxoid areas 30% of tumor.

21.4 Fibrosarcoma, electron microscopic (EM) features.



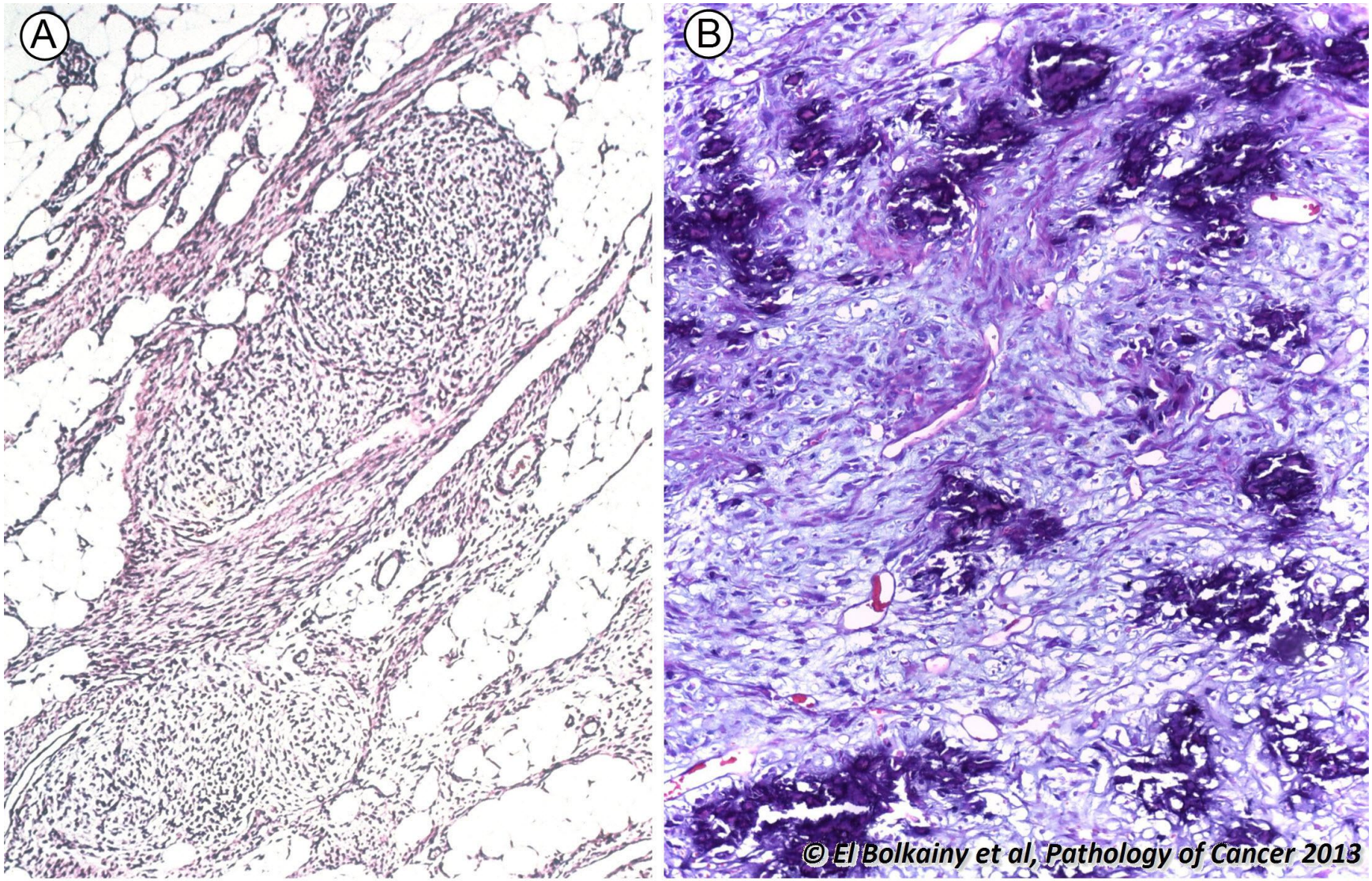
Picture 21-4 **Fibrosarcoma, electron microscopic (EM) features.** Characteristic of fibroblastic differentiation is the presence of many cisternae throughout the cytoplasm.(Reprduced with permission, Fletcher CD, 2007).

21.5 Inflammatory myofibroblastic tumor.



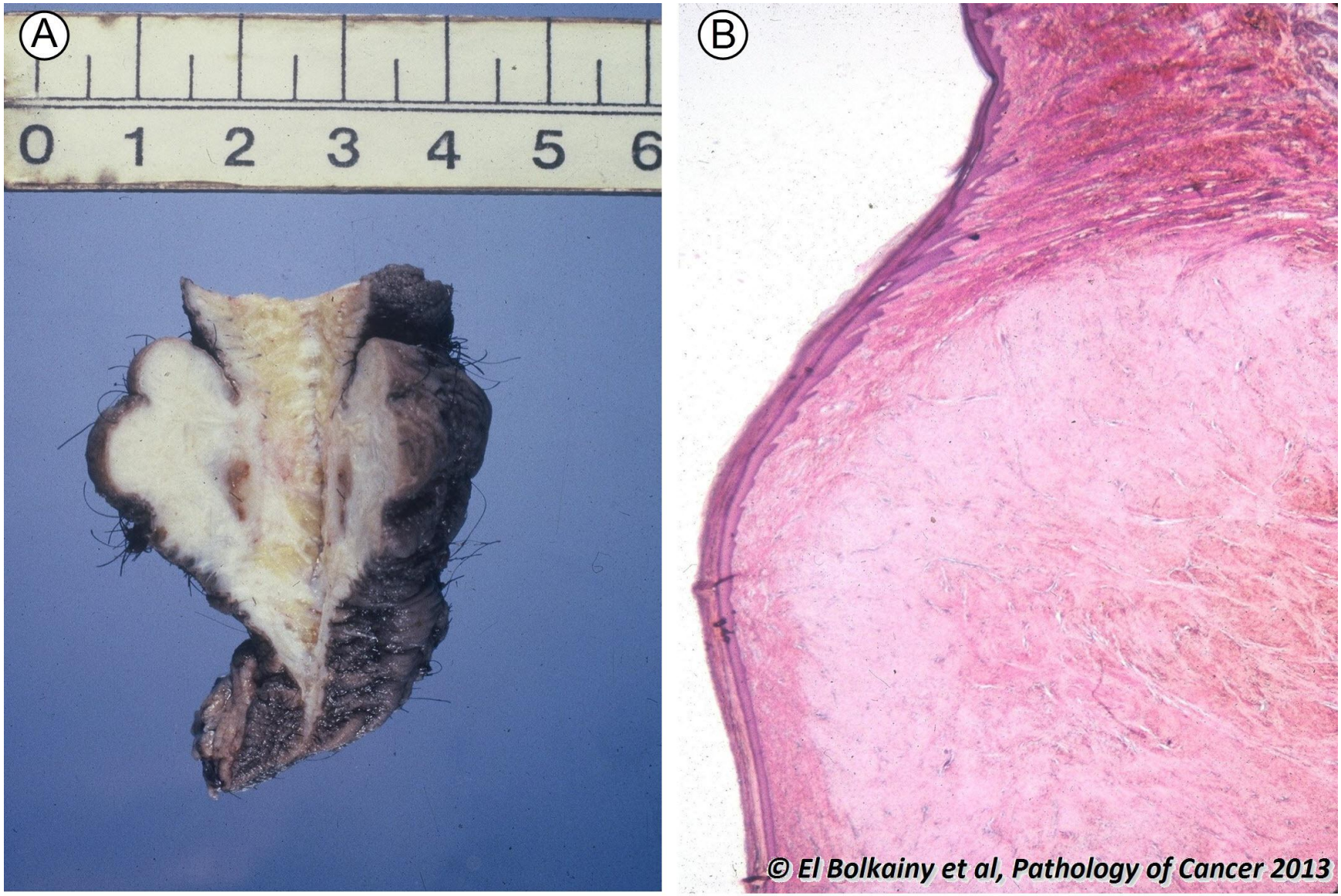
Picture 21-5 **Inflammatory myofibroblastic tumor. A** Grossly, a circumscribed multinodular greyish white rubbery mass with occasional myxoid areas. **B** Myofibroblastic proliferation in bundles with focal inflammatory cells. The tumor cells are positive for actin and ALK-1.

21.6 Fibrous proliferations in infancy.



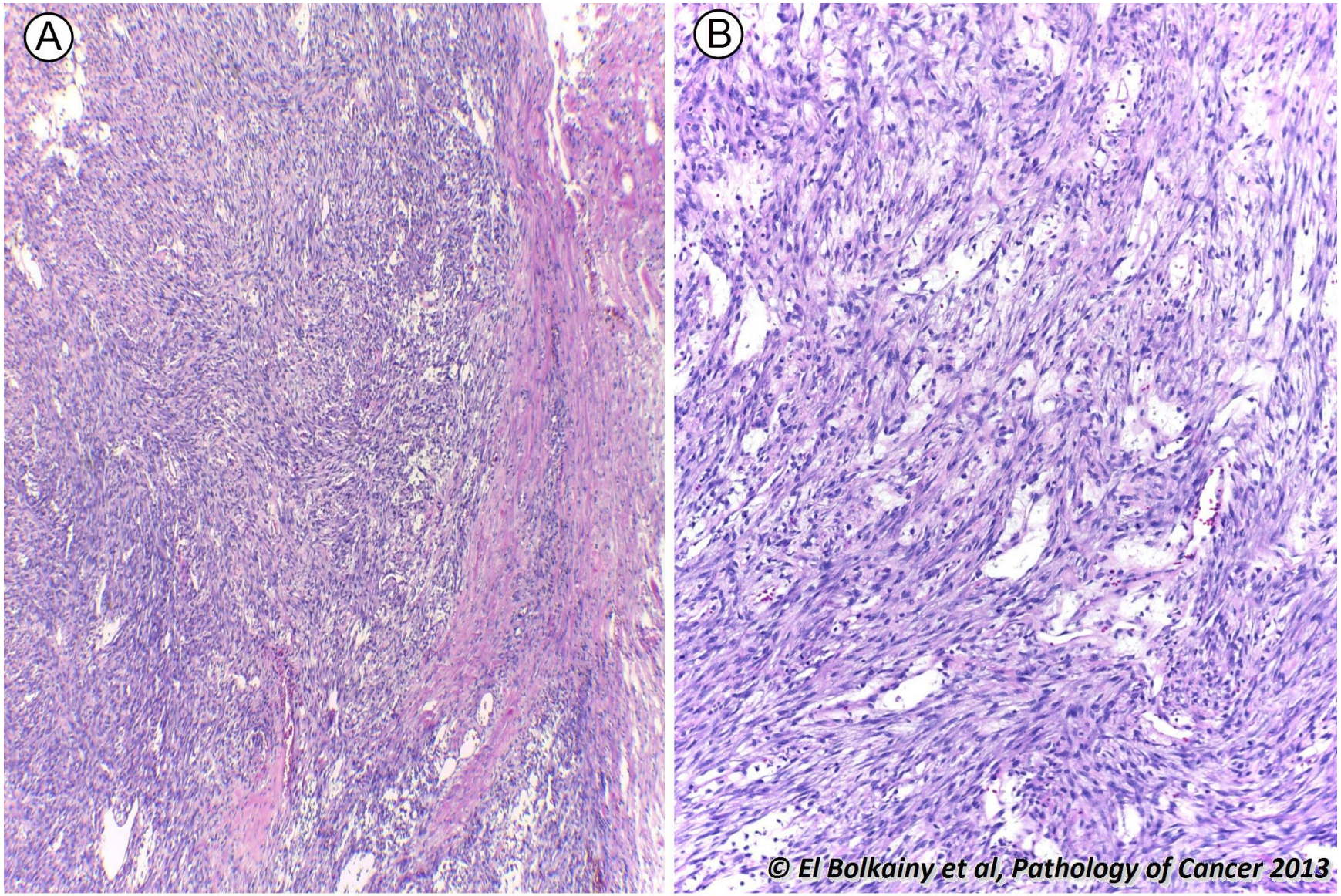
Picture 21-6 **Fibrous proliferations in infancy. A** Fibrous hamartoma of infancy. Three diagnostic cell components (triphasic) are evident in subcutaneous fat; fibroblasts, myofibroblasts, rounded as well as stellate primitive mesenchymal cells. An organoid pattern is characteristic. **B** Calcifying aponeurotic fibroma. Typically affects hands and feet. Fibroblastic proliferation with hyaline stroma is associated with focal calcification and chondroid matrix.

21.7 Keloid after surgery.



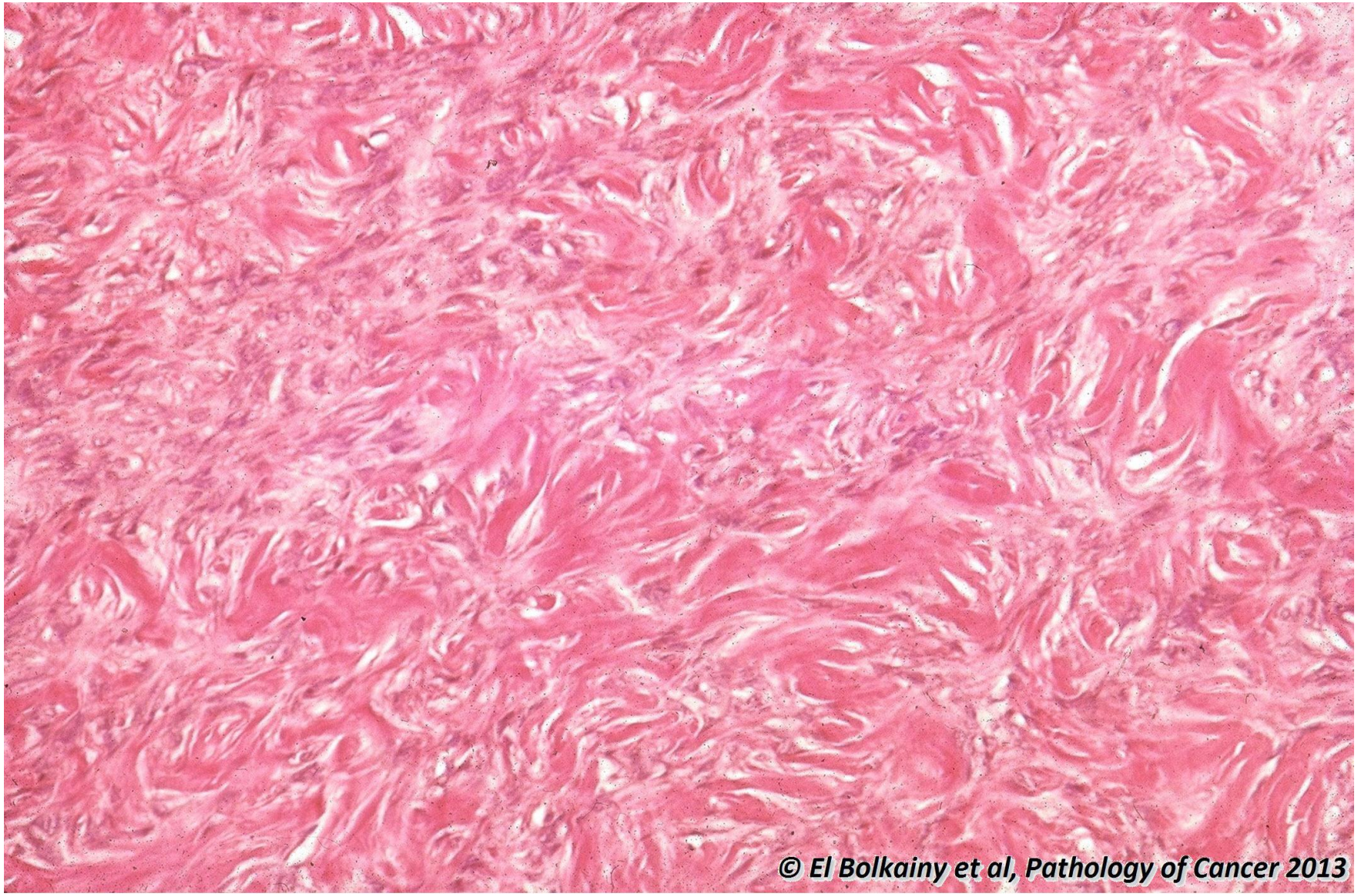
Picture 21-7 **Keloid after surgery. A** Gross features, superficial whitish fibrotic mass related to surgical wound intimately adherent to skin. **B** Histology, the skin lacks adnexal structures. The lesion shows few fibrocytes but abundant thick collagen.

21.8 Nodular fasciitis.



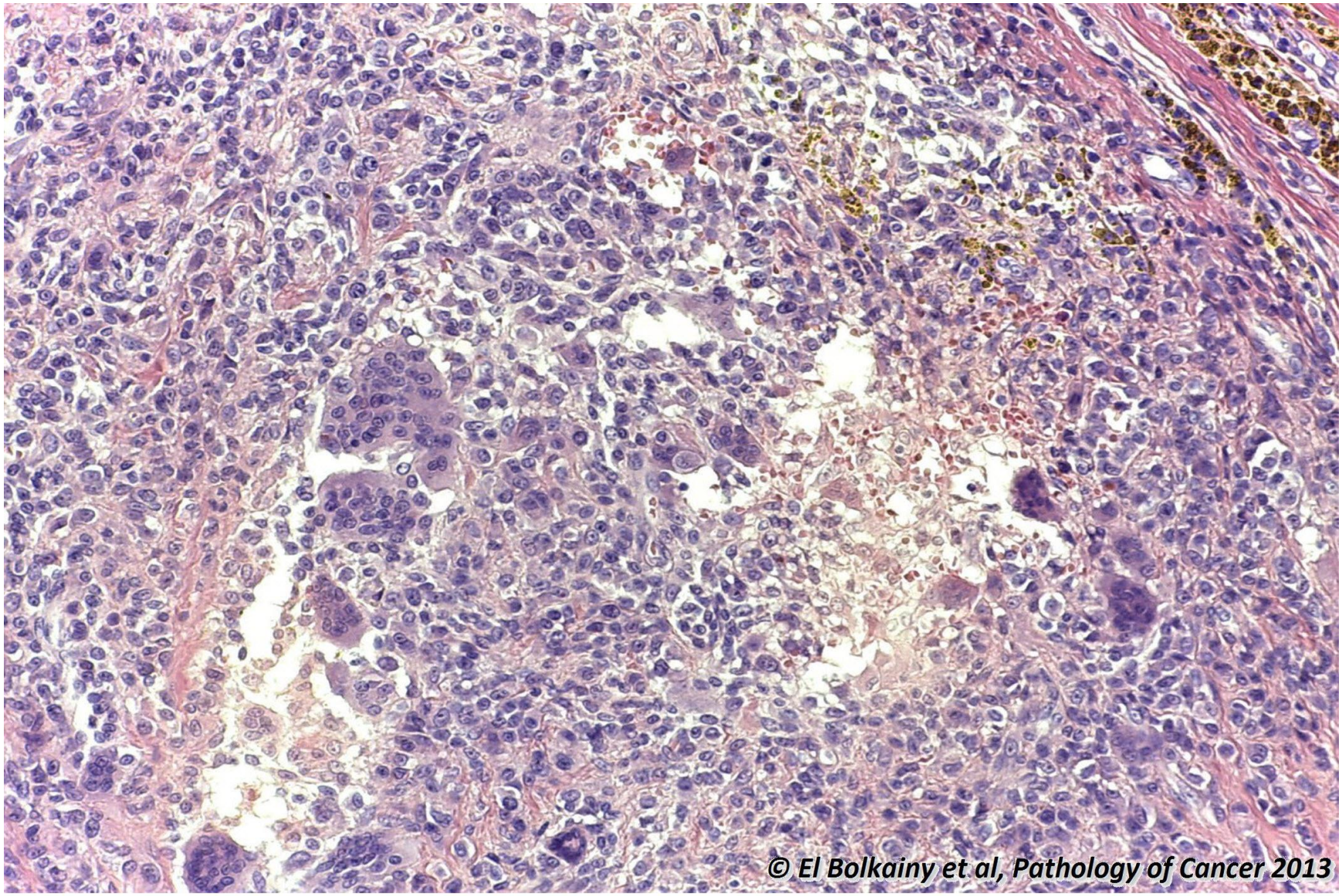
Picture 21-8 **Nodular fasciitis.** This tumor is common in young adults at forearm. **A** Low power, It shows zonal phenomenon (cellular circumscribed periphery and loose arrangement at center). **B** High power, the proliferating fibroblasts are loosely arranged (tissue culture-like), stroma myxoid with microscopic spaces with extravasated red blood cells and lymphocytes. Proliferative fasciitis variant (not shown) contains in addition ganglion-like blasts with abundant cytopasm.

21.9 Benign fibrous histiocytoma (dermatofibroma).



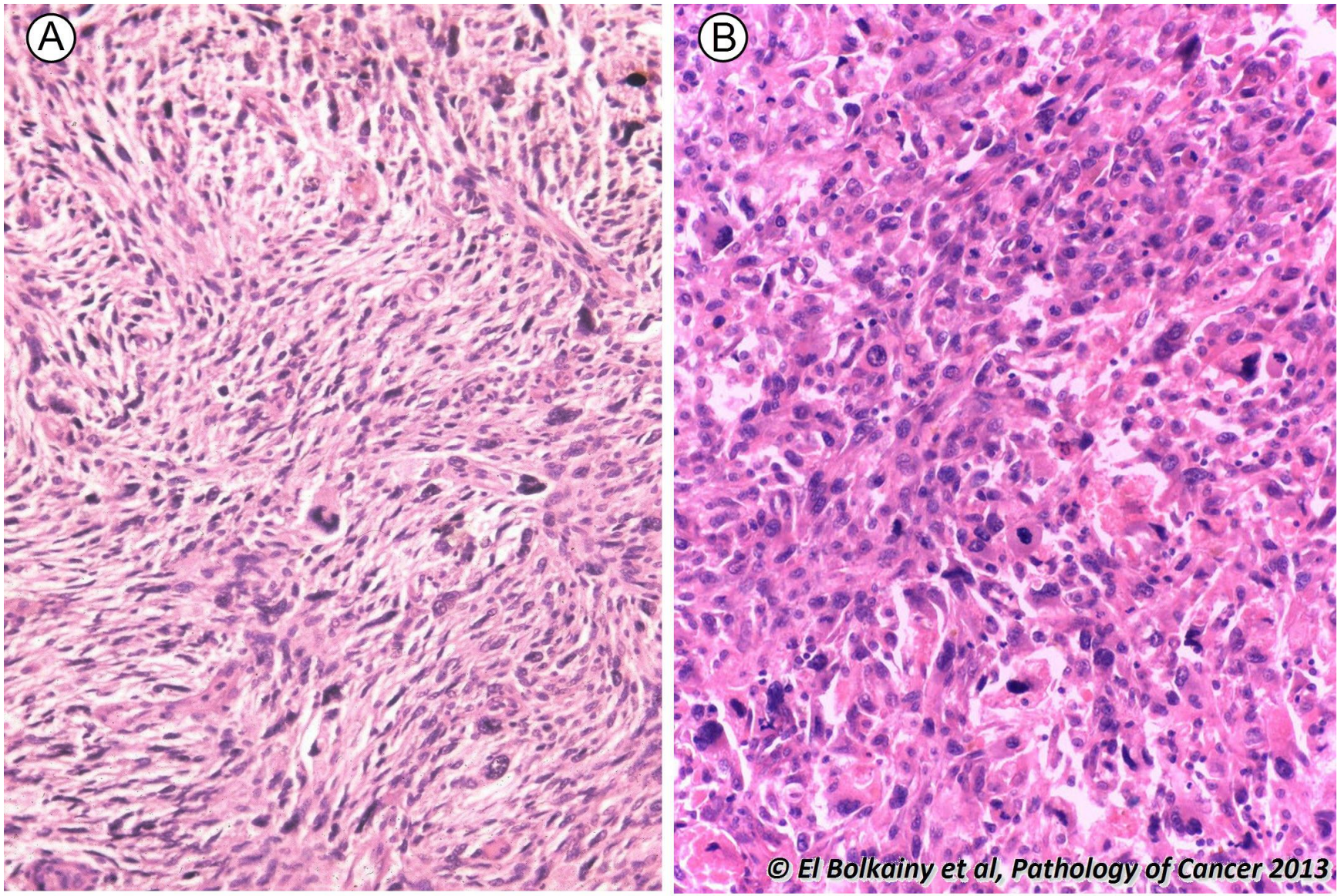
Picture 21-9 **Benign fibrous histiocytoma (dermatofibroma).** It shows bland fibrocytes with storiform pattern. It differs from malignant counterpart by being circumscribed, small sized, located in the lower dermis and lacking giant cells.

