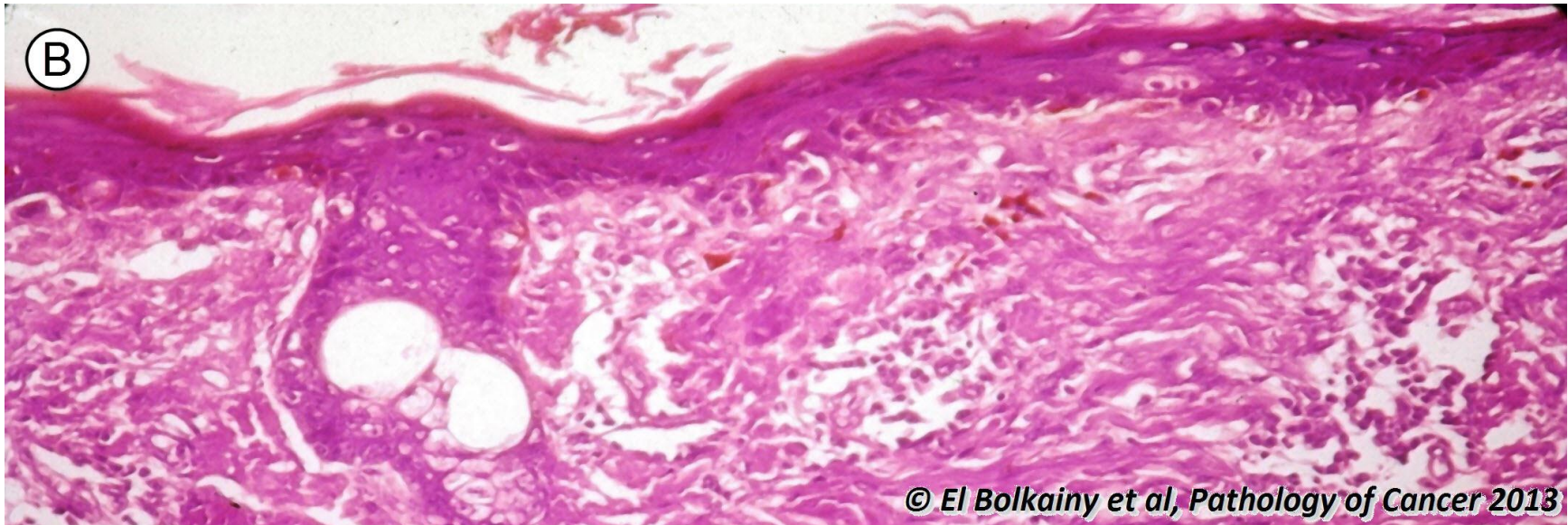
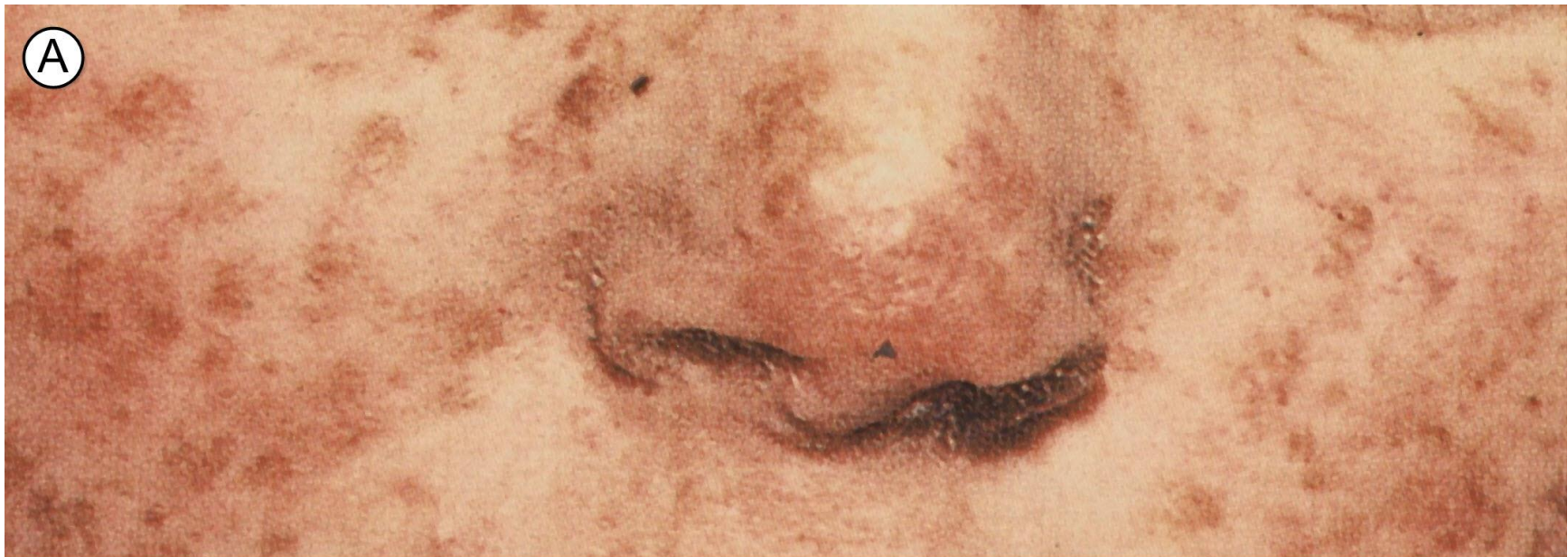


## Chapter 22

# Skin tumors

22.1 Xeroderma pigmentosum.



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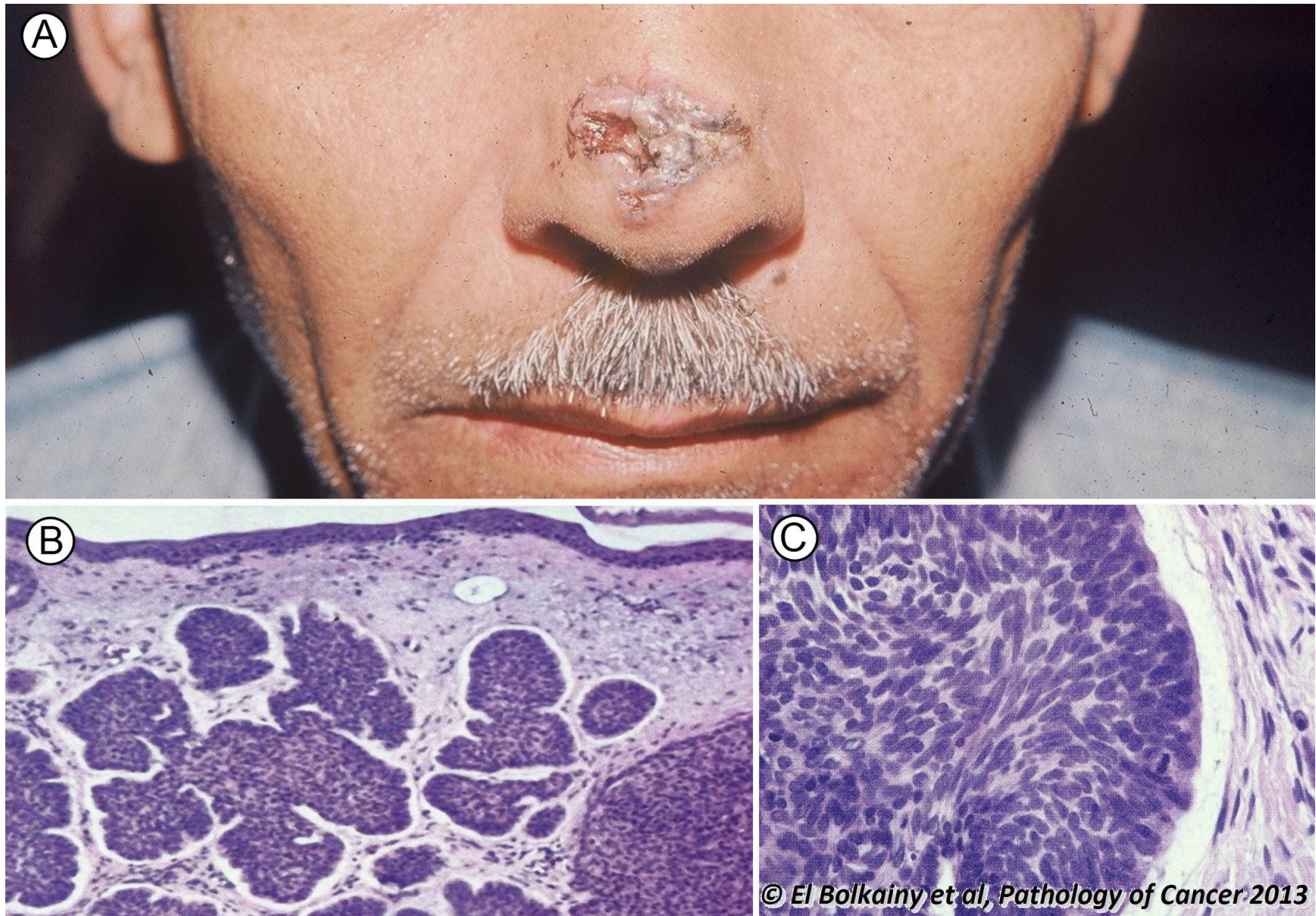
**Picture 22-1** Xeroderma pigmentosum. **A** Clinical picture of face showing multiple tumors and solar keratotic lesions. **B** Histology, atrophy of skin and hair follicles with marked solar degeneration in the dermis.