21.10 Giant cell tumor of tendon sheath.



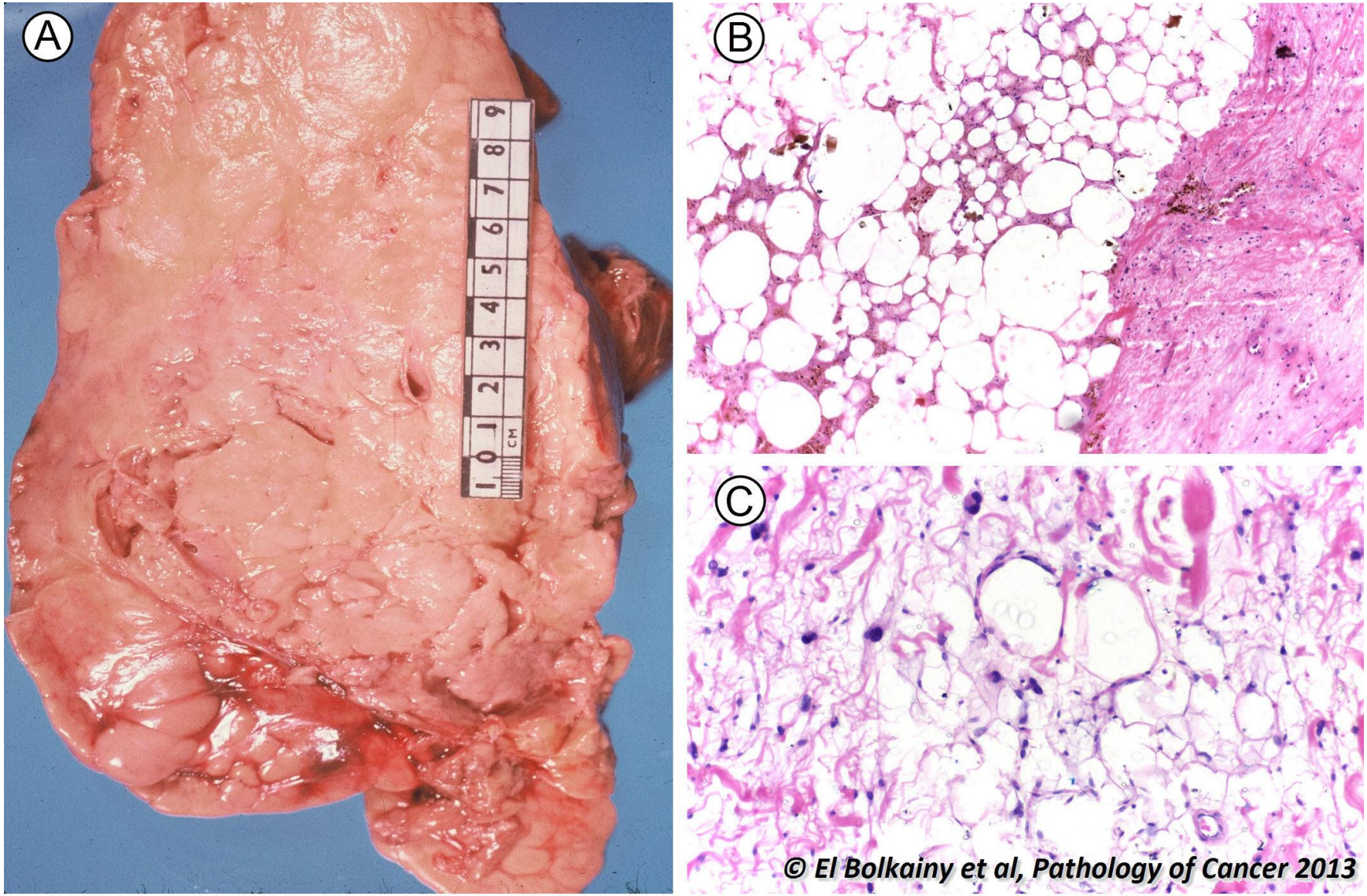
Picture 21-10 **Giant cell tumor of tendon sheath.** The neoplastic cells are small cells which are of fibrohistiocytic nature. The giant cells are reactive in nature. It is subdivided into diffuse and localized types according to the growth patterns.

21.11 Pleomorphic undifferentiated sarcoma/Pleomorphic MFH.



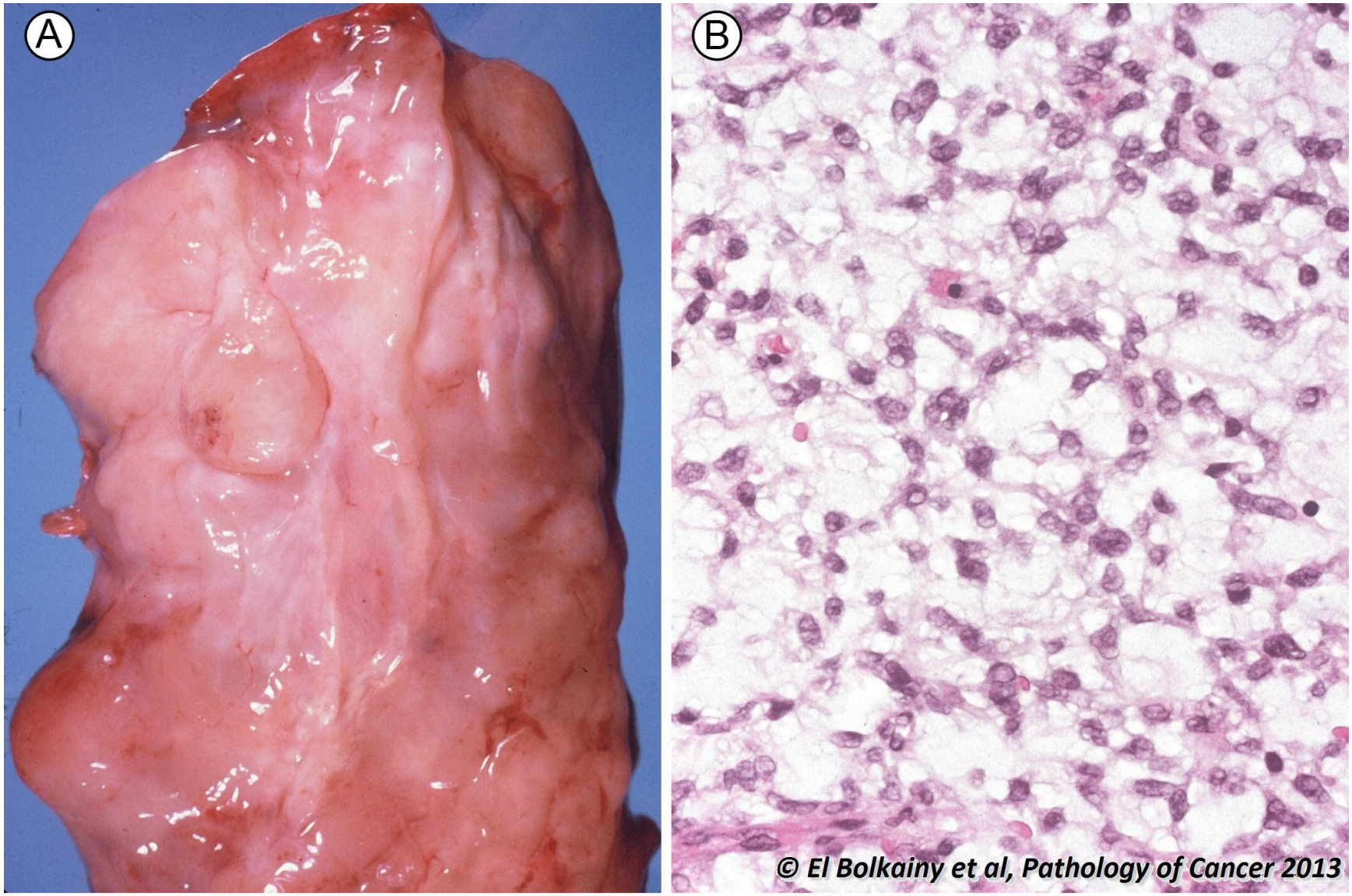
Picture 21-11 **Pleomorphic undifferentiated sarcoma/Pleomorphic MFH. A** Histologicallly, malignant features are evident in both spindle cells and pleomorphic giant cells. **B** The tumor cells show active mitosis. If desmin, actin, or S-100 are positive, the tumor is classified as pleomorphic rhabdomyosarcoma, liposarcoma, leiomyosarcoma respectively. But, if negative, the tumor is diagnosed pleomorphic undifferentiated sarcoma. In addition, CD68 proved to be an unreliable marker, hence plays no role to confirm the diagnosis of pleomorphic sarcomas.

21.12 Atypical lipomatous tumor (well-differentiated liposarcoma).



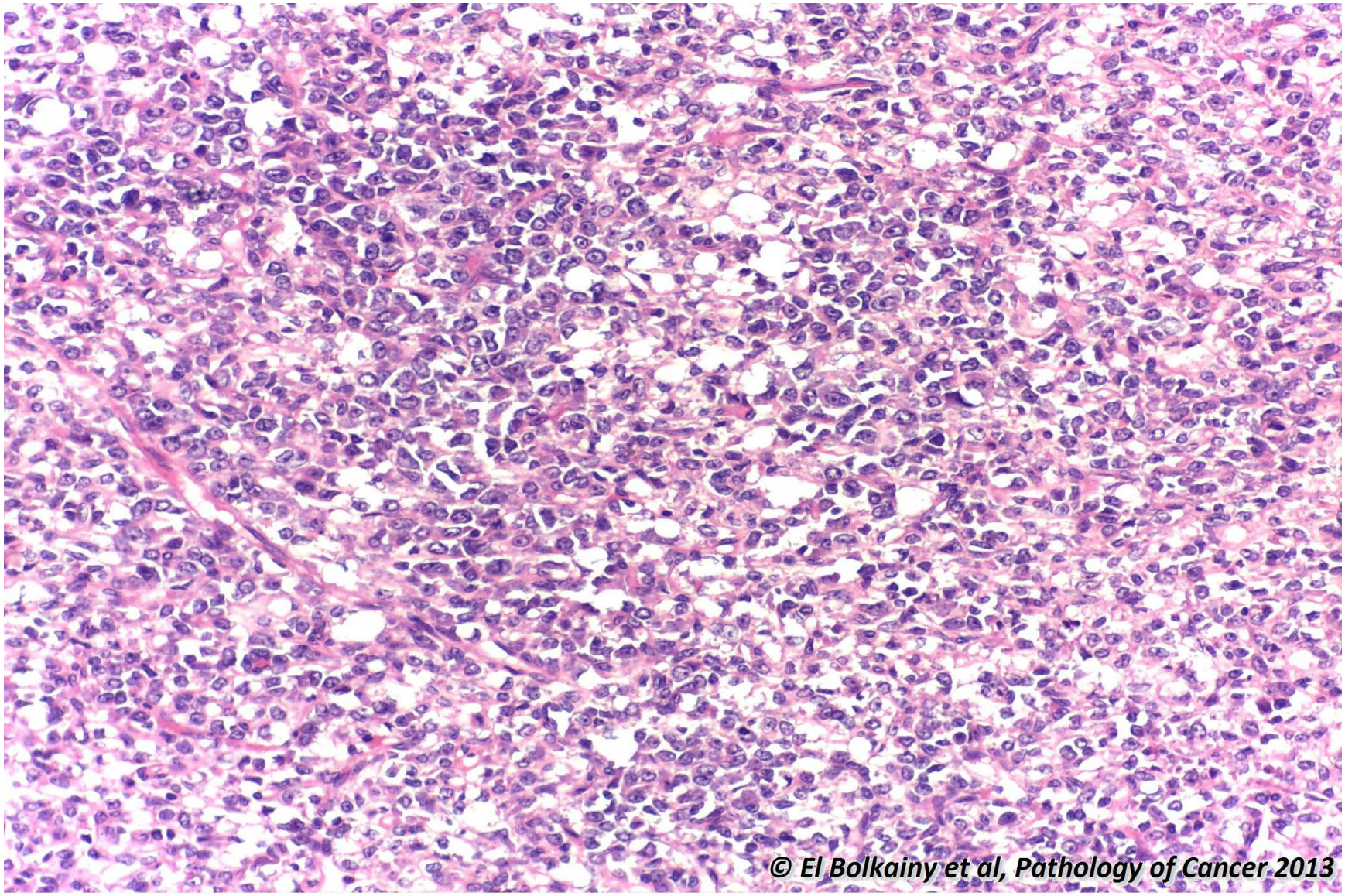
Picture 21-12 **Atypical lipomatous tumor (well-differentiated liposarcoma). A** Grossly, It is a large circumscribed lobulated tumor. **B** and **C** There is marked variation in size of fat cells associated with pleomorphic cells. Lipoblasts exist in fibrovasular trabeculae. Immunoreactive to S-100, MDM-2 and CDK-4.

21.13 Myxoliposarcoma.



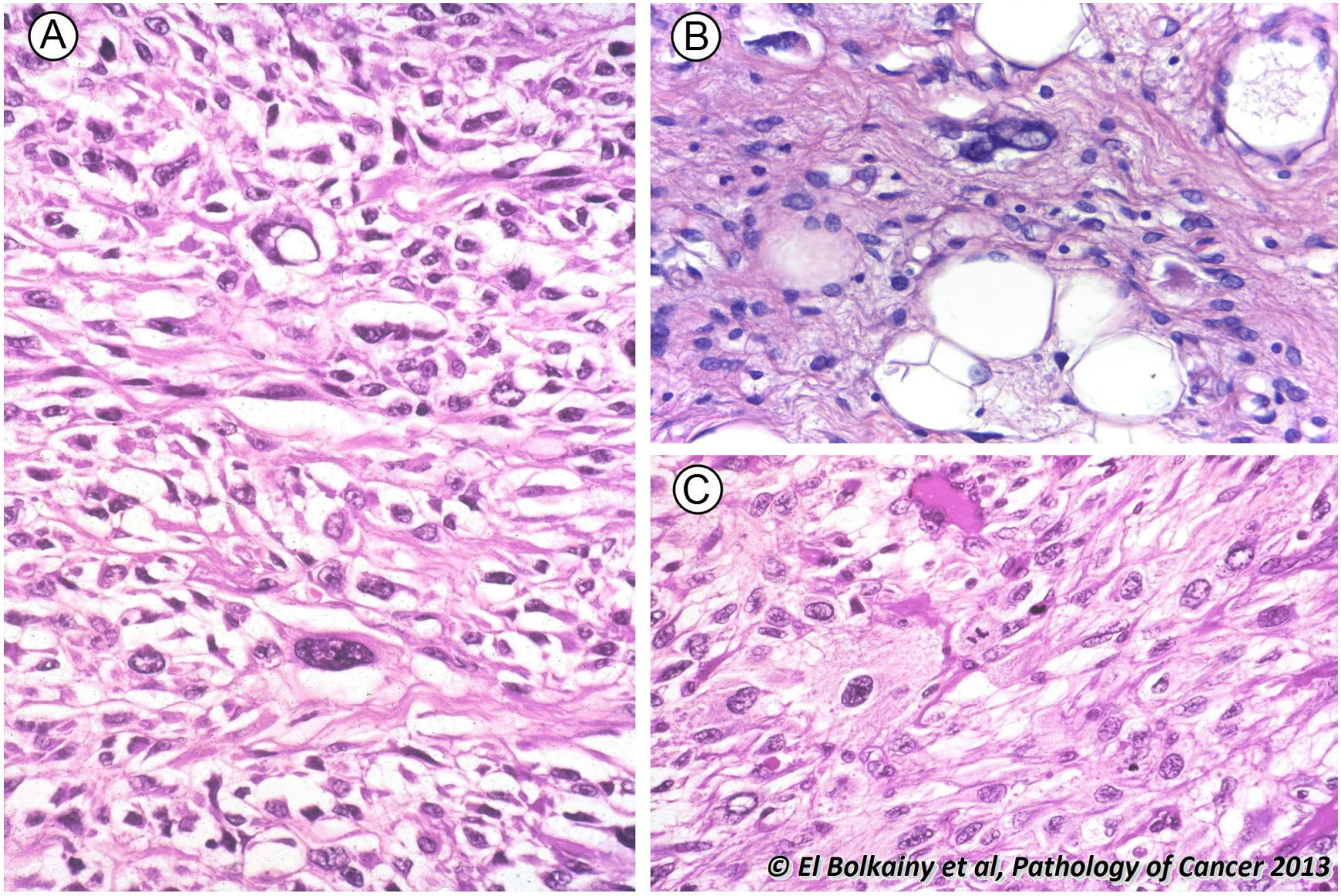
Picture 21-13 **Myxoliposarcoma. A** Grossly, a circumscribed tumor with tan gelatinous cut section. **B** Proliferating spindle cells and lipoblasts (S-100 +) in a myxoid stroma rich in plexiform arborizing blood vessels.

21.14 Round cell liposarcoma.



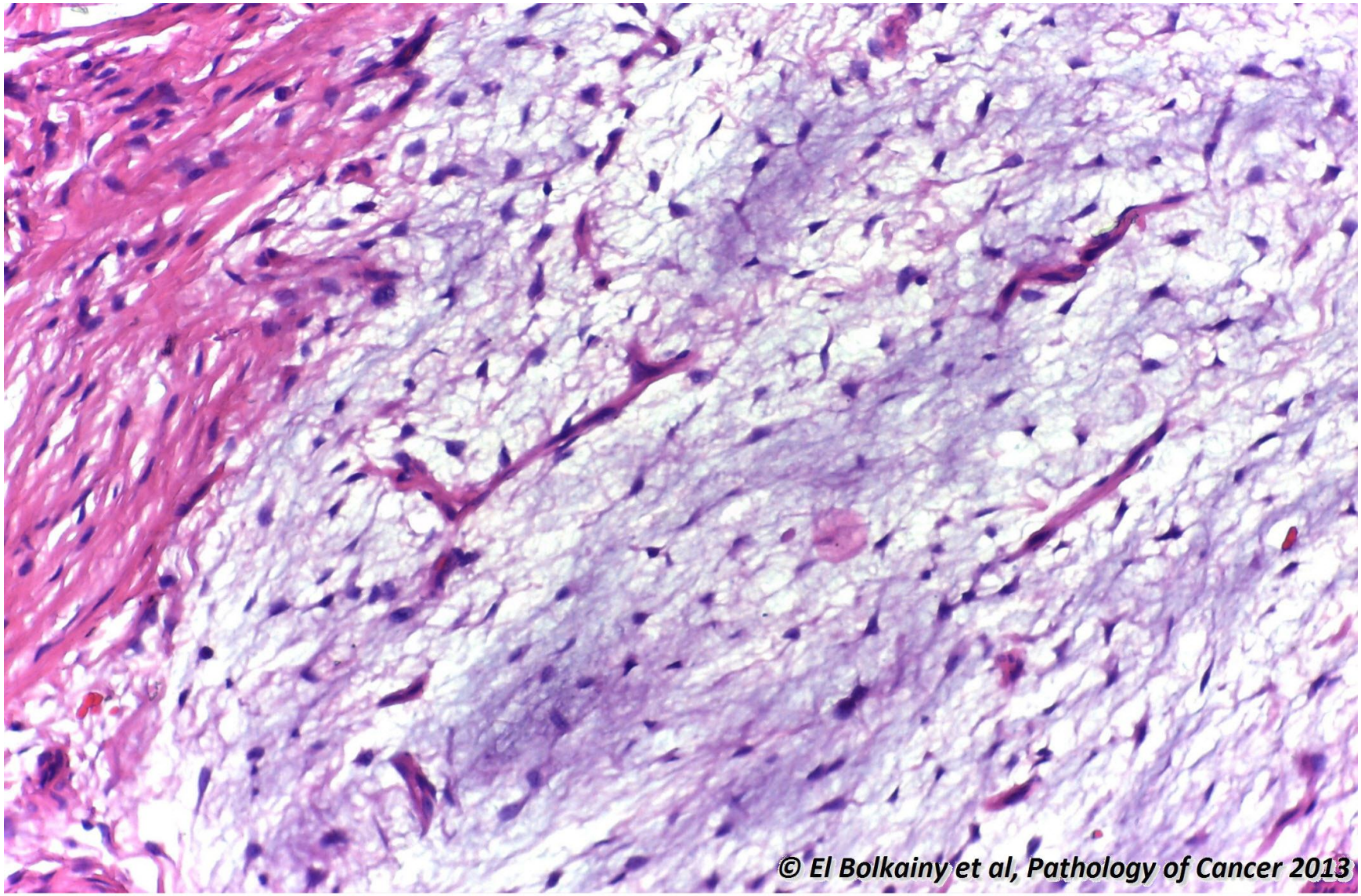
Picture 21-14 **Round cell liposarcoma.** The diagnostic criteria of this subtype is the presence of S-100 positive round cells in > 80 % of tumor. It may be novo or the result of dedifferentiation of a low grade liposarcoma.

21.15 Pleomorphic liposarcoma.



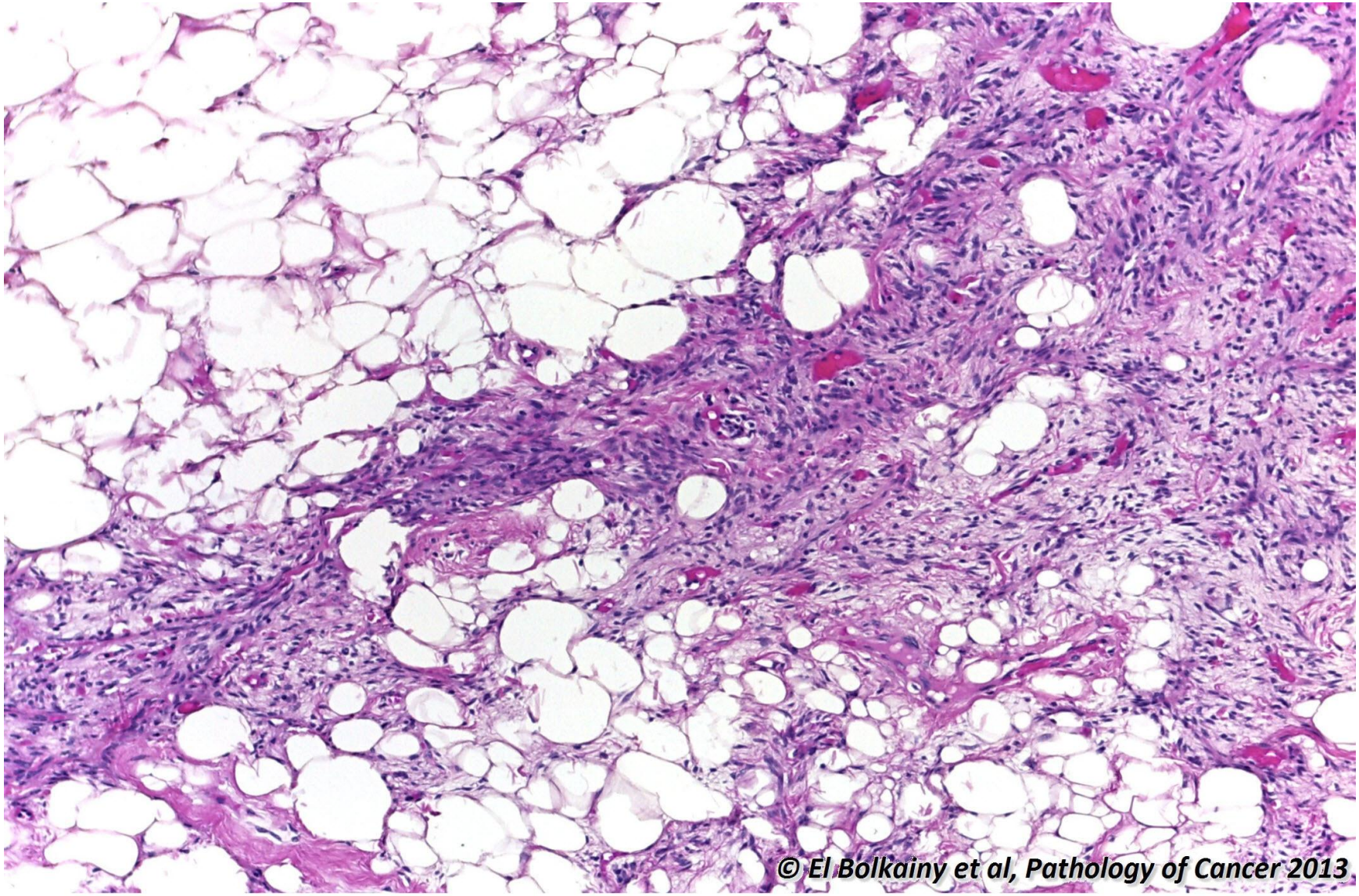
Picture 21-15 **Pleomorphic liposarcoma. A, B** and **C.** It is composed of a mixture of spindle, round and pleomorphic giant cells with multiple cytoplasmic vacuoles, all are positive for S-100.

21.16 Lipoblastoma.



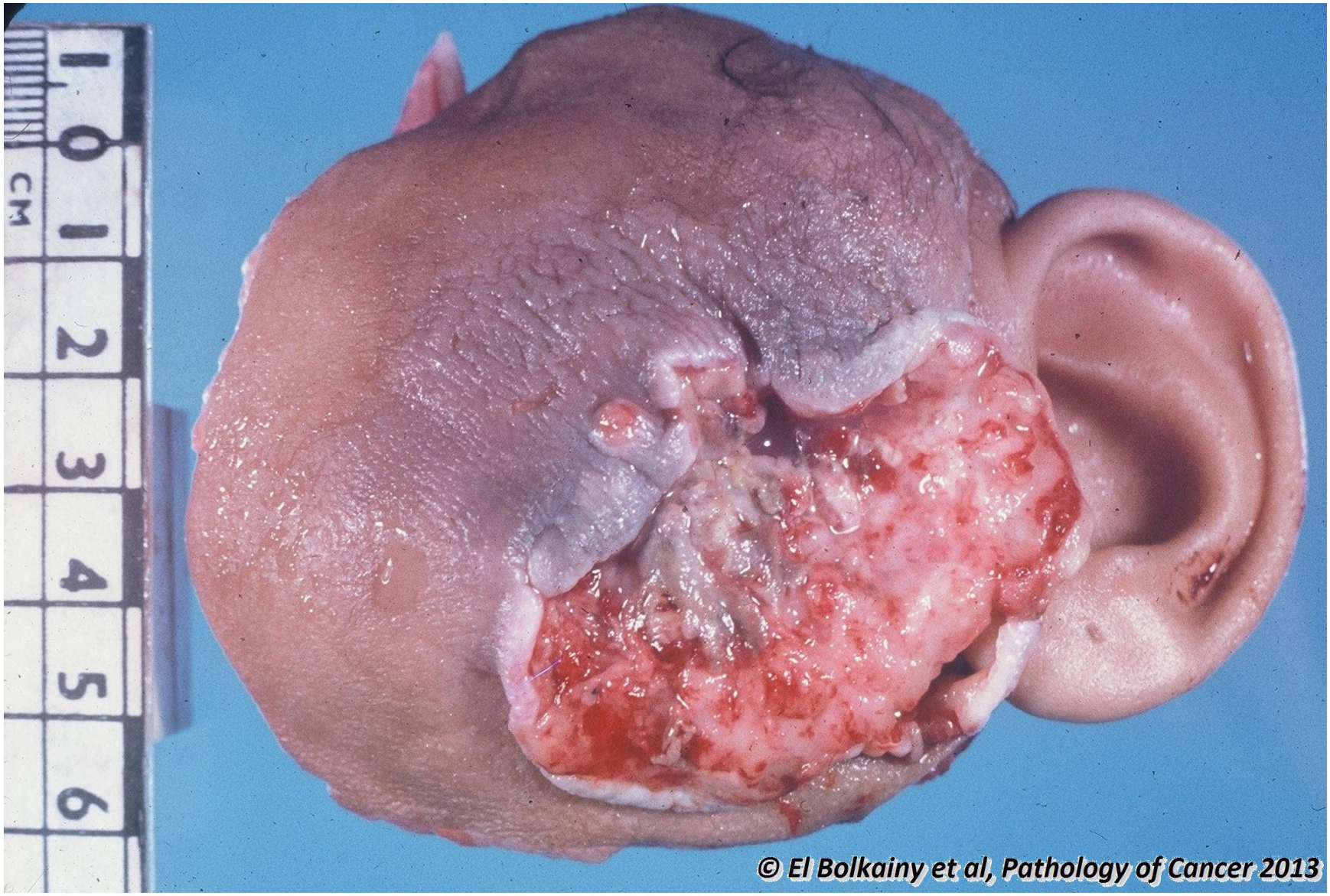
Picture 21-16 **Lipoblastoma.** A benign tumor of infancy (<3 years) composed of arborizing blood vessels, simulating myxoliposarcoma, but the age and lobulated pattern are characteristic.

21.17 Spindle cell / pleomorphic lipoma.



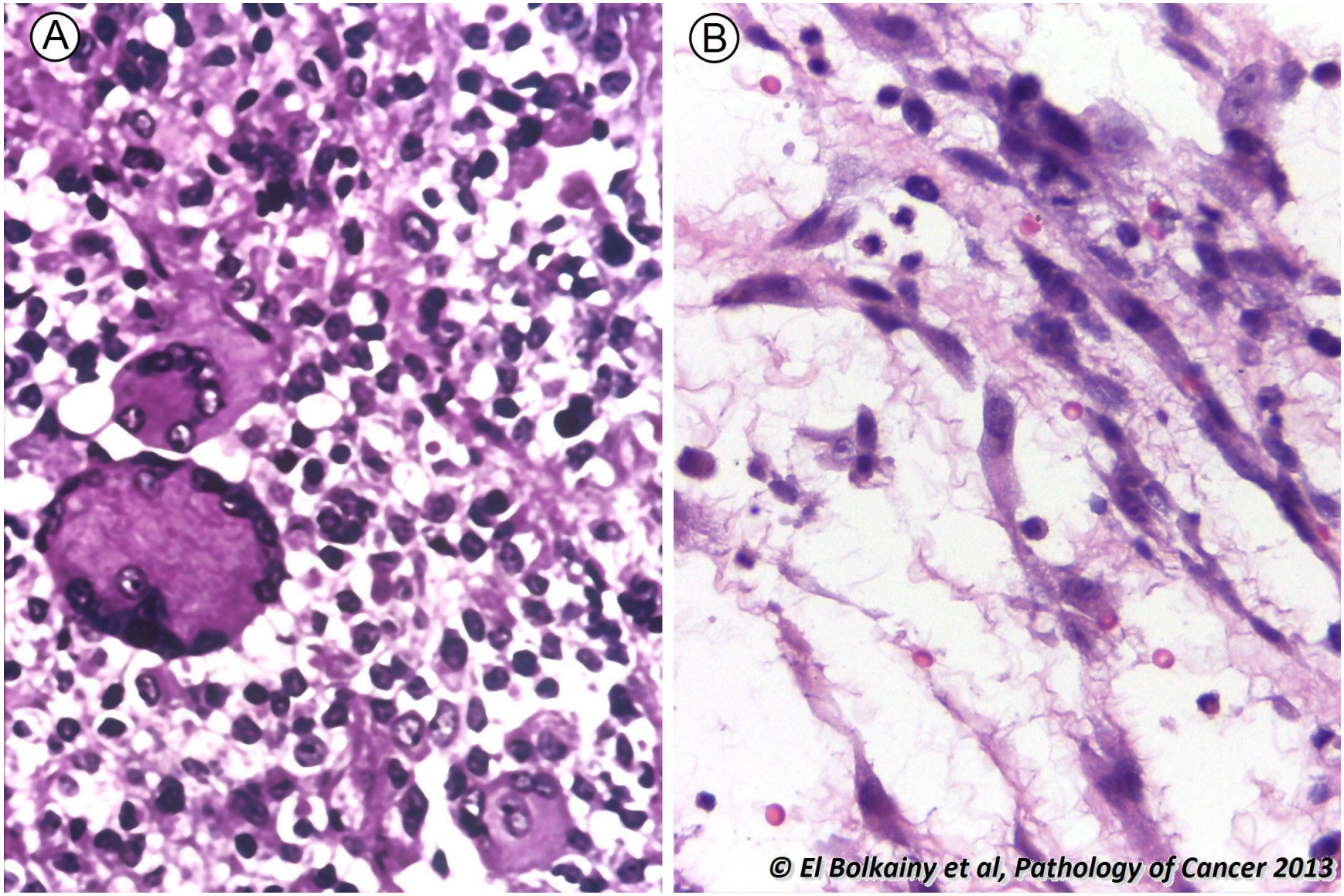
Picture 21-17 **Spindle cell/ pleomorphic lipoma.** A benign tumor of adults showing an admixture of spindle cells, collagen, myxoid areas and multinucleated floret-like giant cells.

21.18 Embryonal rhabdomyosarcoma, gross appearance.



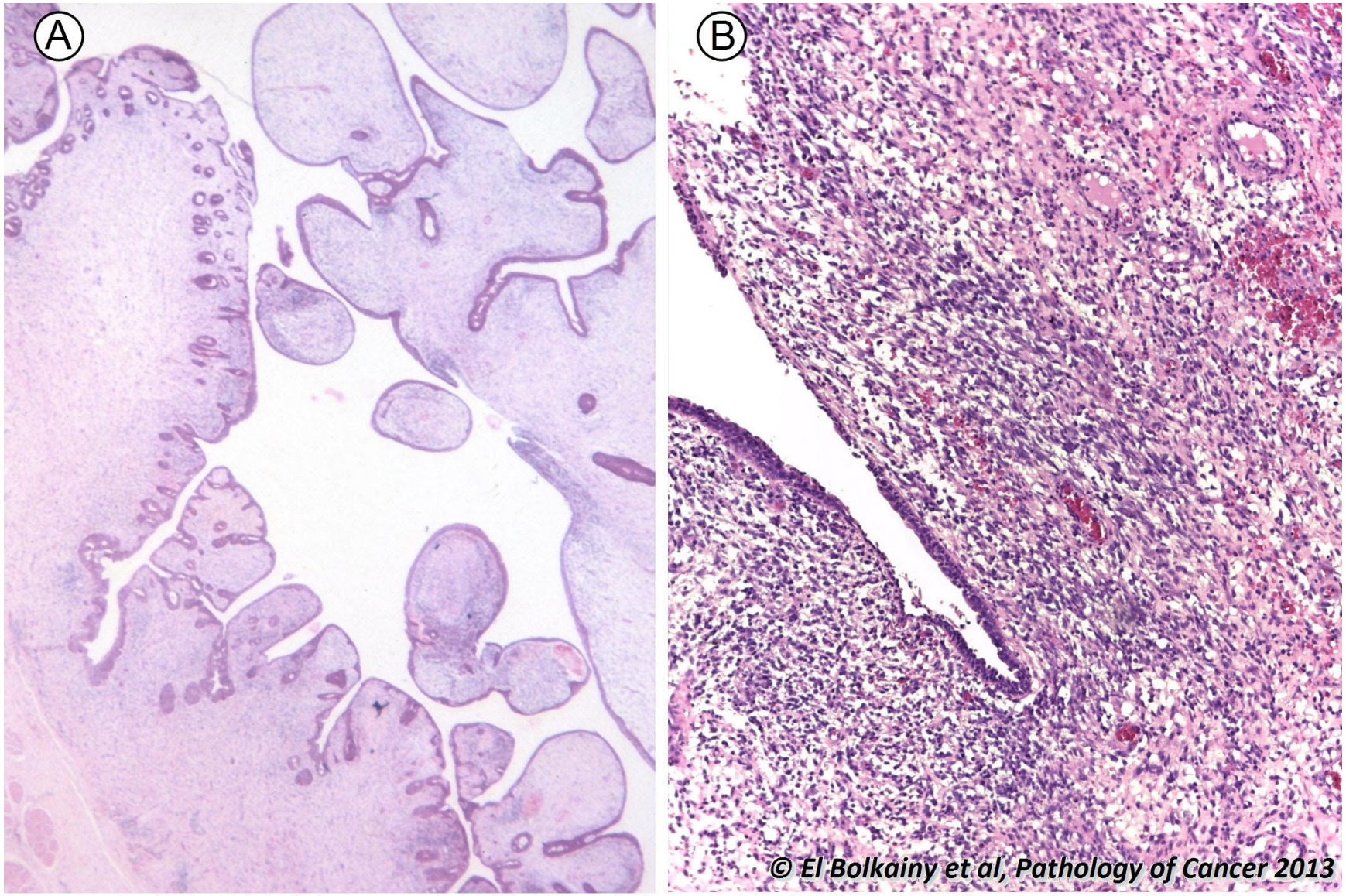
Picture 21-18 **Embryonal rhabdomyosarcoma, gross appearance.** An ulcerating mass in the pre-auricular region with necrotic hemorrhagic floor.

21.19 Embryonal rhabdomyosarcoma, histology.



Picture 21-19 **Embryonal rhabdomyosarcoma, histology. A** and **B** The various cell shapes include: spindle, stellate, strap cells and pleomorphic giant cells. The cells are immunoreactive to: desmin, myogenin and myo D-1.

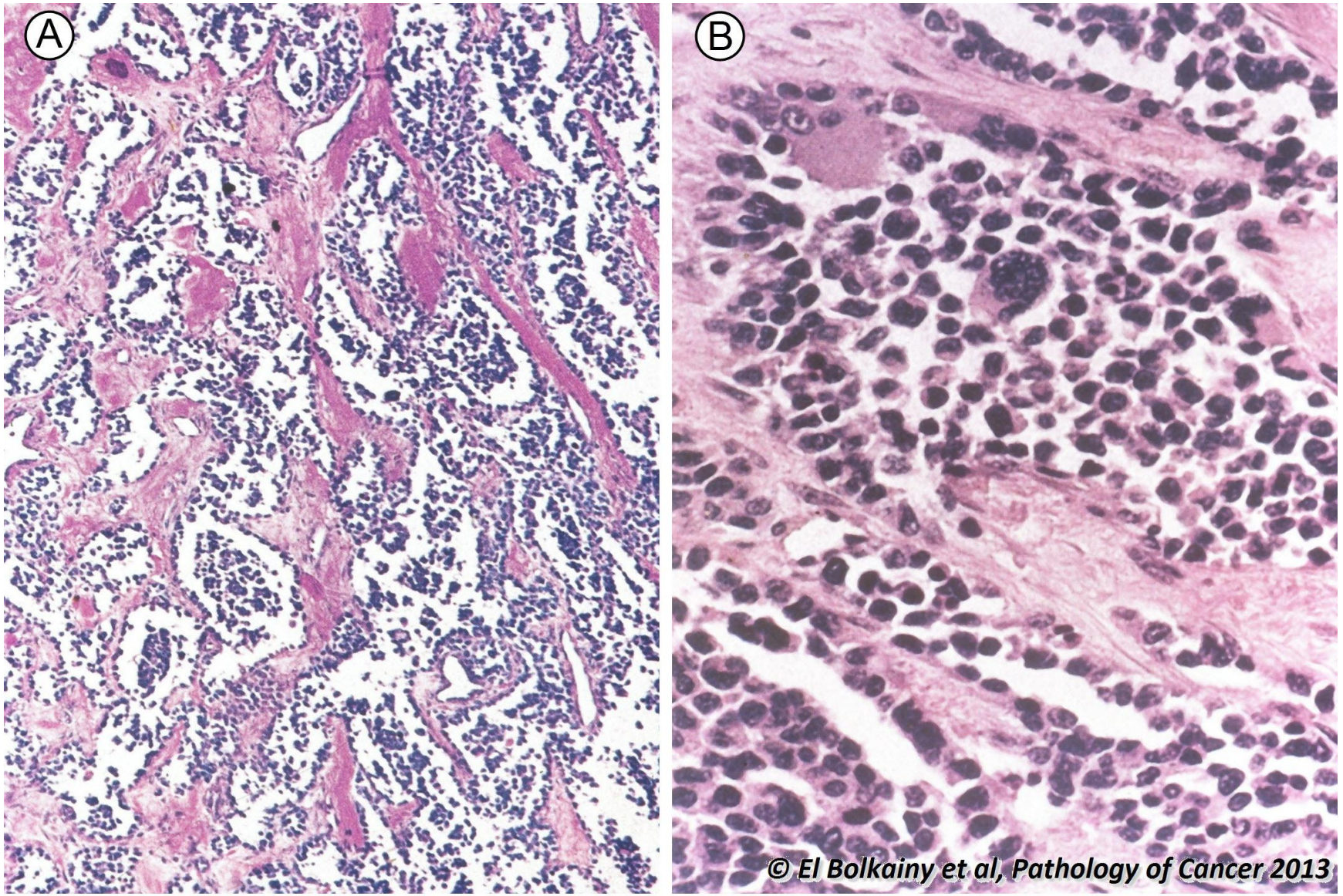
21.20 Botryoid rhabdomyosarcoma, histology.



Picture 21-20

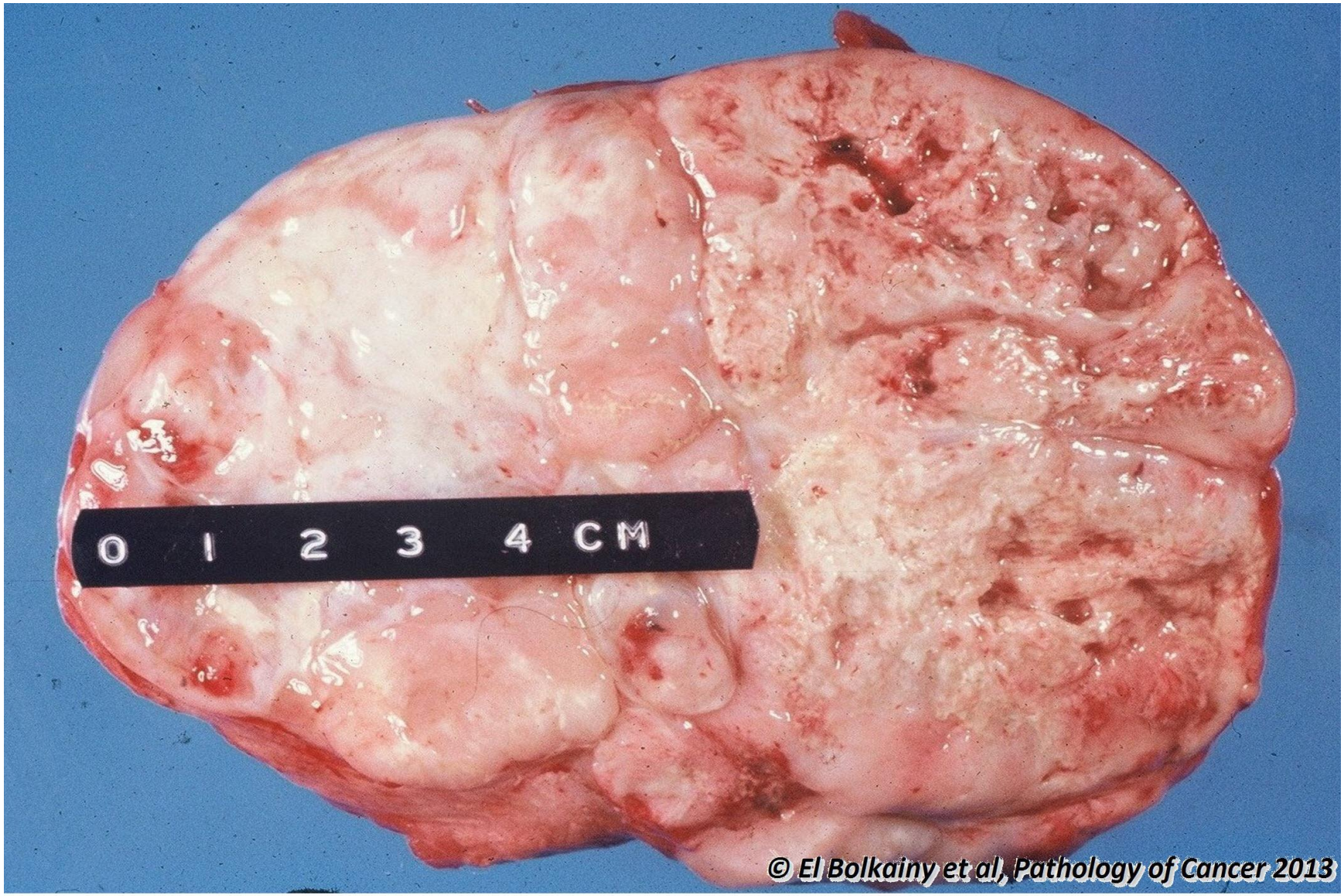
Botryoid rhabdomyosarcoma, histology. A Tumors arising in relation to cavities develop a multipolypoid grape-like structures. **B** There is concentration of tumor cells under surface epithelium (cambium layer), whereas, the rest of stroma is hypocellular.