22.2 Multiple basal cell carcinomas in an albino patient.



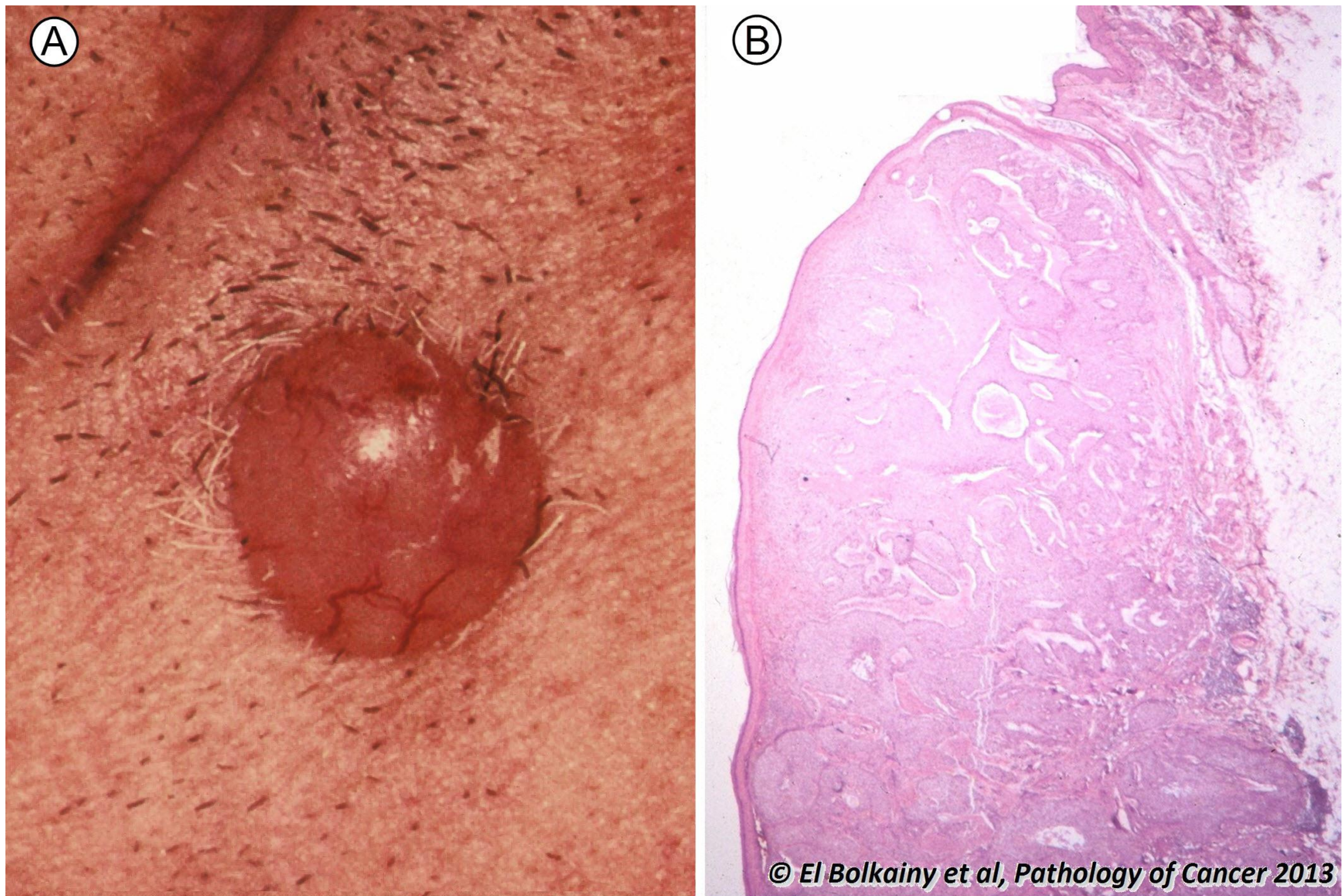
**Picture 22-2** Multiple basal cell carcinomas in an albino patient. A small tumor at skin of lower neck and another in the shoulder region showing characteristic rolled-in edge.