21.21 Alveolar rhabdomyosarcoma, histology.



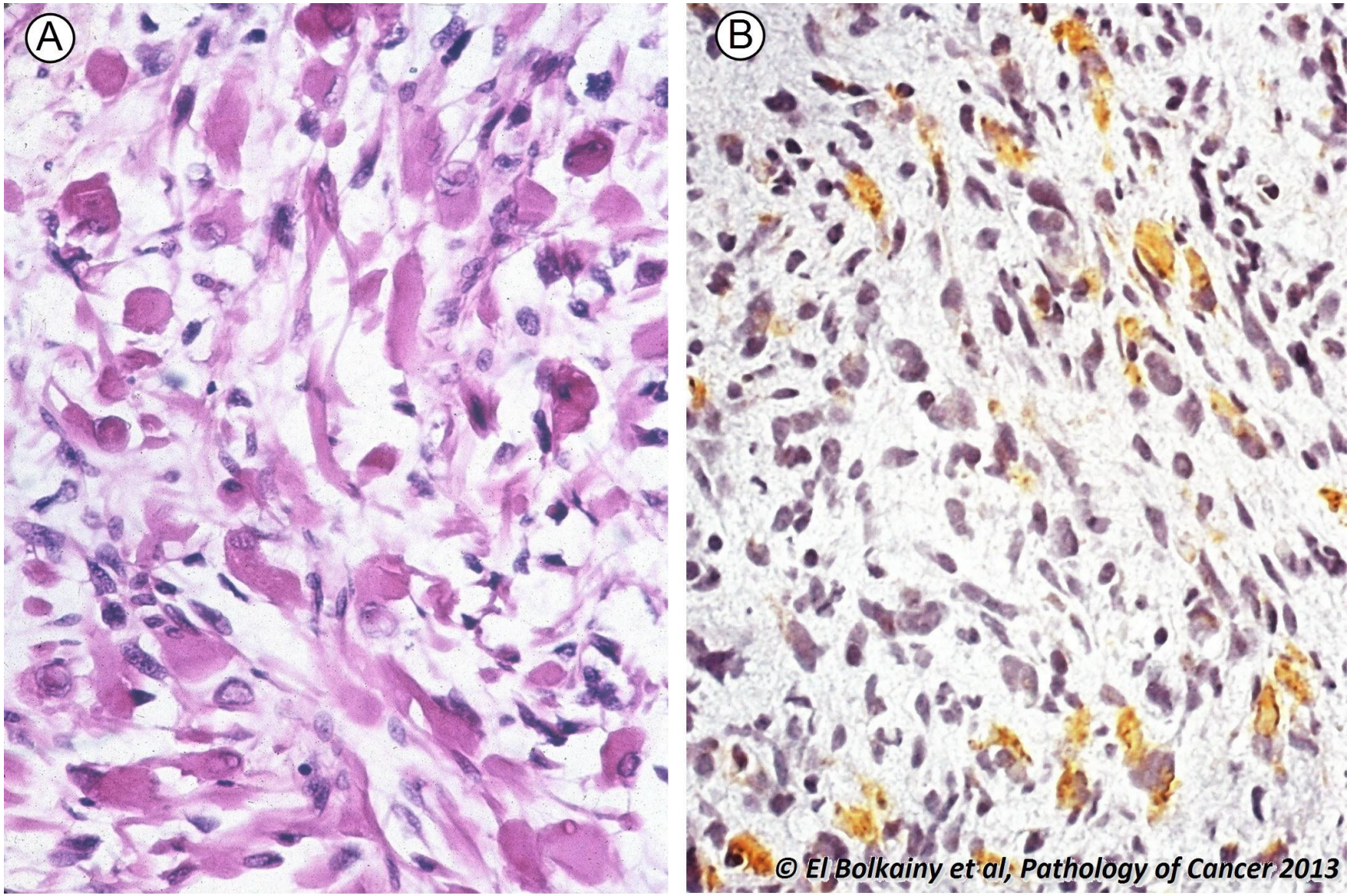
Picture 21-21 **Alveolar rhabdomyosarcoma, histology.** This unfavorable subtype is characterized by nests of very small round discohesive tumor cells (desmin positive) in abundant fibrotic stroma. **A** Low power. **B** High power.

21.22 Pleomorphic rhabdomyosarcoma, gross appearance.



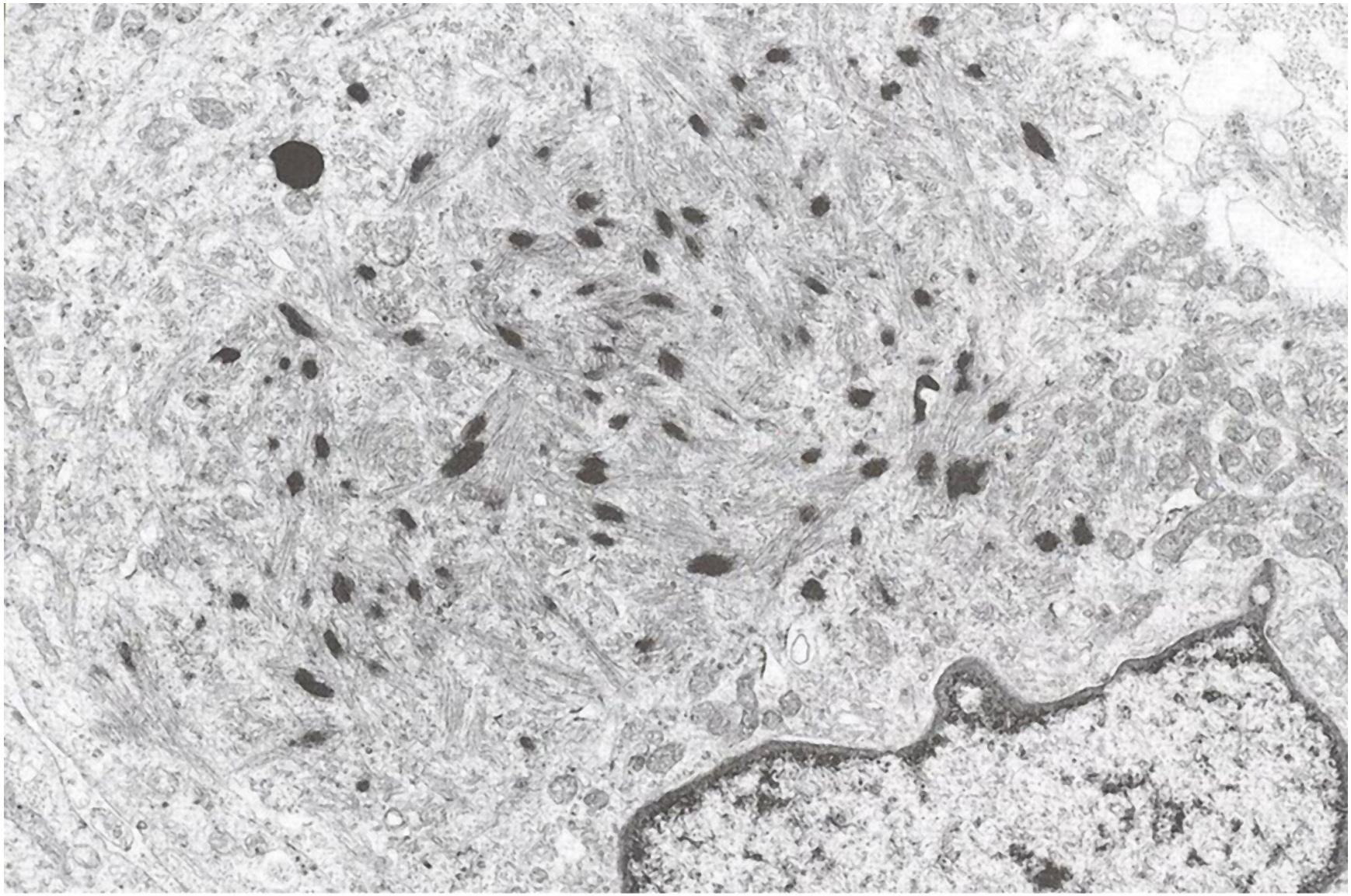
Picture 21-22 **Pleomorphic rhabdomyosarcom, gross appearance.** A bulky tumor mass showing variablity in color with grey, whitish, yellow (necrosis), and reddish areas (hemorrhage). The tumor is surrounded by a very thin pseudocapsule of compressed normal tissue.

21.23 Pleomorphic rhabdomyosarcoma, histology.



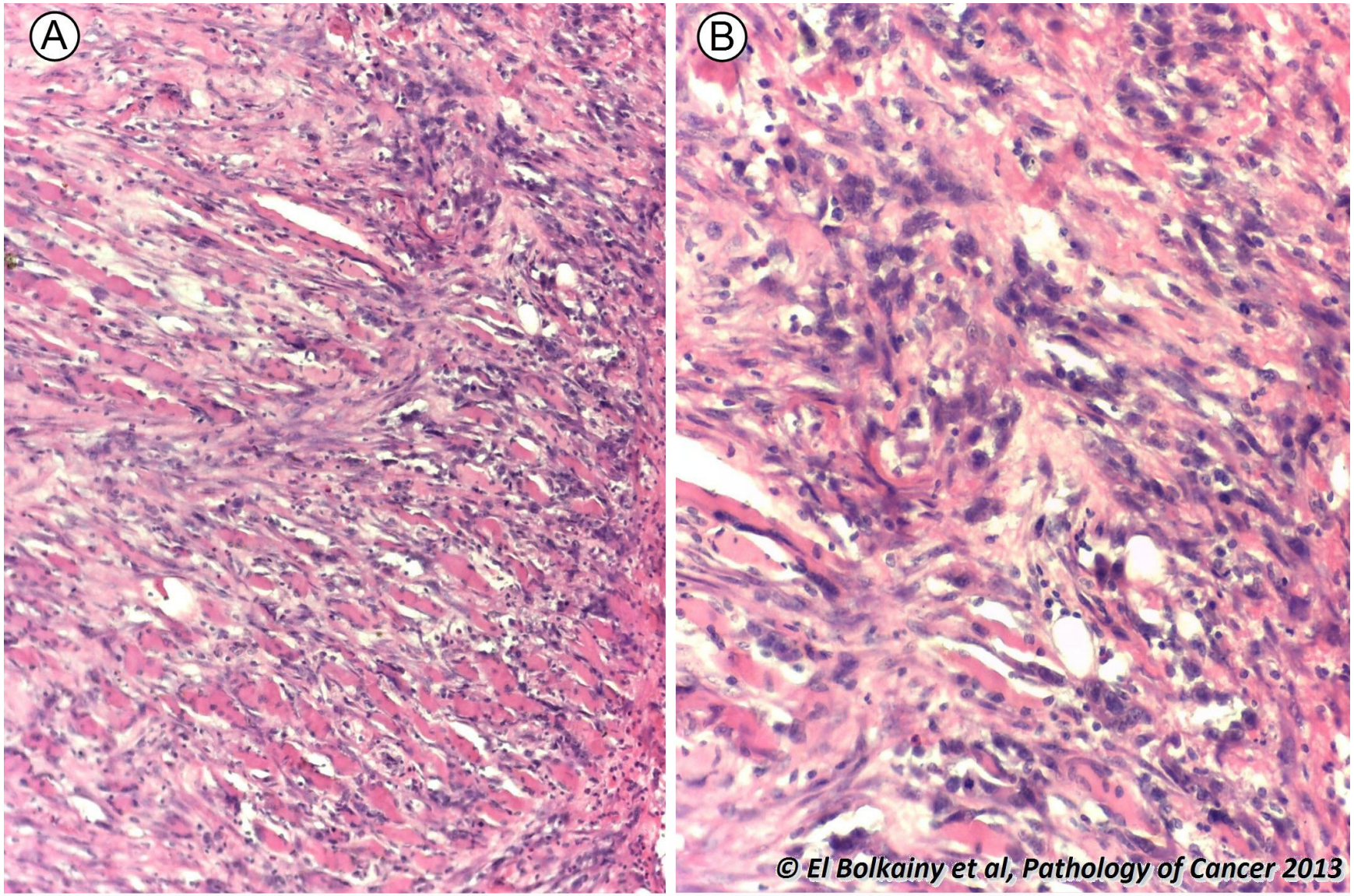
Picture 21-23 **Pleomorphic rhabdomyosarcoma, histology. A** Strap and pleomorphic cells. **B** Positivity to desmin confirms the diagnosis and distinguishes it from other pleomorphic malignant tumors.

21.24 Rhabdomyosarcoma, electron microscopic features.



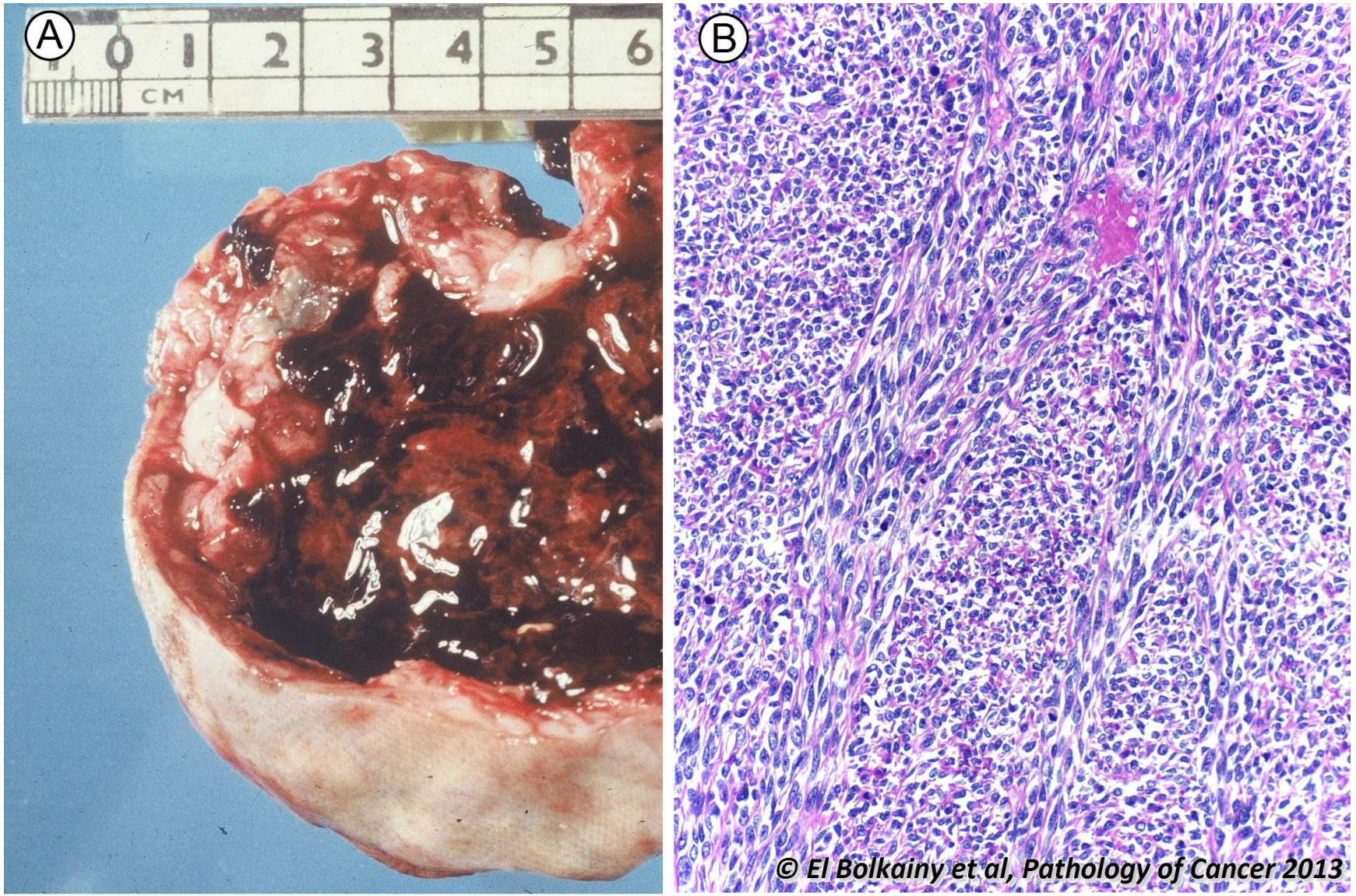
Picture 21-24 **Rhabdomyosarcoma, electron microscopic features.** Rhabdomyoblastic differentiation is characterized by dense Z-band material in the center of myofibrils. (Reproduced with permission, Fletcher CD, 2007).

21.25 Proliferative myositis.



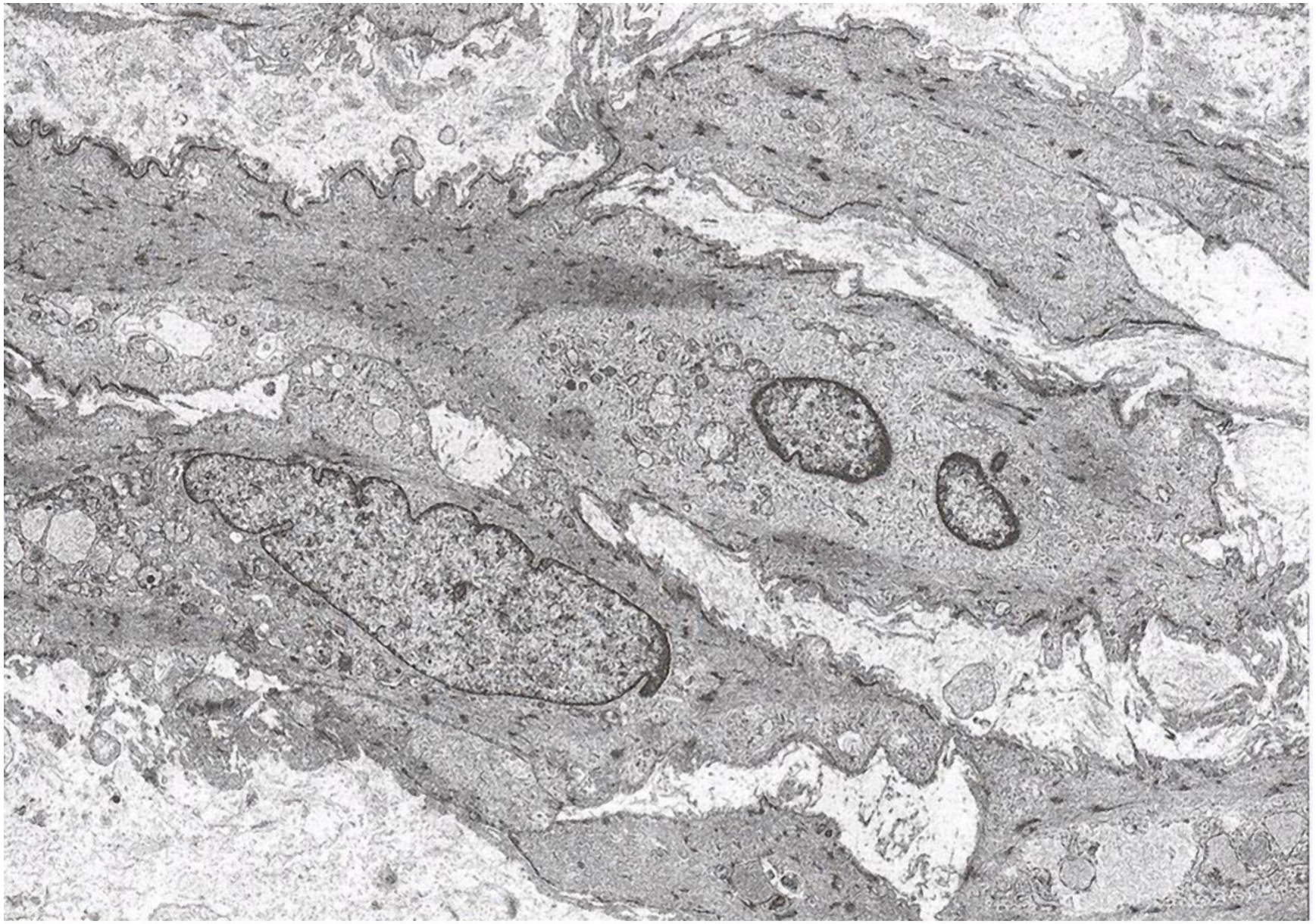
Picture 21-25 **Proliferative myositis. A** Low power. **B** High power. Many spindle cells (fibroblasts and myofibroblasts) are evident inbetween skeletal muscles. Focal inflammation and ganglion-like cells may be seen (not shown).

21.26 Leiomyosarcoma.



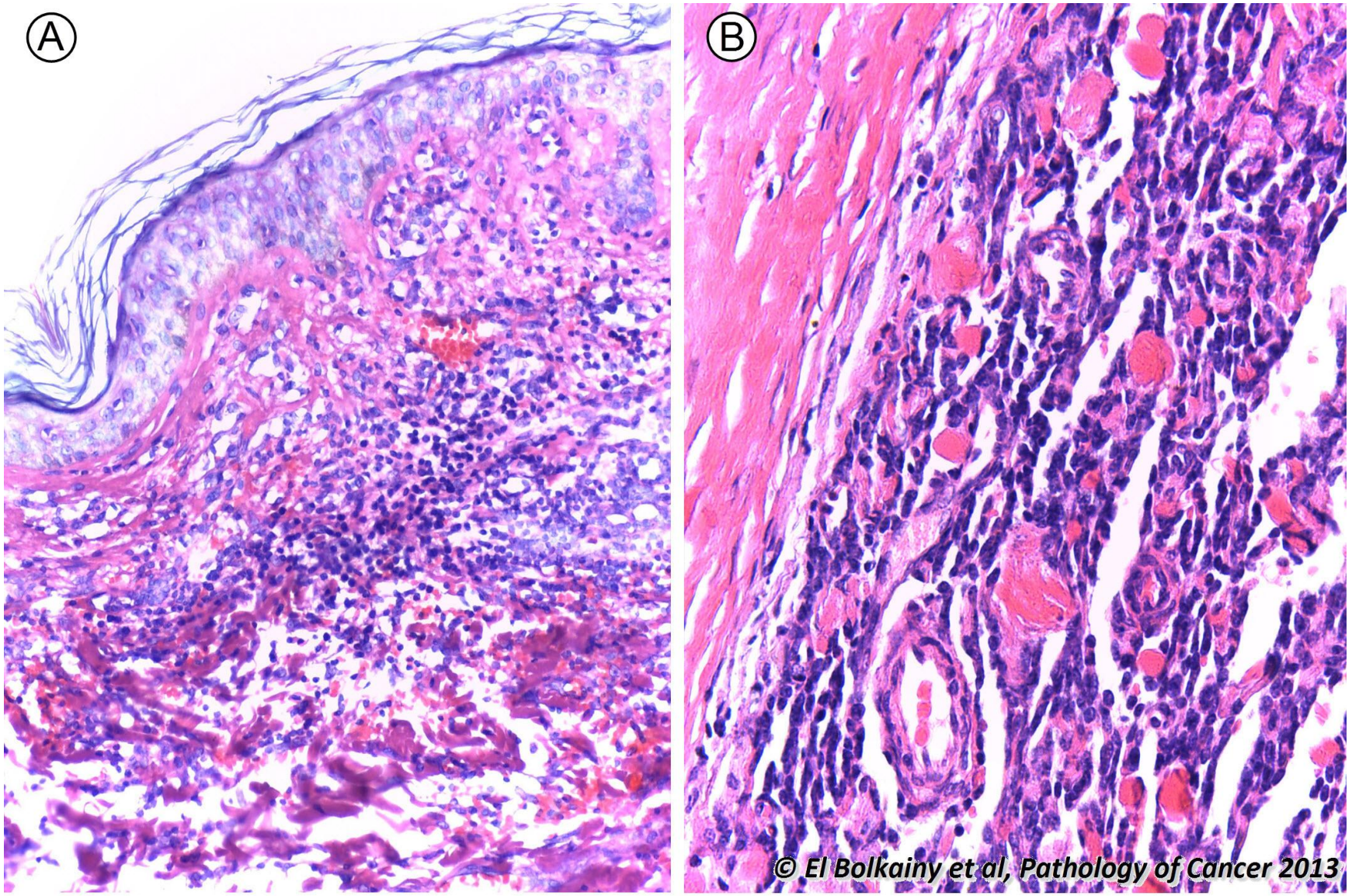
Picture 21-26 **Leiomyosarcoma. A** Retroperitoneal leiomyosarcoma, gross appearance. It shows marked cystic degeneration with hemorrhage. **B** Histology, It is composed of spindle cells with blunt-ended nuclei and single paranuclear vacuoles (actin positive). The bundles cross each other at right angle (interdigitating pattern).

21.27 Leiomyosarcoma, electron microscopic features.



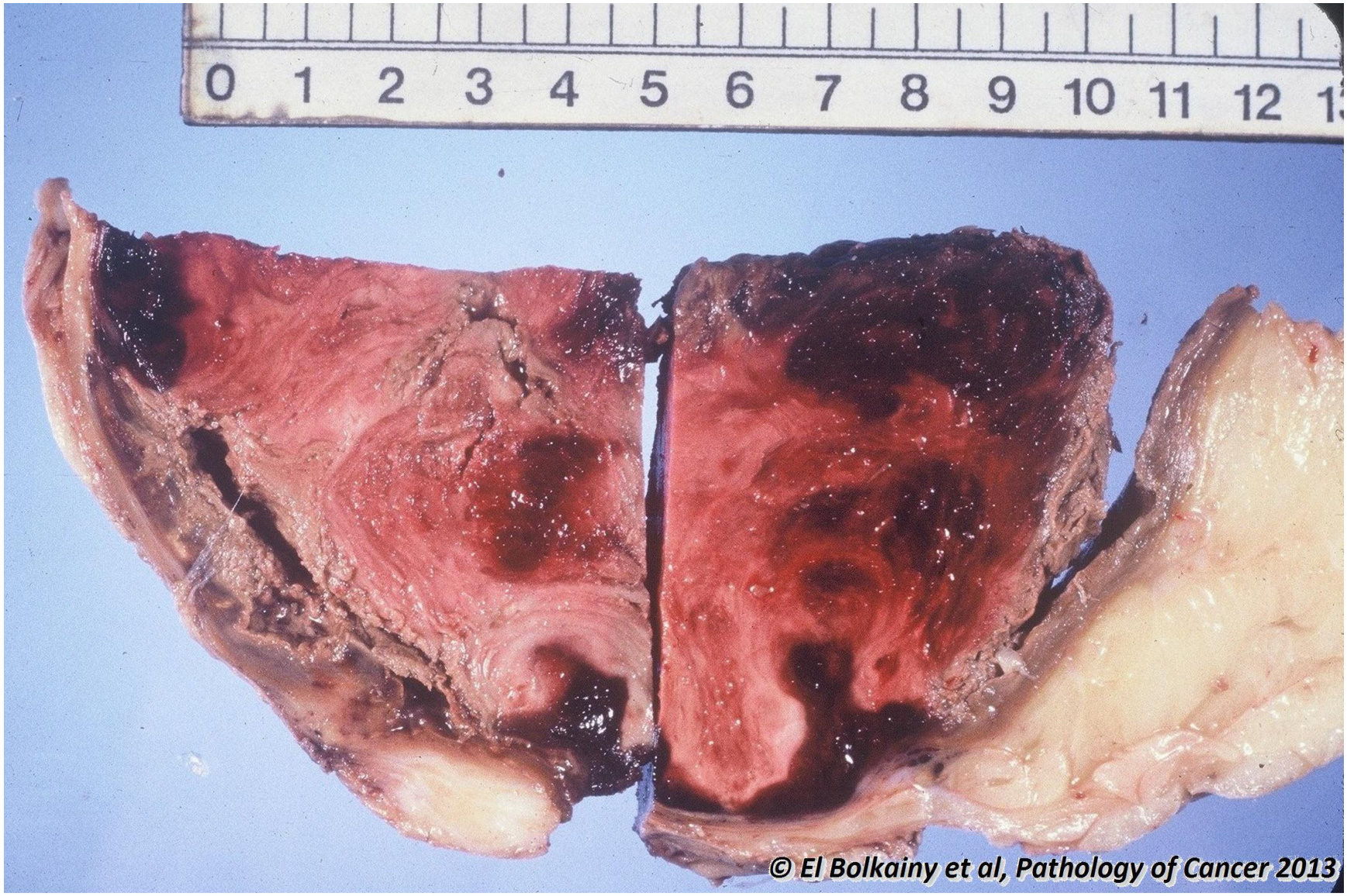
Picture 21-27 **Leiomyosarcoma, electron microscopic features.** Myofilaments are evident in cytoplasm and surface contacts between the cells. (Reproduced with permission, Fletcher CD, 2007).

21.28 Hemangioendothelioma.



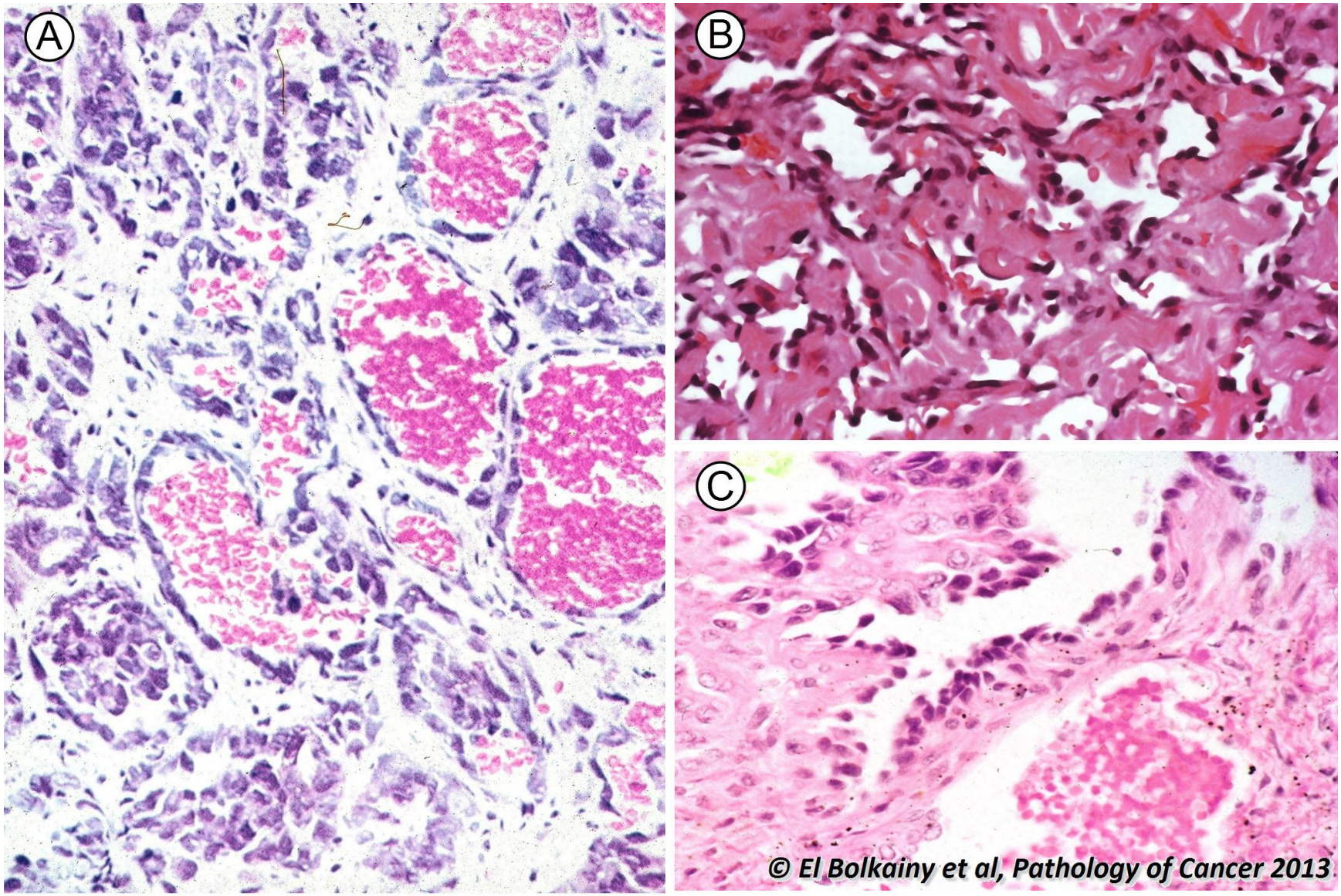
Picture 21-28 **Hemangioendothelioma. A** Proliferation of endothelium is confined to vessel lining and lumen (CD 34 positive). The three variants are: Kaposiform variant (cellular), pediatric Dabska type (papillary) and epithelioid variant (bland cells) which may complicate radiotharapy. **B** High power.

21.29 Angiosarcoma, gross features.



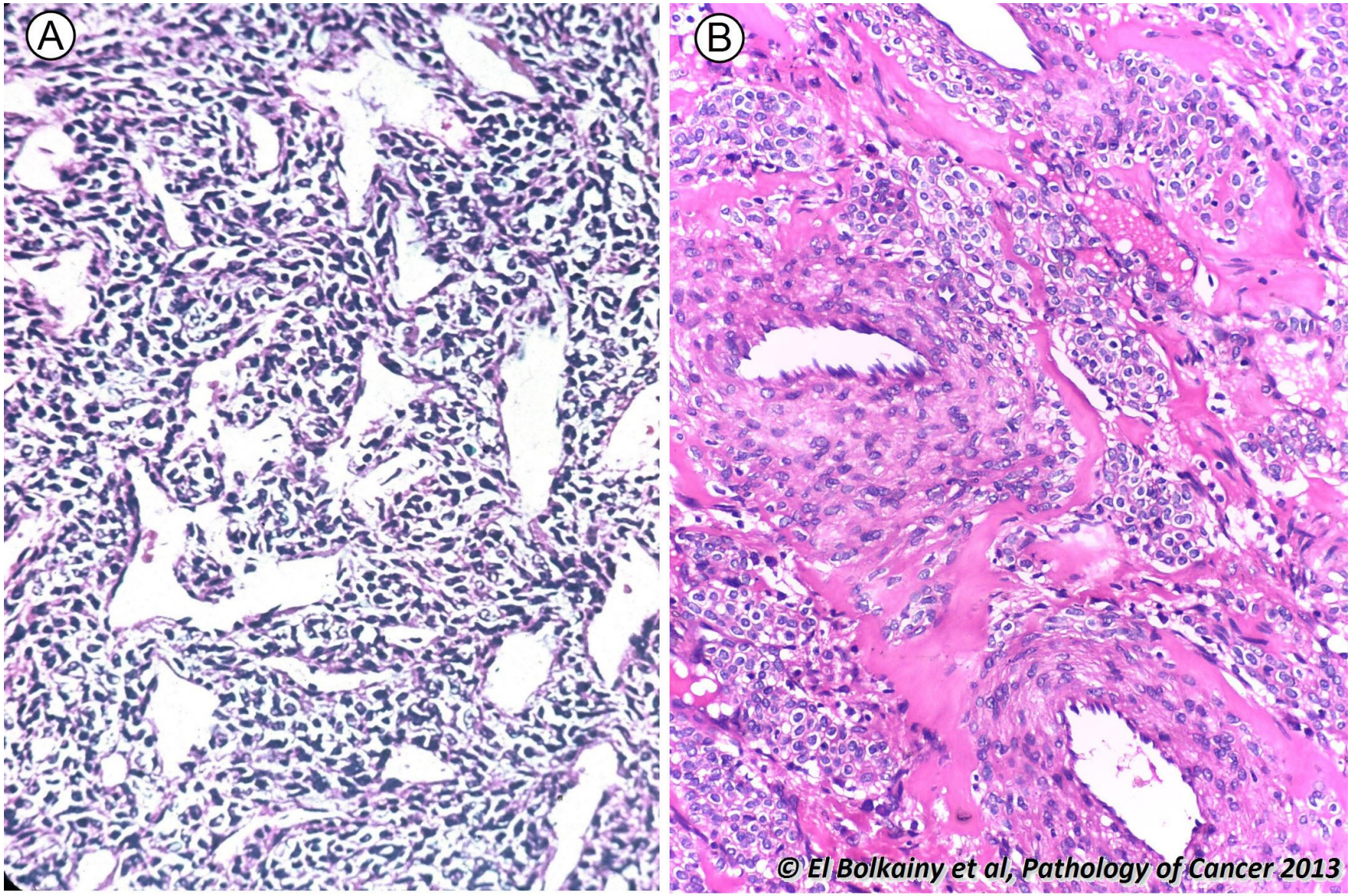
Picture 21-29 **Angiosarcoma, gross features.** It appears as a hemorrhagic soft mass lesion which may be misdiagnosed as hematoma.

21.30 Angiosarcoma, histology.



Picture 21-30 **Angiosarcoma, histology. A** Irregular vascular spaces lined by anaplastic pleomorphic endothelium, with active mitosis. **B** and **C** Seive-like and diffuse patterns are common, as well as, associated marked hemorrhage and necrosis.

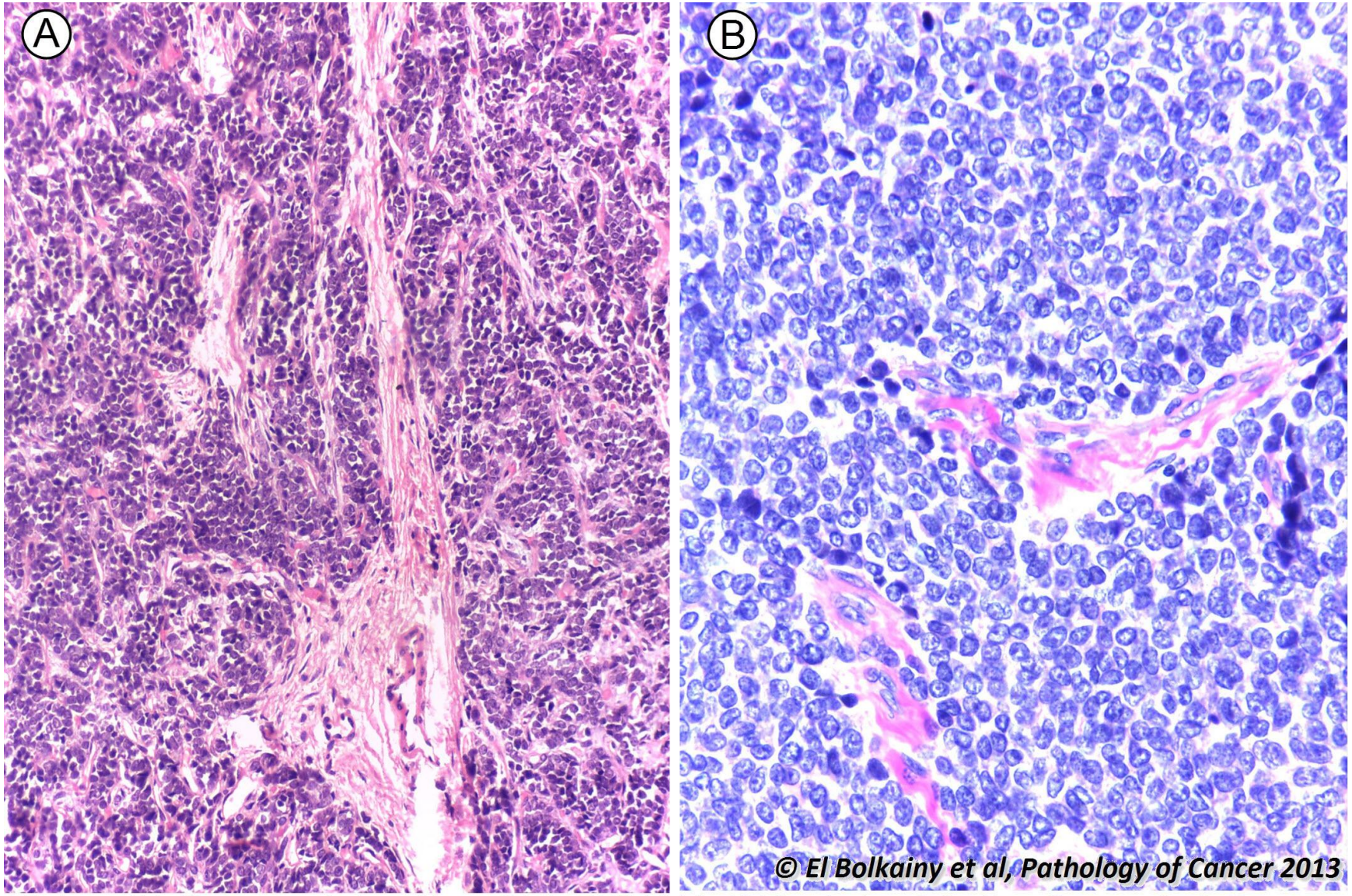
21.31 Perivascular tumors.



Picture 21-31

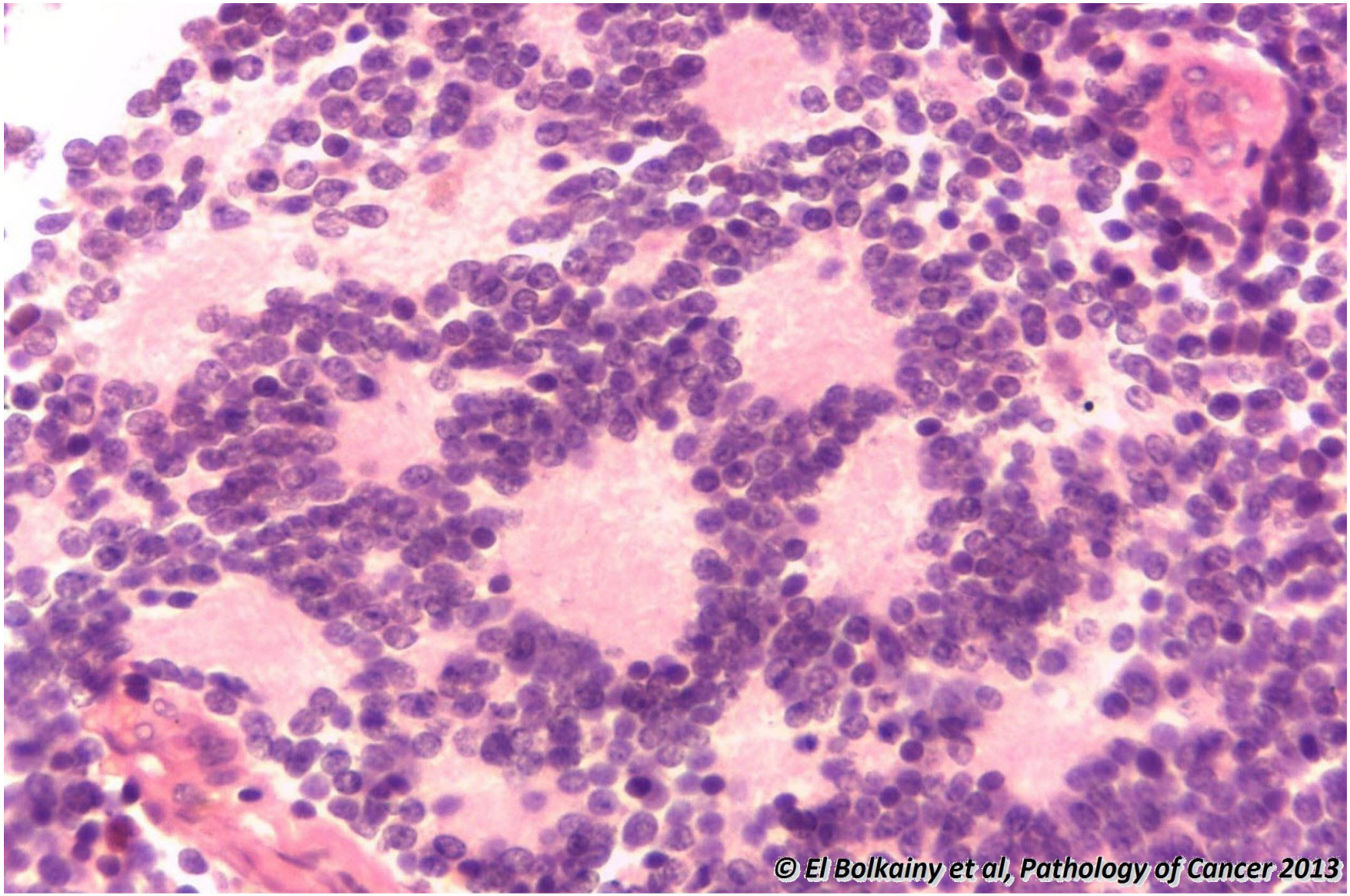
Perivascular tumors. A Hemangiopericytoma is characterized by actin negative spindle cells around thin-walled branching vessels (staghorn shape). **B** Glomus tumor is composed of actin positive round cells around thick-walled non-branching hyalinized vessels.