## 22.3 Basal cell carcinoma.



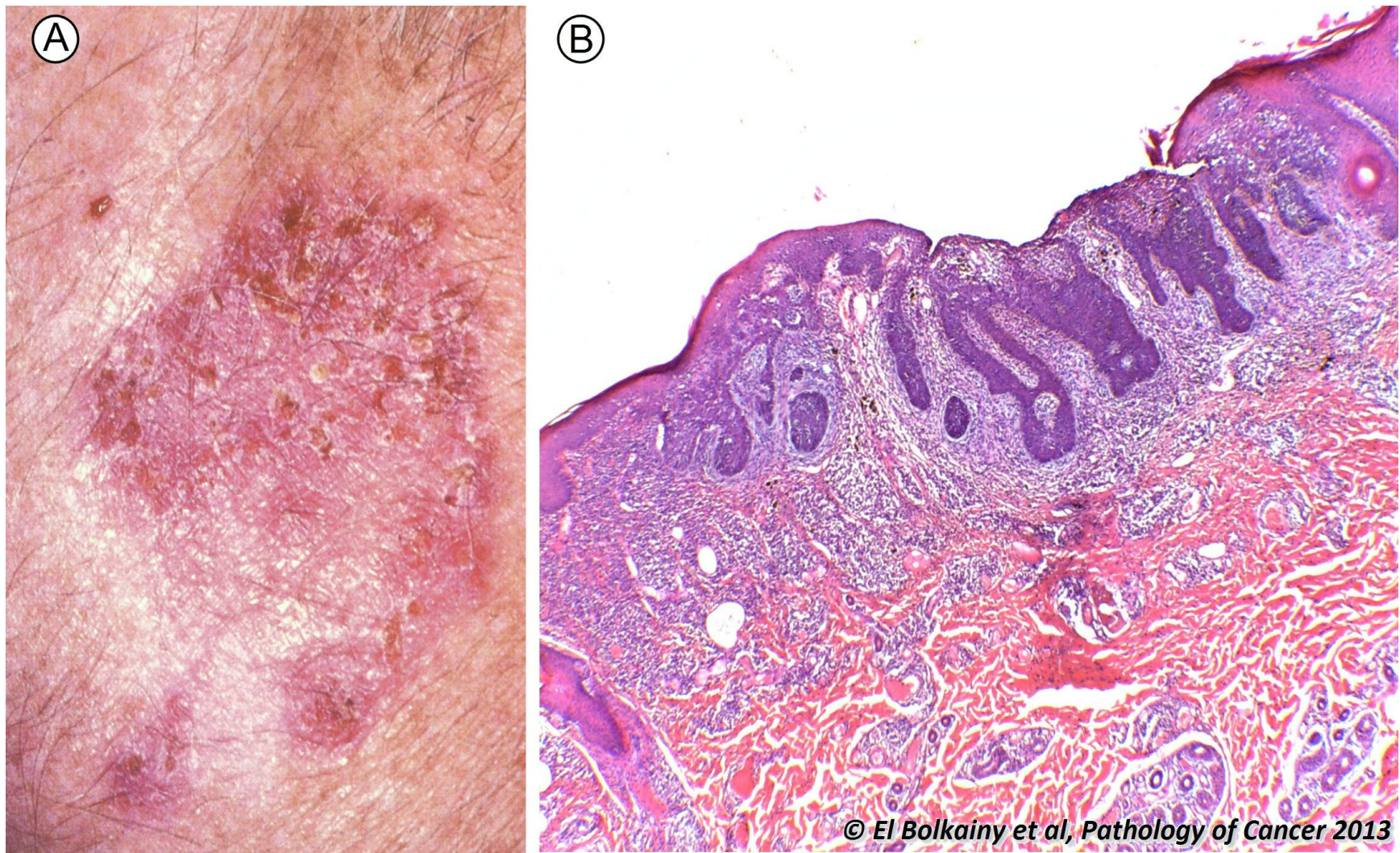
**Picture 22-3** Basal cell carcinoma. **A** Gross features, showing an ulcer with beaded rolled-in edge. **B** and **C** The histology is characterized by basaloid cells with peripheral palisading and retraction artifact from stroma.