21.32 Primitive neuroectodermal tumor (Ewing / PNET).



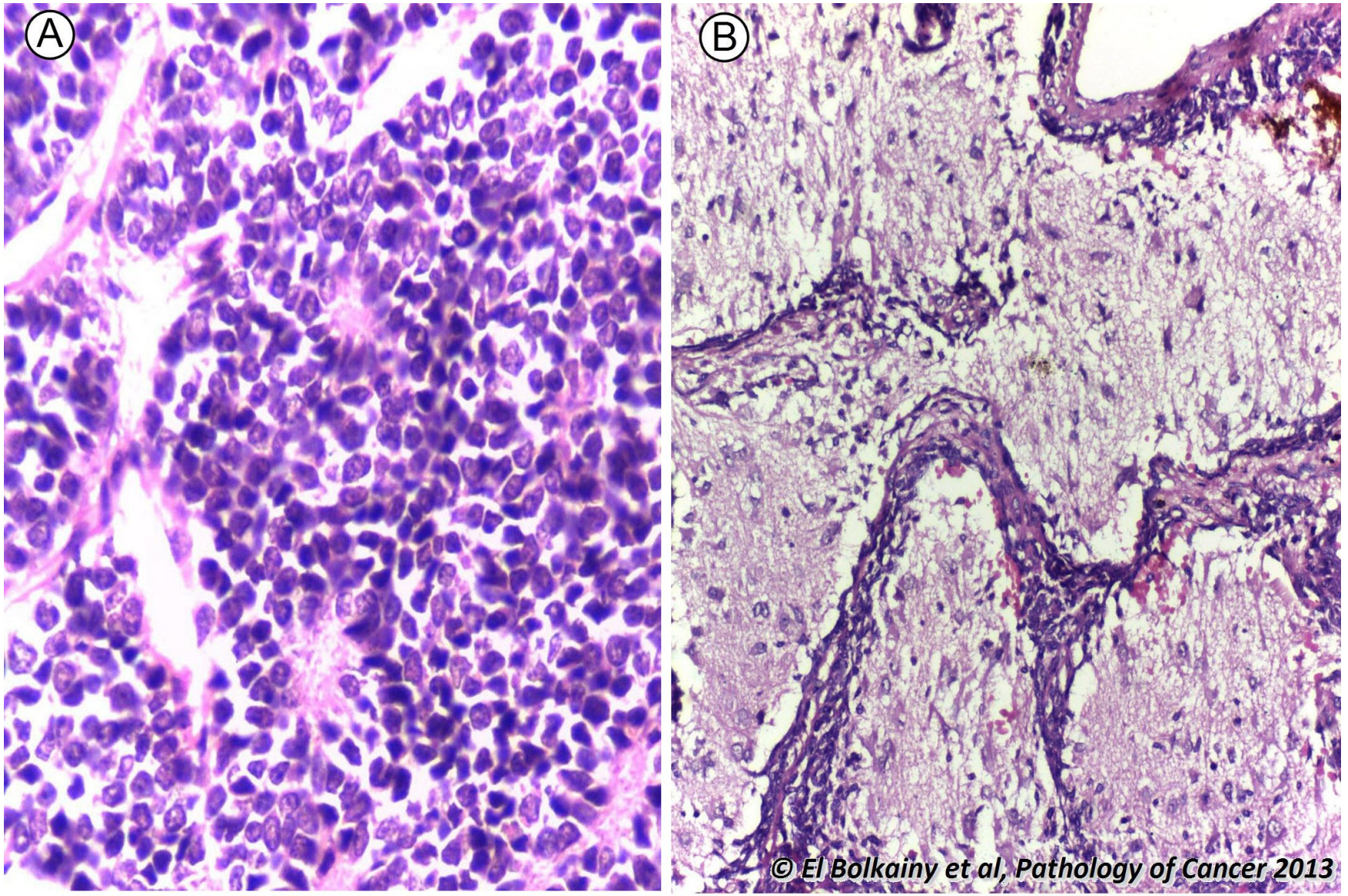
Picture 21-32 **Primitive neuroectodermal tumor (Ewing / PNET).** The histology is characterized by uniform round cells, fine dispersed chromatin and vascular stroma. The pattern is solid and vaguely nodular. Surface immunoreactivity to CD99 (MIC2). FLI-1 are confirmatory markers. **A** Low power. **B** High power.

21.33 Neuroepithelioma.



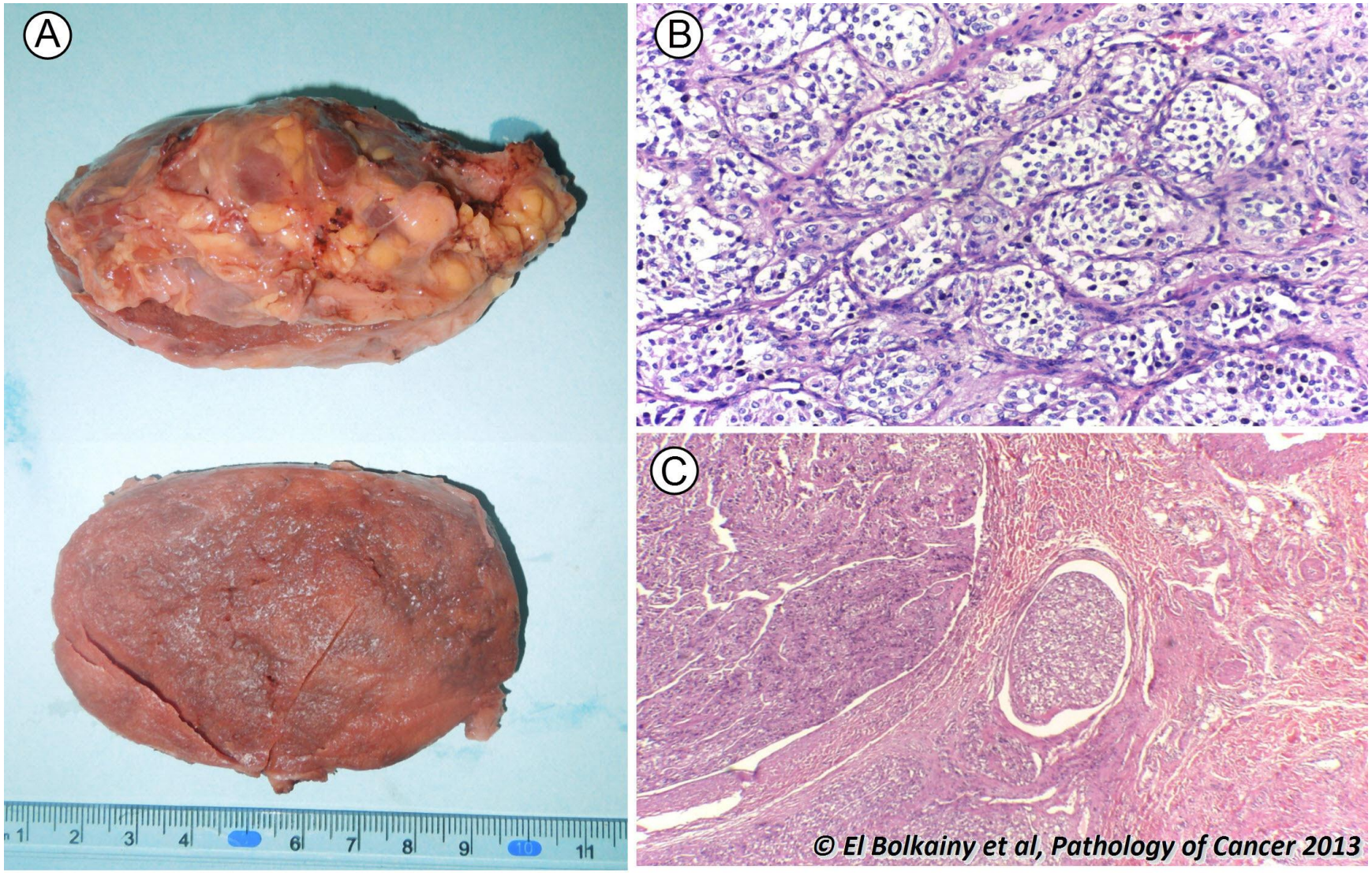
Picture 21-33 **Neuroepithelioma.** It is composed of round cells as PNET with solid vaguely nodular pattern and shows numerous pseudorosettes of the Homer-wright type (lumen filled with fibrillary material).

21.34 Neuroblastoma, histology.



Picture 21-34 **Neuroblastoma, histology. A** It is composed of round cells with few pseudorosettes or ganglionic cells, organized in groups, surrounded by neurilemmal stromal cells. Tumor cells are immunoreactive to chromogranin and synaptophysin. Elevated levels of ctecholamines in blood and urine. **B** Stroma-rich neuroblastoma. Pale areas represent abundant neurilemmal stroma while bluish areas show neuroblastic component.

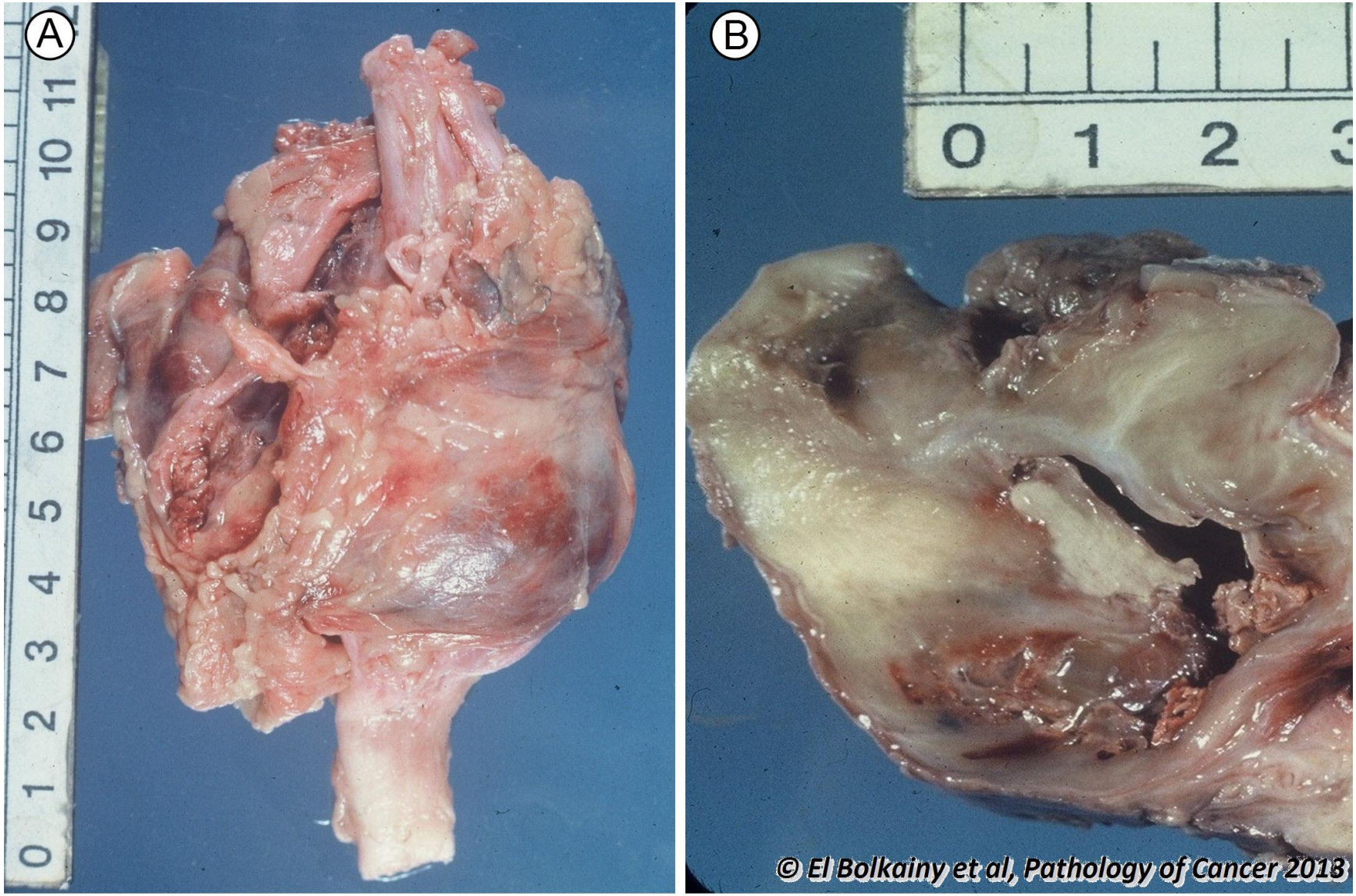
21.35 Paraganglioma.



Picture 21-35

Paraganglioma. A Gross appearance. Outer surface showing fibrofatty pseudocapsule (upper specimen) with brownish cut section (lower specimen). **B** Histology, the cells are rounded and show a nesting pattern (Zellbalen) surrounded by fibrovascular stroma containing supporting sustentacular cells. Tumor cells are positive for chromogranin and sustentacular cells are positive for S-100. **C** Malignant paraganglioma with vascular and stromal invasion. Note the loss of nesting pattern.

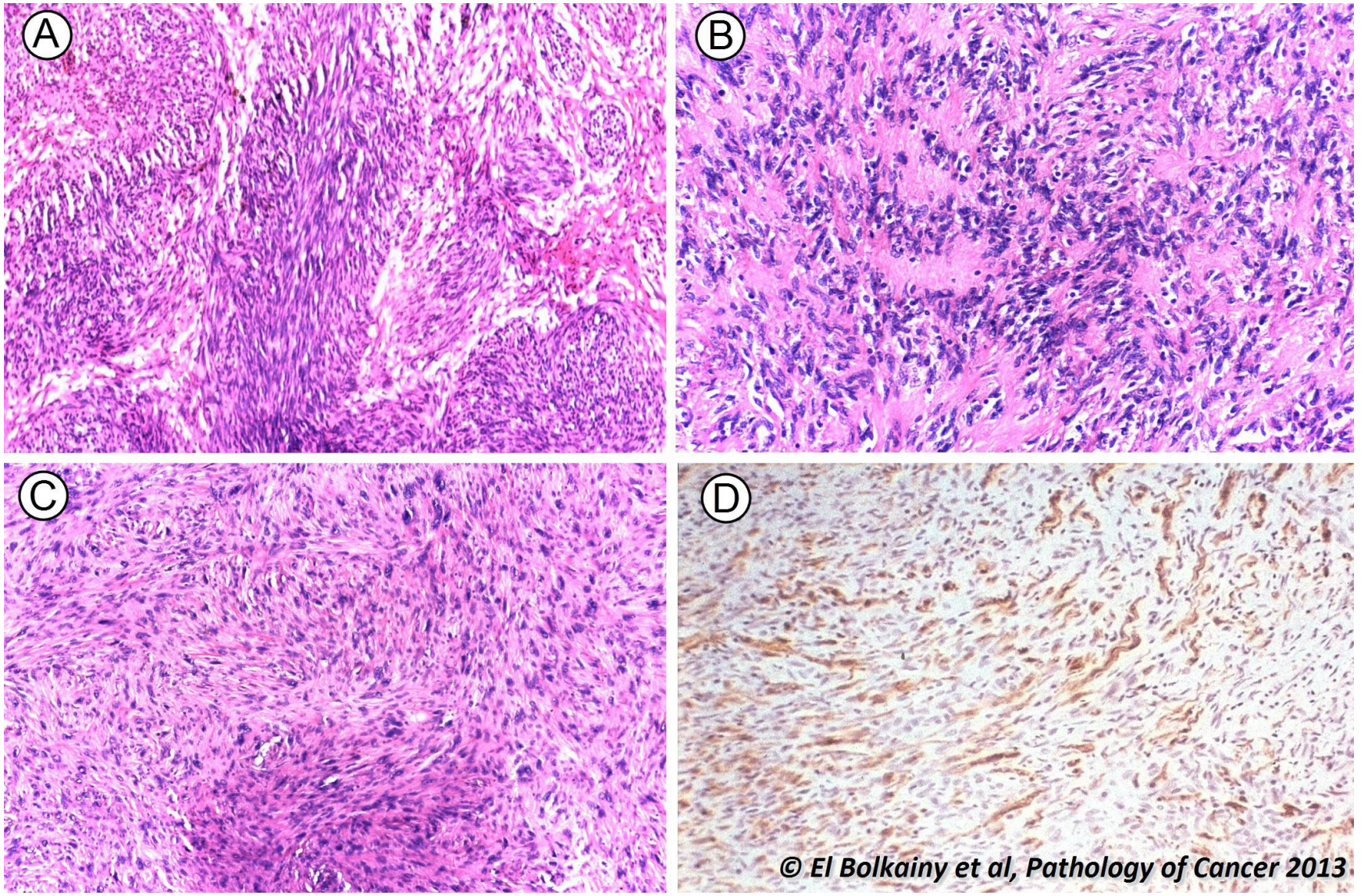
21.36 Malignant peripheral nerve sheath tumor (MPNST) of sciatic nerve, gross features.



Picture 21-36

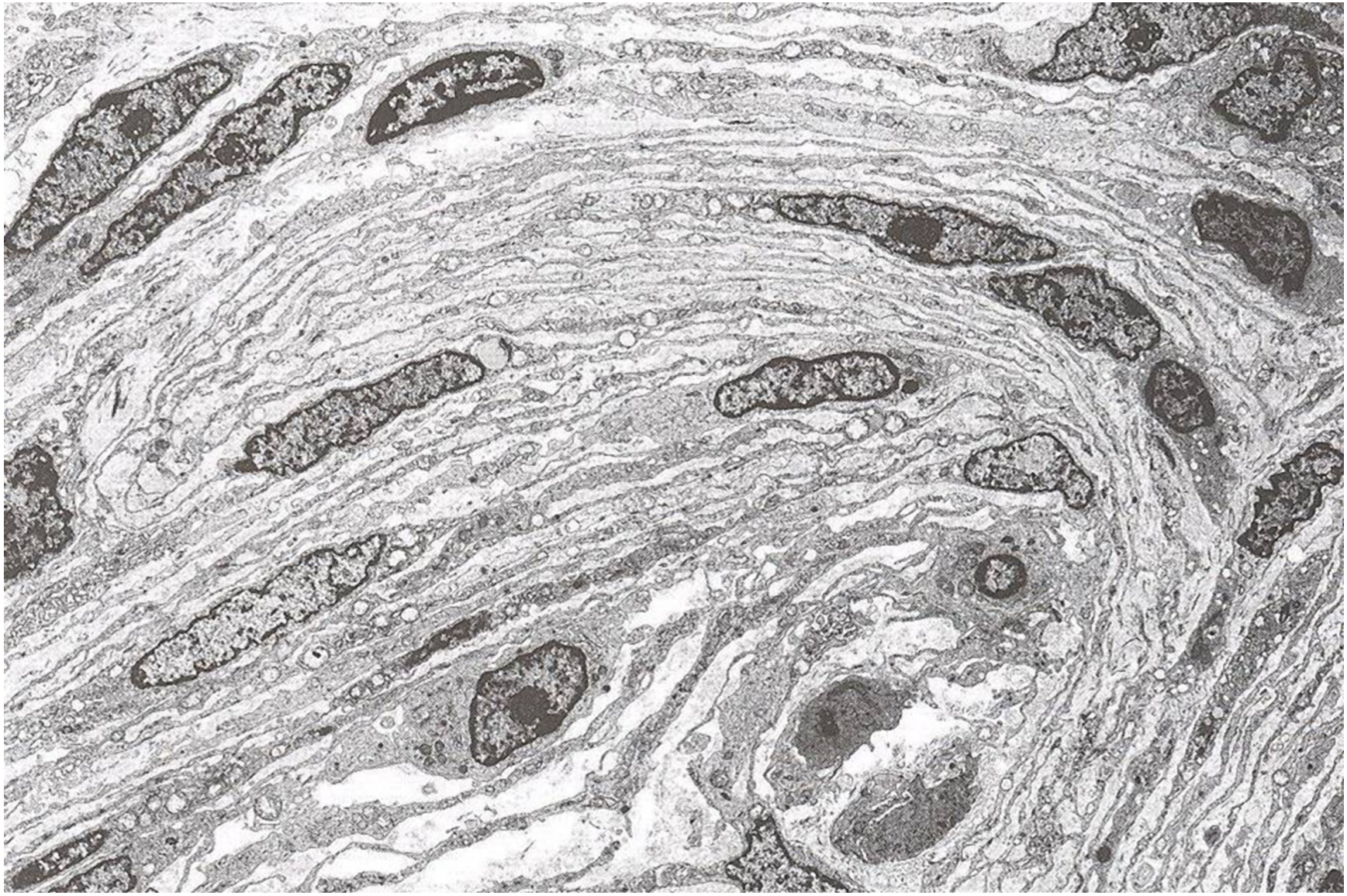
Malignant peripheral nerve sheath tumor (MPNST) of sciatic nerve, gross features. A Surface view showing the charcteristic fusiform shape of tumor along the nerve. **B** Cross section, demonstrating gray white color, focal areas of hemorrhage, necrosis, and cystic change. This tumor may complicate multiple neurofibromatosis (NF-1 syndrome).

21.37 Malignant peripheral nerve sheath tumor, histology.



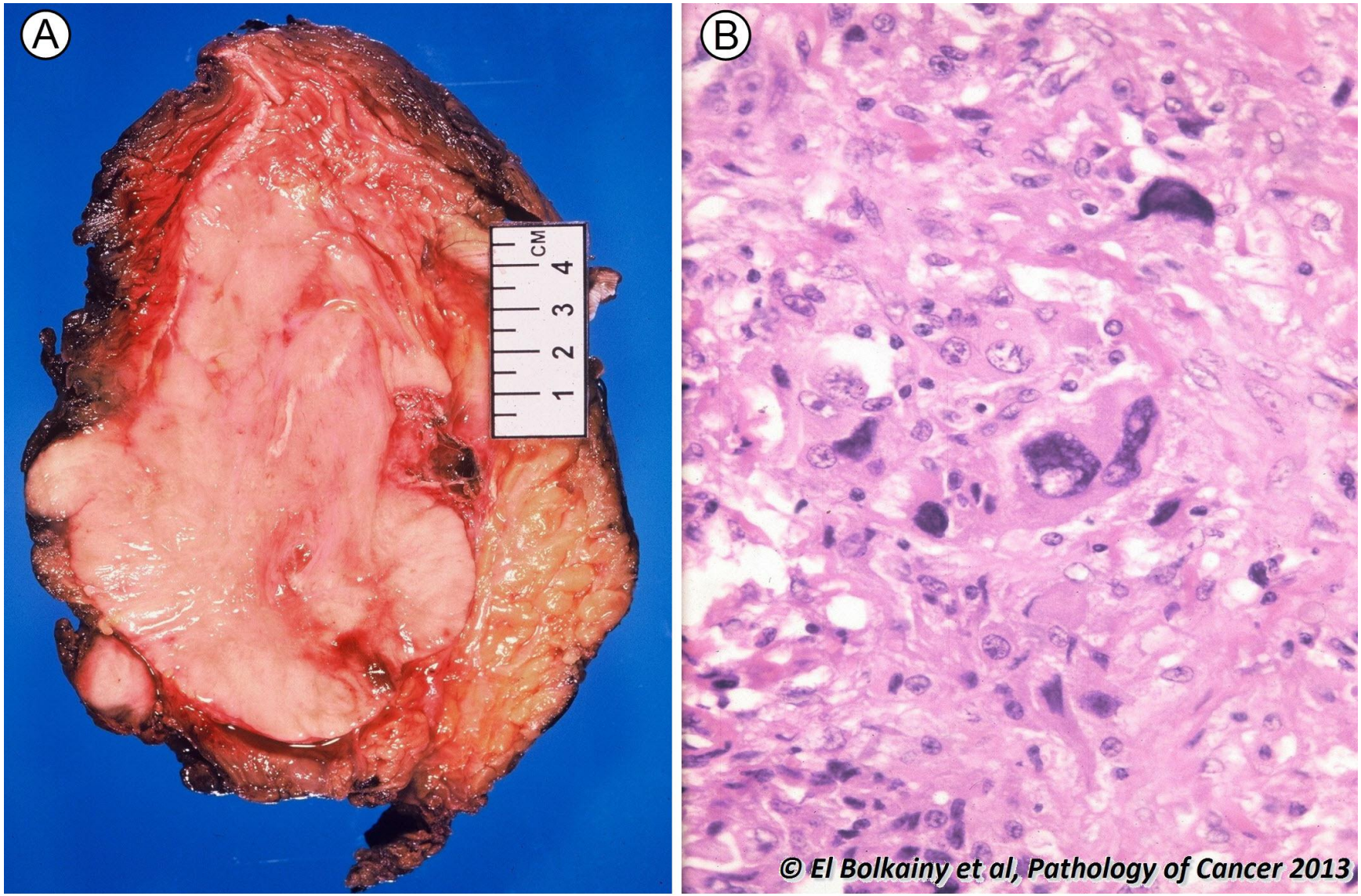
Picture 21-37 **Malignant peripheral nerve sheath tumor, histology . A** and **B** Characteristic features are marked nuclear hyperchromasia compared to cytoplasm, and areas of hypercellularity alternating with hypocellualrity (marble-like appearance). **C** The bundles are patternless, but, rarely may show palisading or whorled patterns. **D** Immunoreactivity: S-100, rarely desmin positive (Triton tumor).

21.38 Malignant peripheral nerve sheath tumor (MPNST), electron microscopic features.



Picture 21-38 **Malignant peripheral nerve sheath tumor (MPNST), electron microscopic features.** Complex cytoplasmic processes form rounded spaces around collagen fibrils (mesaxon formation) characteristic of neurilemmal differentiation. (Reproduced with permission, Fletcher CD, 2007).

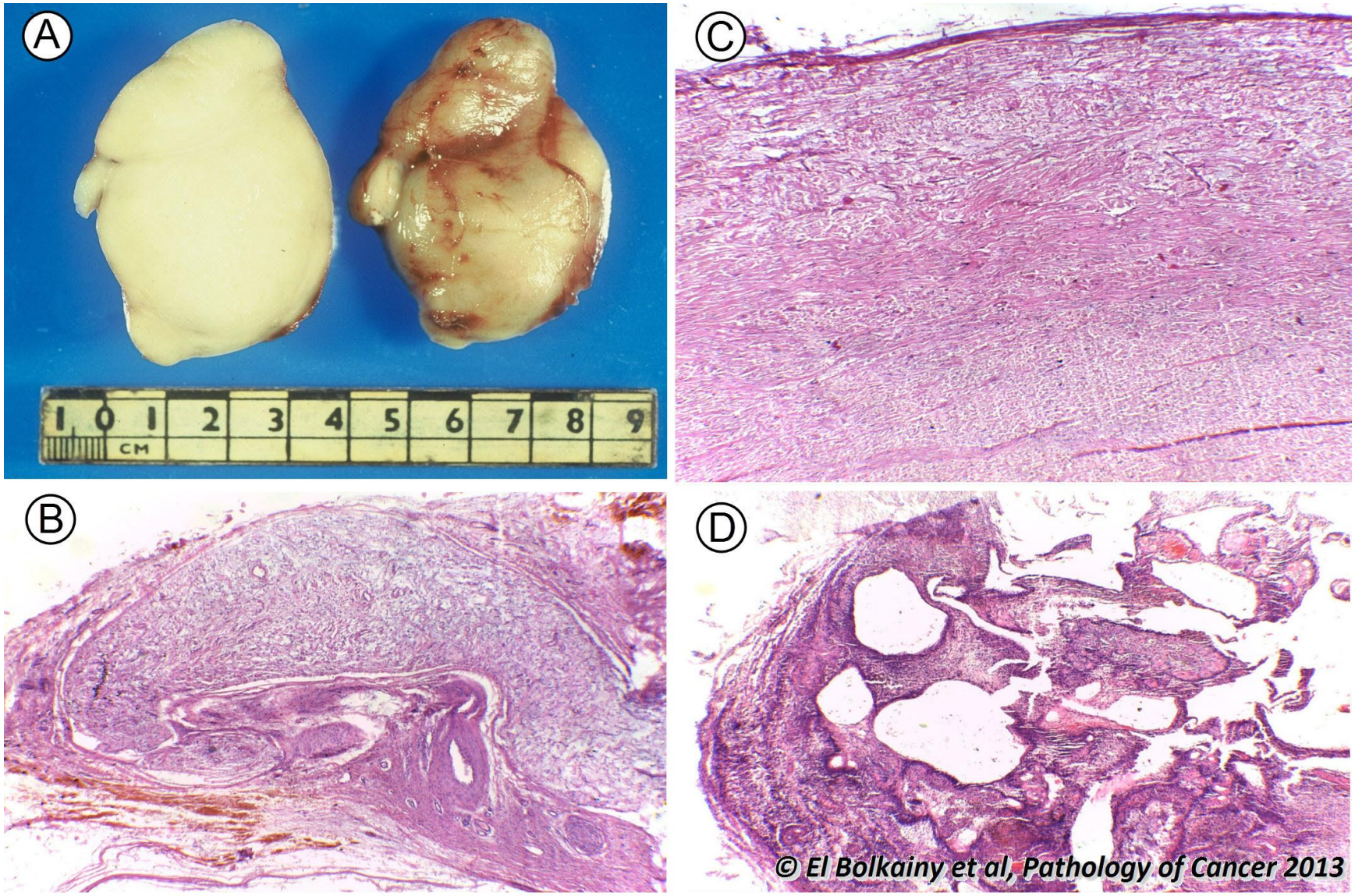
21.39 Malignant granular cell tumor of thigh.



Picture 21-39

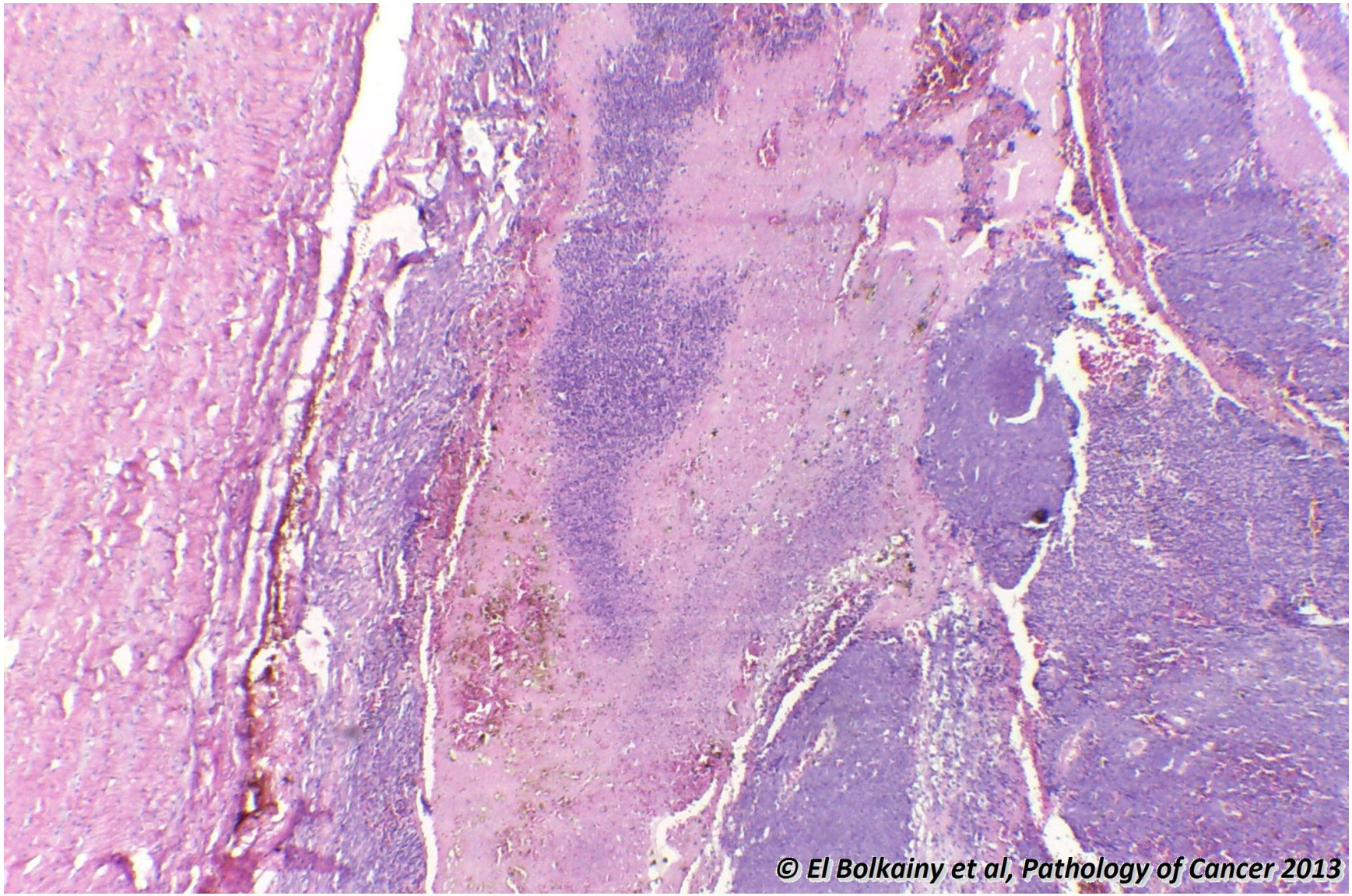
Malignant granular cell tumor of thigh. A Gross appearance: Note the greyish color, large size, areas of degeneration as well as invasion of surrounding fatty soft tissue. **B** Histollogically, it is composed of large polygonal cells with eosinophilic granular cytoplasm (S-100 and CD 68 positive), mitosis > 2/10 HPF, and focal necrosis. The large tumor size and prominent nuclei distinguish it from the benign type.

21.40 Neurofibroma.



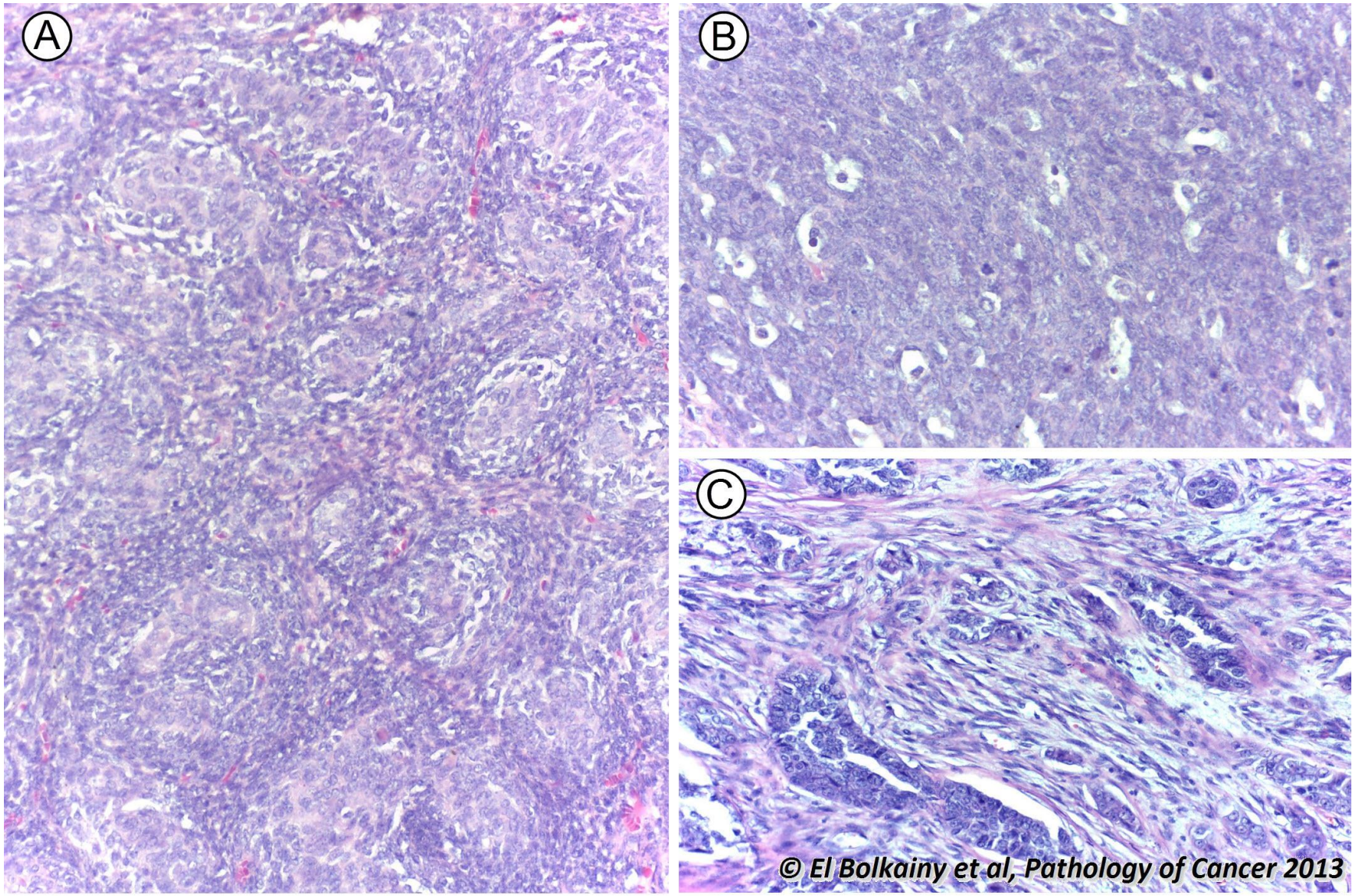
Picture 21-40 **Neurofibroma. A** Gross appearance: a well-defined, gray white, small tumor mass. **B, C,** and **D** The histology is characterized by slender spindle cells (S-100 positive) with wavy (serpentine) pattern, plisade pattern (verocay bodies), associated fibrosis, vascularity and cystic change.

21.41 Monophasic synovial sarcoma, histology, low power.



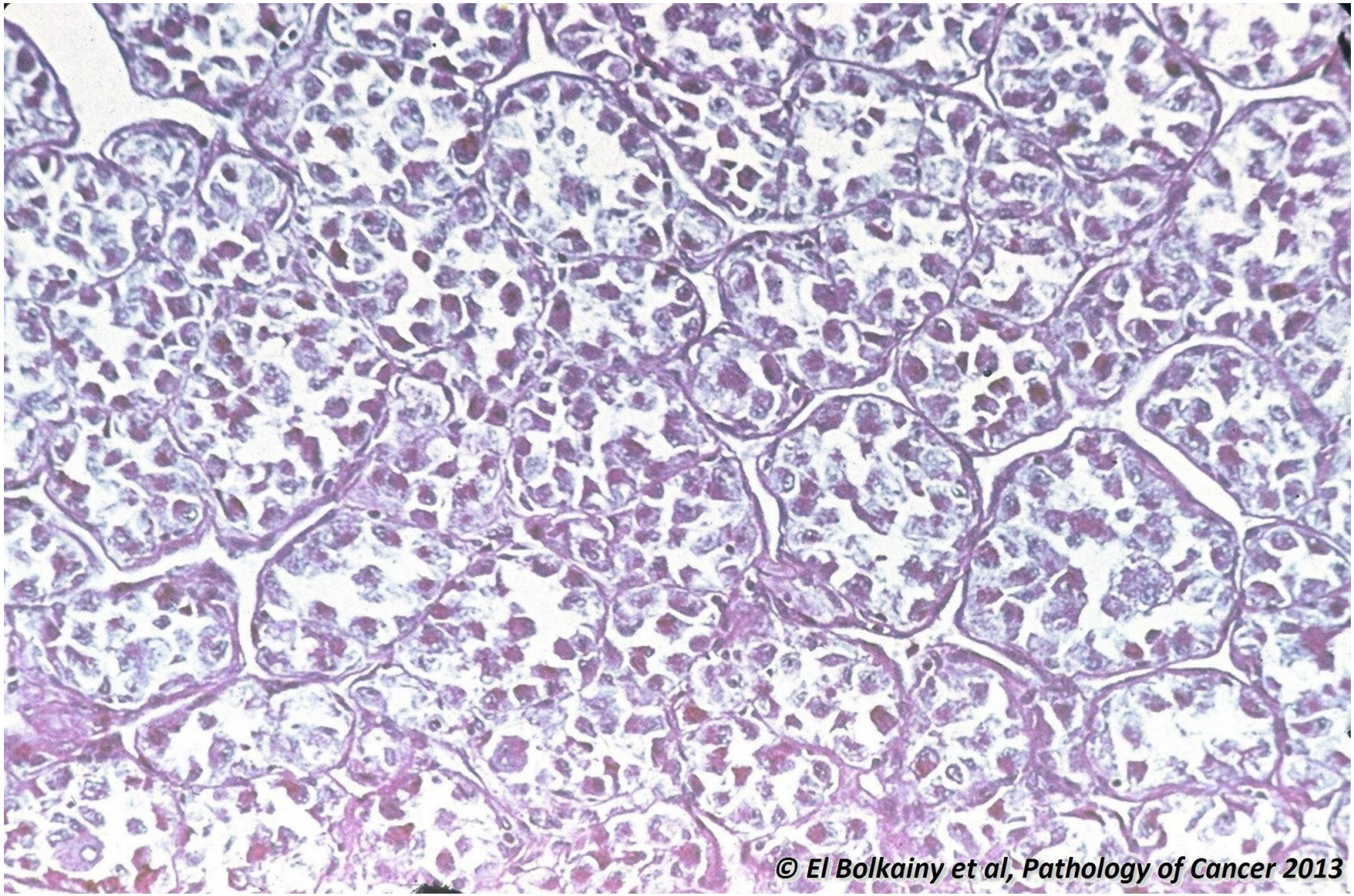
Picture 21-41 **Monophasic synovial sarcoma, histology, low power.** Areas of hypercellularity alternating with hypocellularity (marble-like appearance). Both are bluish in color with variable intensity. eosinophilic tissue represent normal stroma infiltrated by the tumor.

21.42 Synovial sarcoma, histology.



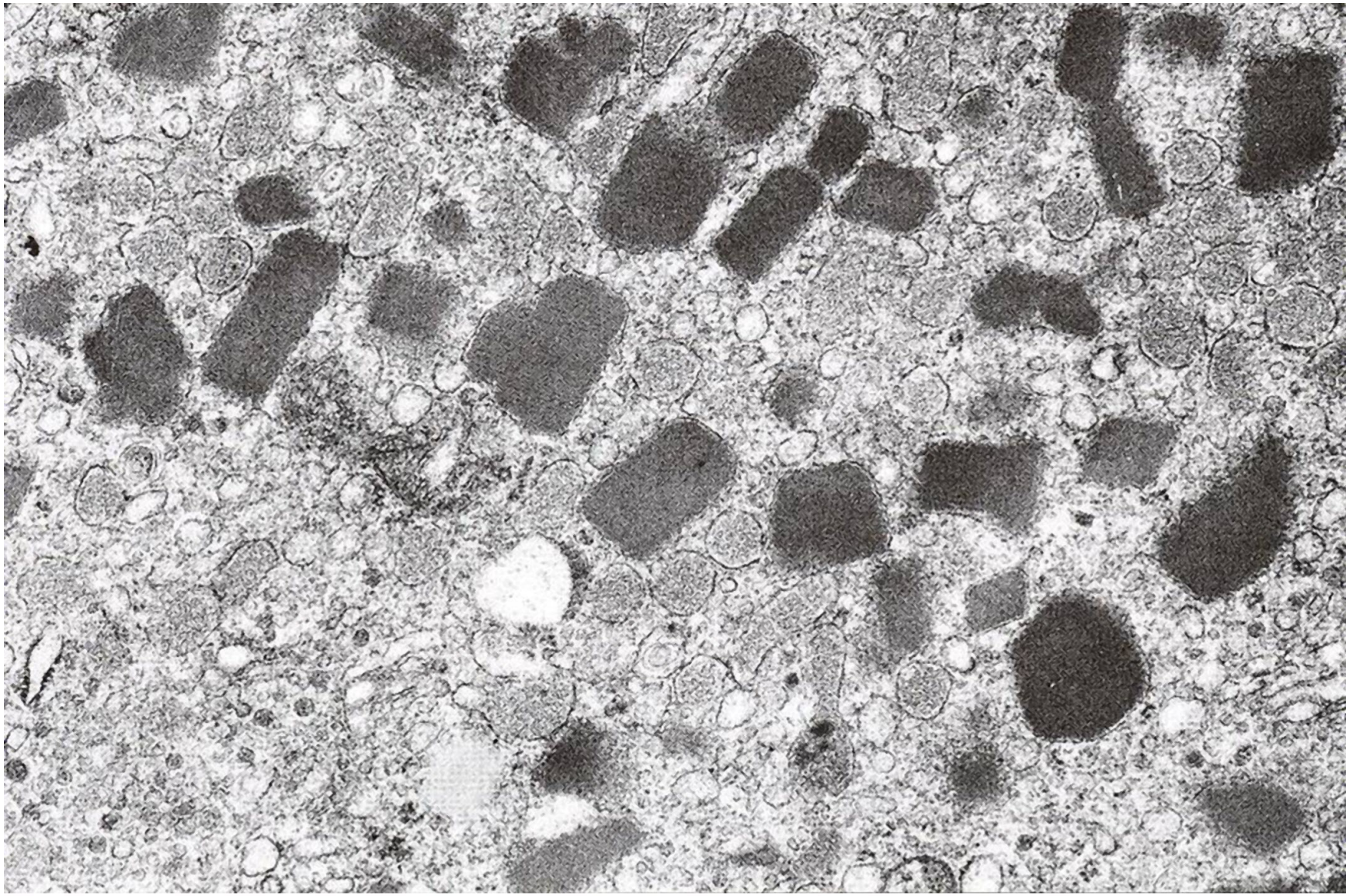
Picture 21-42 **Synovial sarcoma, histology. A** The biphasic tumor is composed of spindle cells with carrot-shaped overlaping nuclei as well as solid groups of epithelial cells (pale areas). **B** Epithelial-lined spaces. **C** Biphasic tumor with stromal and glandular components. Pericytoma-like pattern and focal calcification may be seen (not shown). Immunoreactivity to CK (70%), CD 99 (65%) and TLE-1 (92%). t(X;18) is confirmatory.