22.4 Basal cell carcinoma, nodular type.



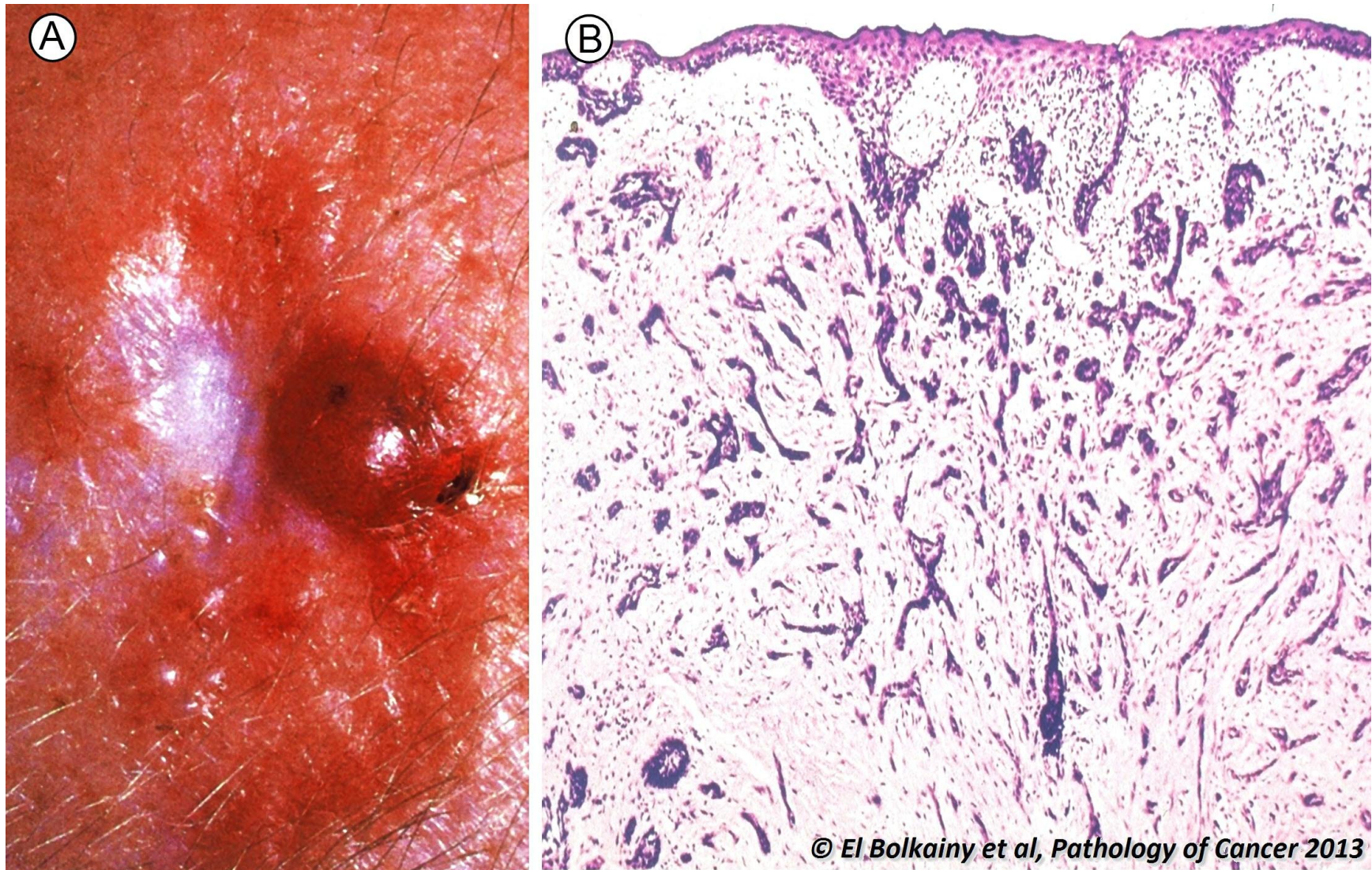
**Picture 22-4** Basal cell carcinoma, nodular type. **A** Gross features, presents as a rounded nodule. **B** Histologically, it is rather circumscribed, easily excised with safety margin, hence, the lower incidence of recurrence than infiltrating BCC.

22.5 Basal cell carcinoma, superficial type.



**Picture 22-5** Basal cell carcinoma, superficial type. **A** Gross features. **B** Histology, showing multicentric origin and superficial nature. Recurrence is not uncommon at lateral margins of excision.

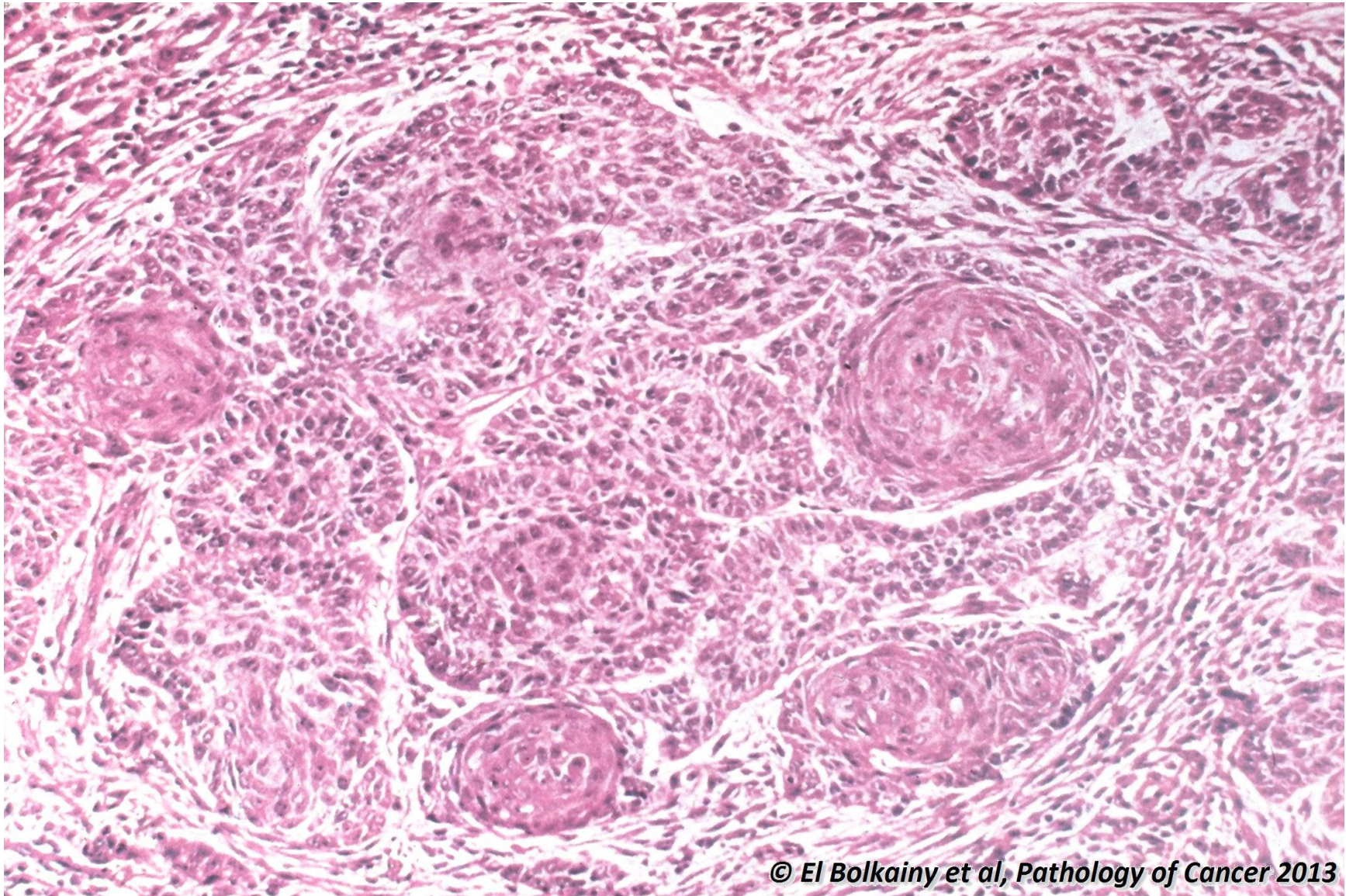
## 22.6 Basal cell carcinoma, infiltrating morpheiform type.



**Picture  
22-6**

**Basal cell carcinoma, infiltrating morpheiform type.** **A** Grossly, it appears whitish with induration due to associated fibrosis and it is difficult to identify the edge of tumor during excision, hence, high rate of recurrence. **B** Histology shows scattered small groups of carcinoma with desmoplasia.

22.7 Keratotic basal cell carcinoma.

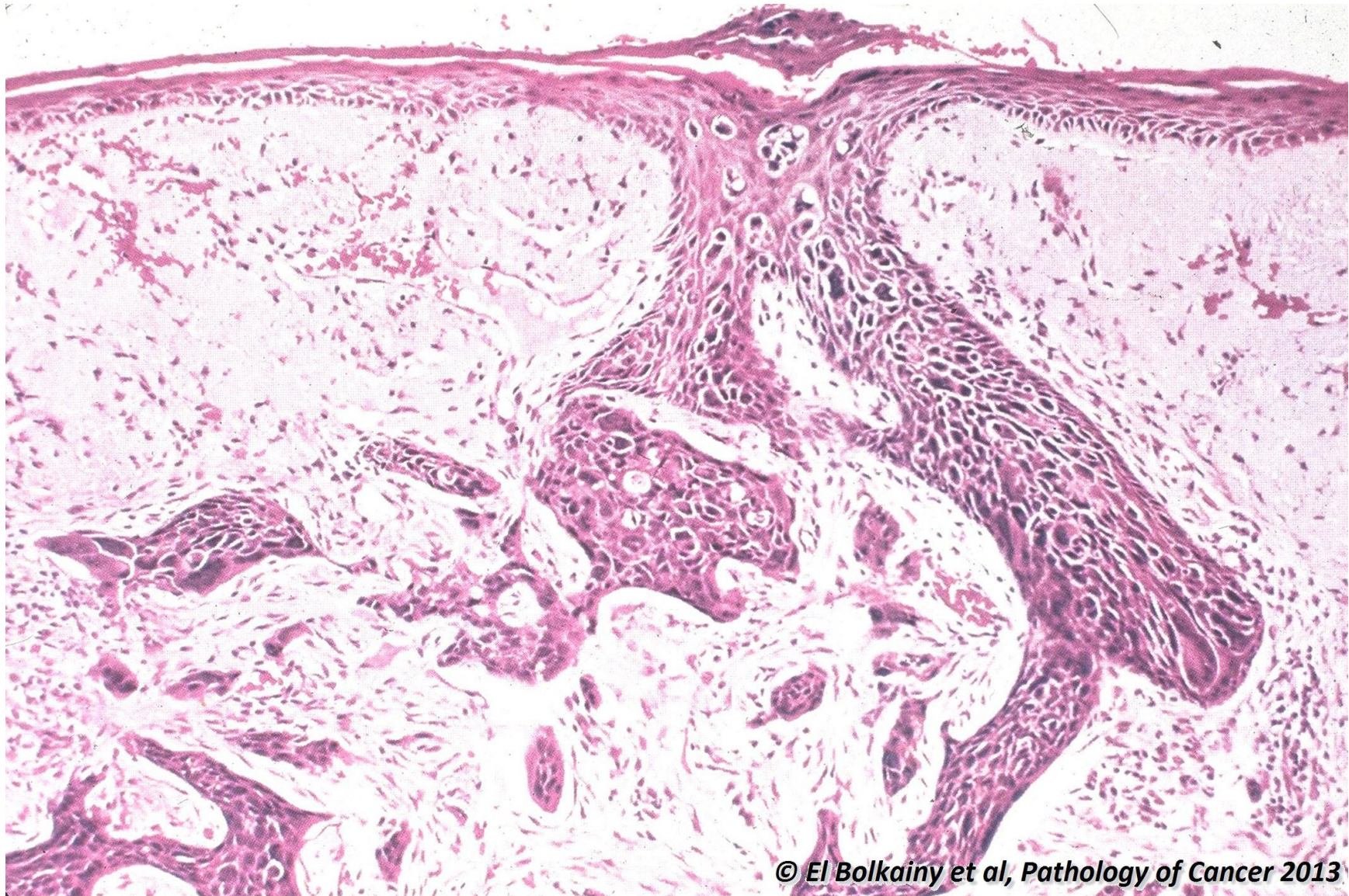


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**Picture 22-7** Keratotic basal cell carcinoma. A classic basal cell carcinoma showing focal central squamous differentiation in the neoplastic cell groups. The tumor behaves as basal cell carcinoma.



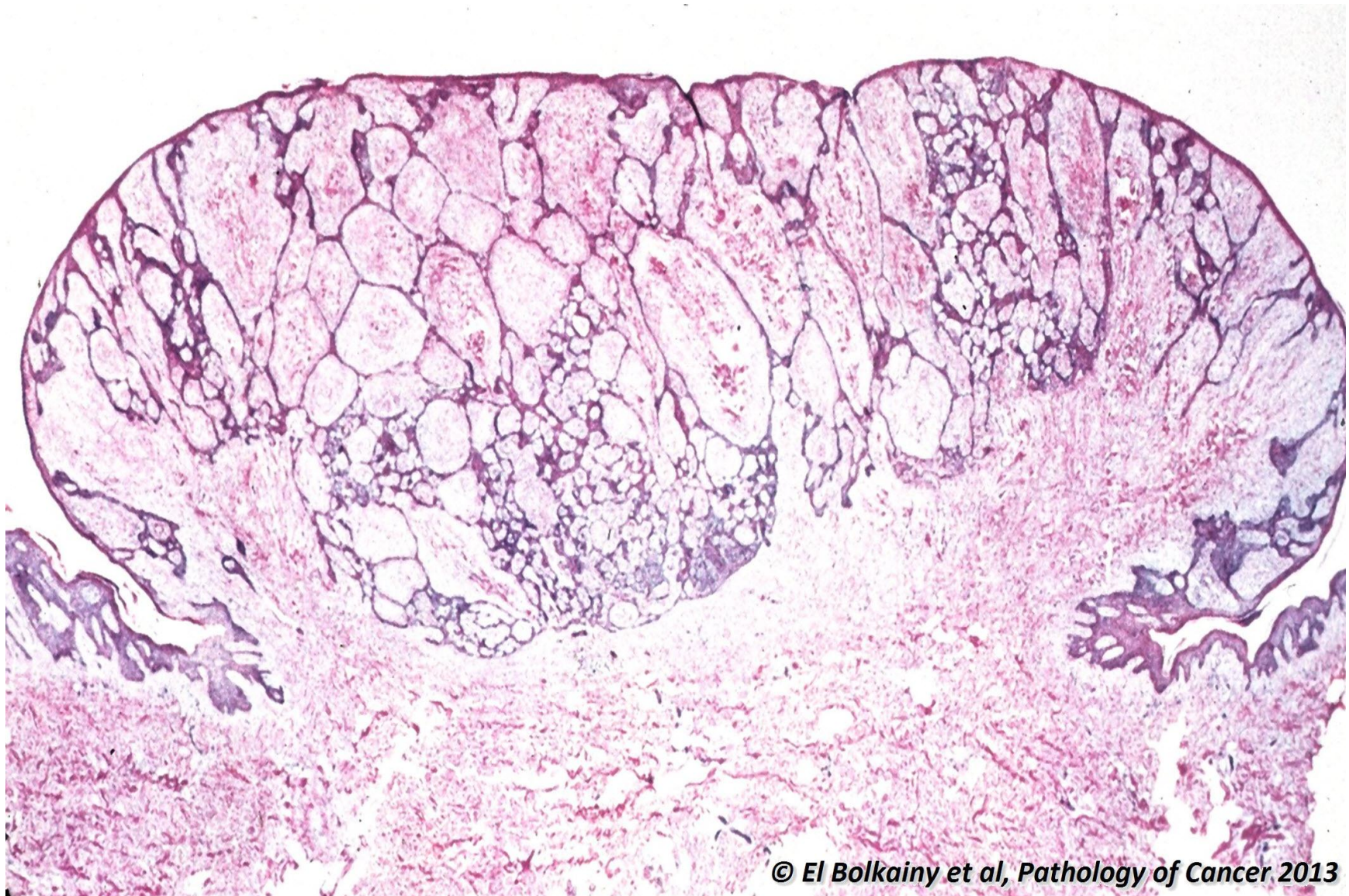
## 22.8 Basosquamous cell carcinoma, histology.