21.43 Alveolar soft part sarcoma.



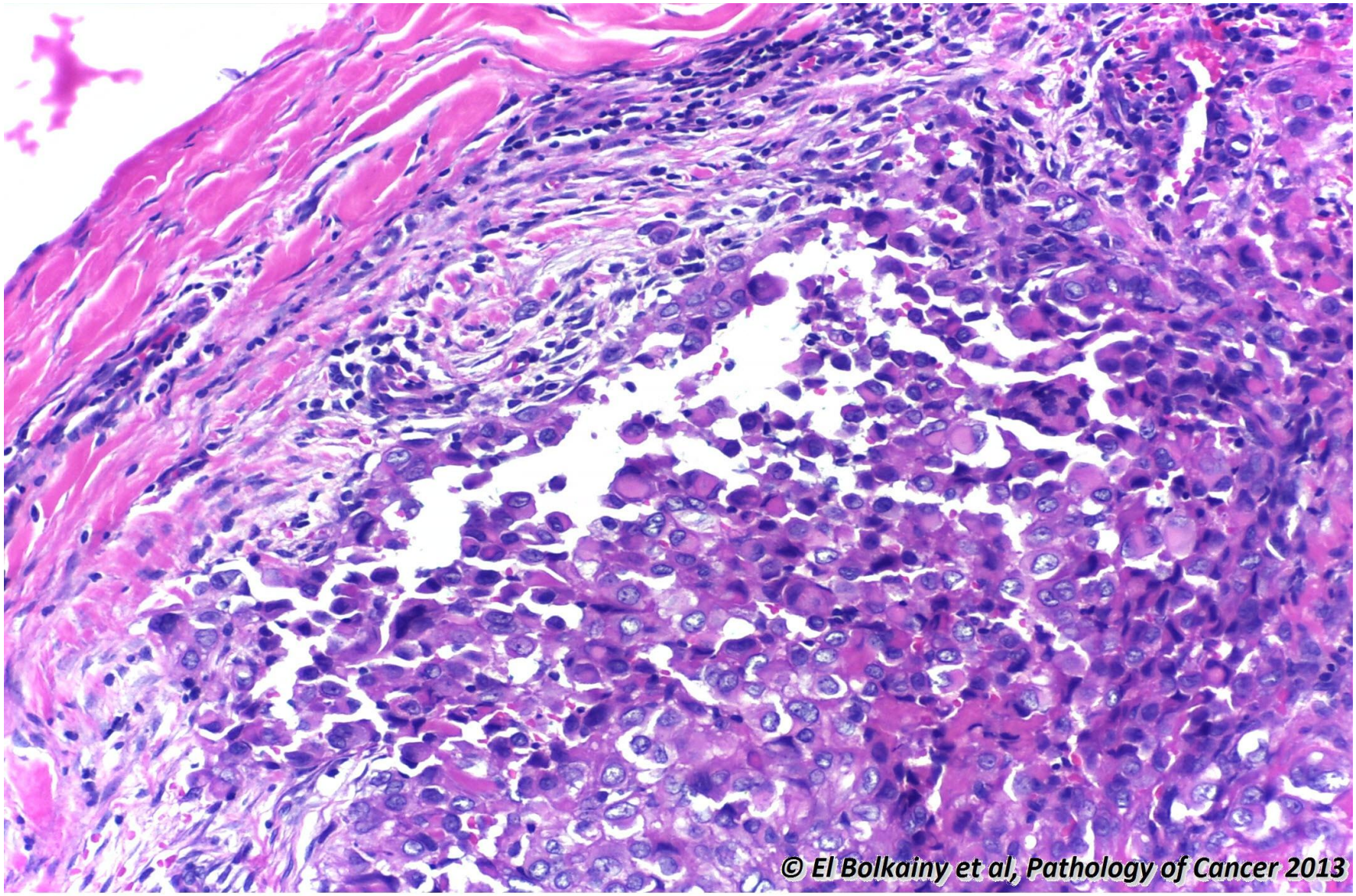
Picture 21-43 **Alveolar soft part sarcoma.** It is composed of nests of polygonal cells with abundant eosinophilic granular cytoplasm, eccentric nuclei with prominent nucleoli. Apart from nuclear positivity to TEE-3, immunohistochemistry plays a limited diagnostic role.

21.44 Alveolar soft part sarcoma, electron microscopic features.



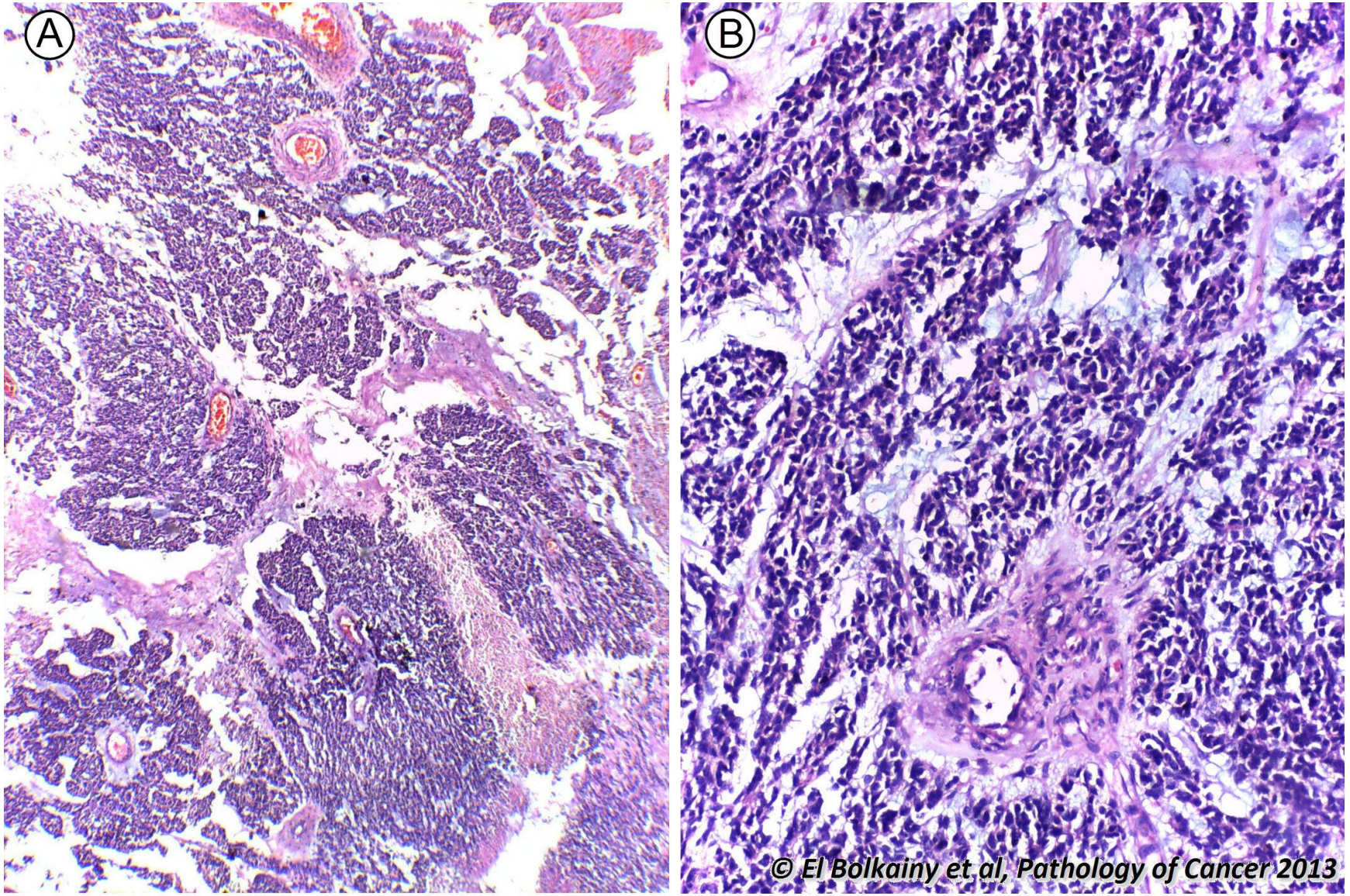
Picture 21-44 **Alveolar soft part sarcoma, electron microscopic features.** Characteristic structures are: membrane-bound rhomboid crystalline material in cytoplasm and small granules found close to Golgi apparatus.(Reproduced with permission, Fletcher CD, 2007)

21.45 Epithelioid sarcoma, histology.



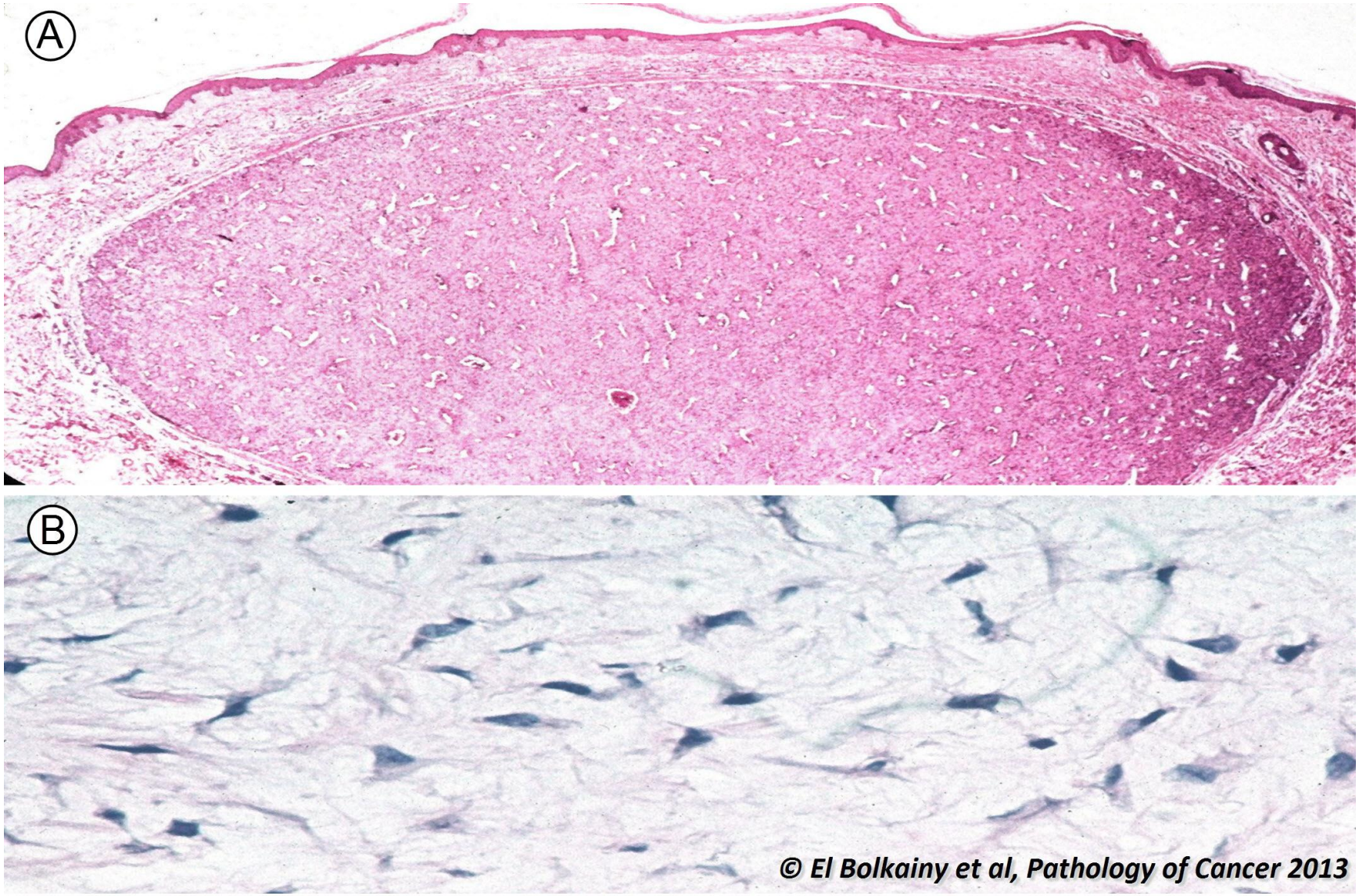
Picture 21-45 **Epithelioid sarcoma, histology.** It is characterized by proliferating atypical spindle and epithelioid cells associated with marked necrosis. Co-expression of CK, vimentin, and CD 34 (50%) is characteristic. Loss of INI-1 protein expression helps to differentiate it from squamous carcinoma and other sarcomas which usually express this protein.

21.46 Desmoplastic round cell tumor (DRCT), histology.



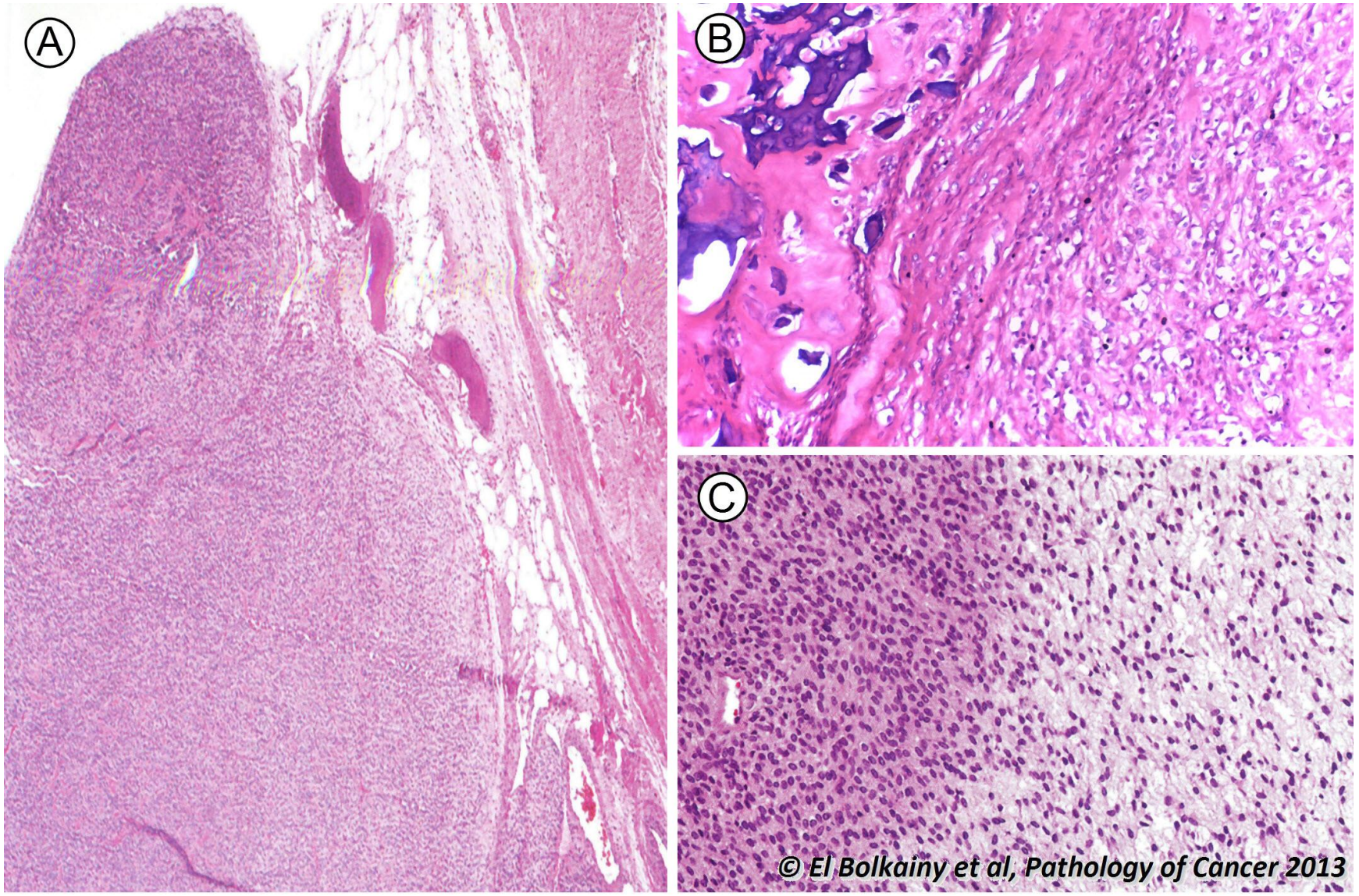
Picture 21-46 **Desmoplastic round cell tumor (DRCT), histology.** It is composed of nests of small round cells in abundant fibrotic stroma. Co-expression of desmin and CK (dot-like pattern) is characteristic. t (11:22) and positive EWS-WT-1 protein are confirmatory. **A** Low power. **B** High power.

21.47 Myxoma, histology.



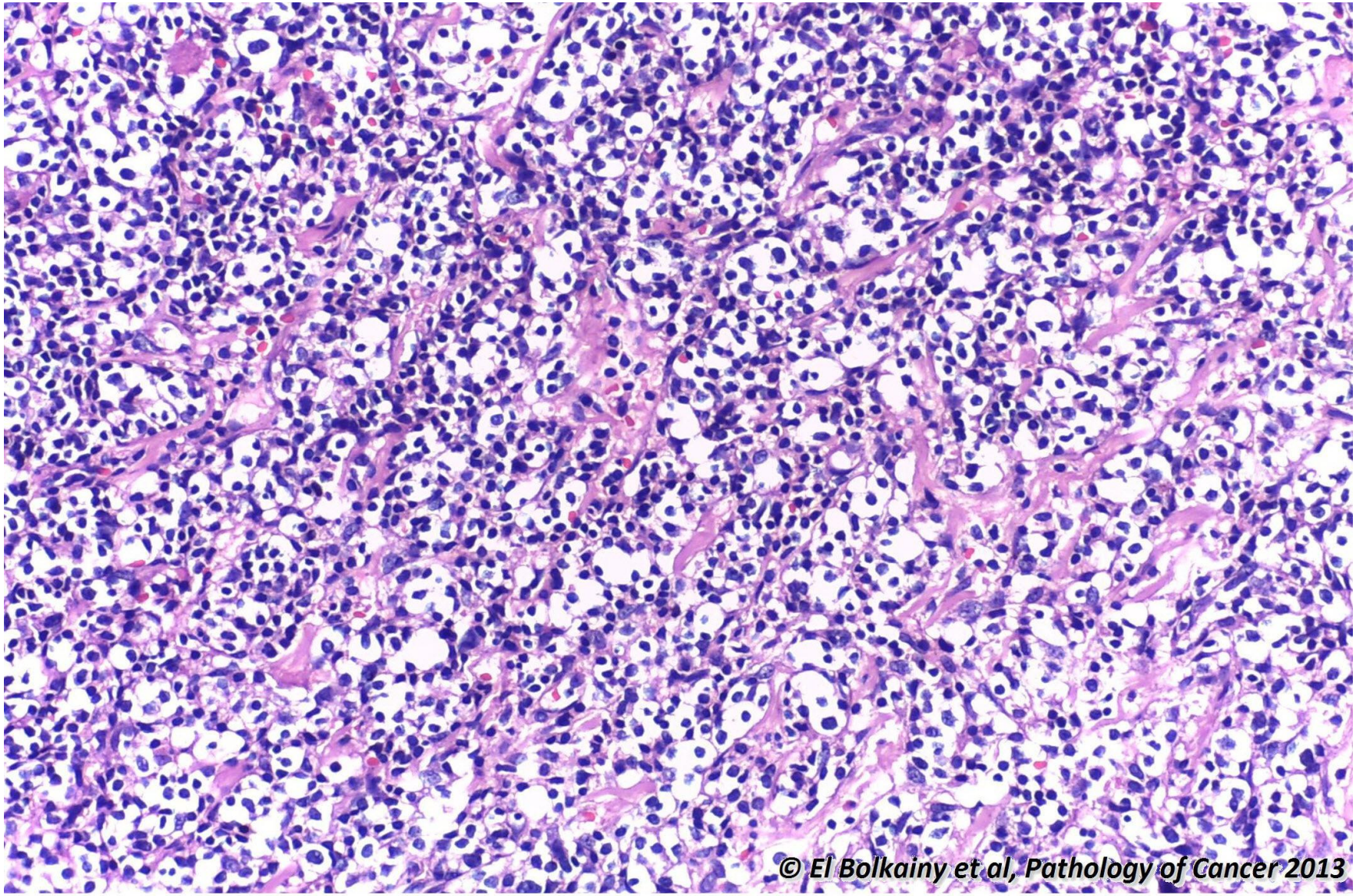
Picture 21-47 **Myxoma, histology. A** The conventional type is composed of stellate and spindle cells (vimentin positive) in myxoid matrix (Alcian blue positive). **B** Aggressive angimyxoma affects genital areas and shows a prominent vascular component (desmin +, ER and PR+) and associated with high risk of local recurrence.

21.48 Ossifying fibromyxoid tumor, histology.



Picture 21-48 **Ossifying fibromyxoid tumor, histology**. **A** It is composed of oval and spindle cells in fibromyxoid matrix, surrounded by a partial shell of mature bone. Its immunoreactivity is complex: S-100 positive (70%), rarely CK, GFAP, desmin and actin are positive. **B** High power of the shell. **C** High power of the tumor cells.

21.49 Perivascular epithelioid cell tumor (PECOMA), histology.



Picture 21-49 **Perivascular epithelioid cell tumor (PECOMA), histology.** It is characterized by perivascular arrangement of epithelioid cells with clear or granular cytoplasm. The co-expression of actin and melanocytic markers (Melan-A and HMB-45) is diagnostic.