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**Picture 22-8** Basosquamous cell carcinoma, histology. The pattern is that of basal cell carcinoma, however, the entire cell population has undergone squamous differentiation. This tumor behaves as squamous cell carcinoma.

22.9 Fibroepithelial basal cell carcinoma, histology.

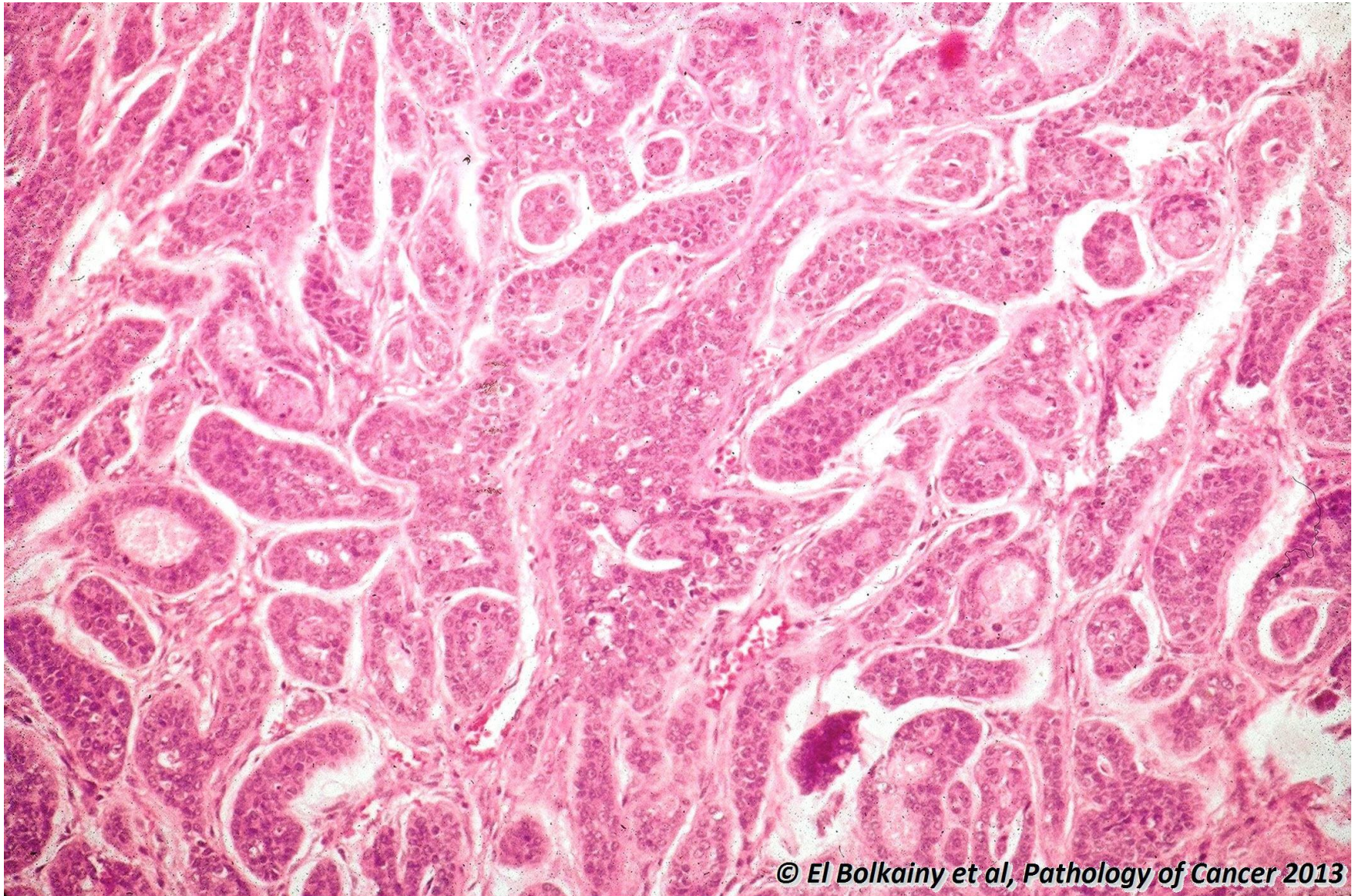


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**Picture 22-9**

**Fibroepithelial basal cell carcinoma, histology.** This rare histological variant may be underdiagnosed because of the marked predominance of the stroma. The epithelium is reduced to a very thin network of basaloid cells.

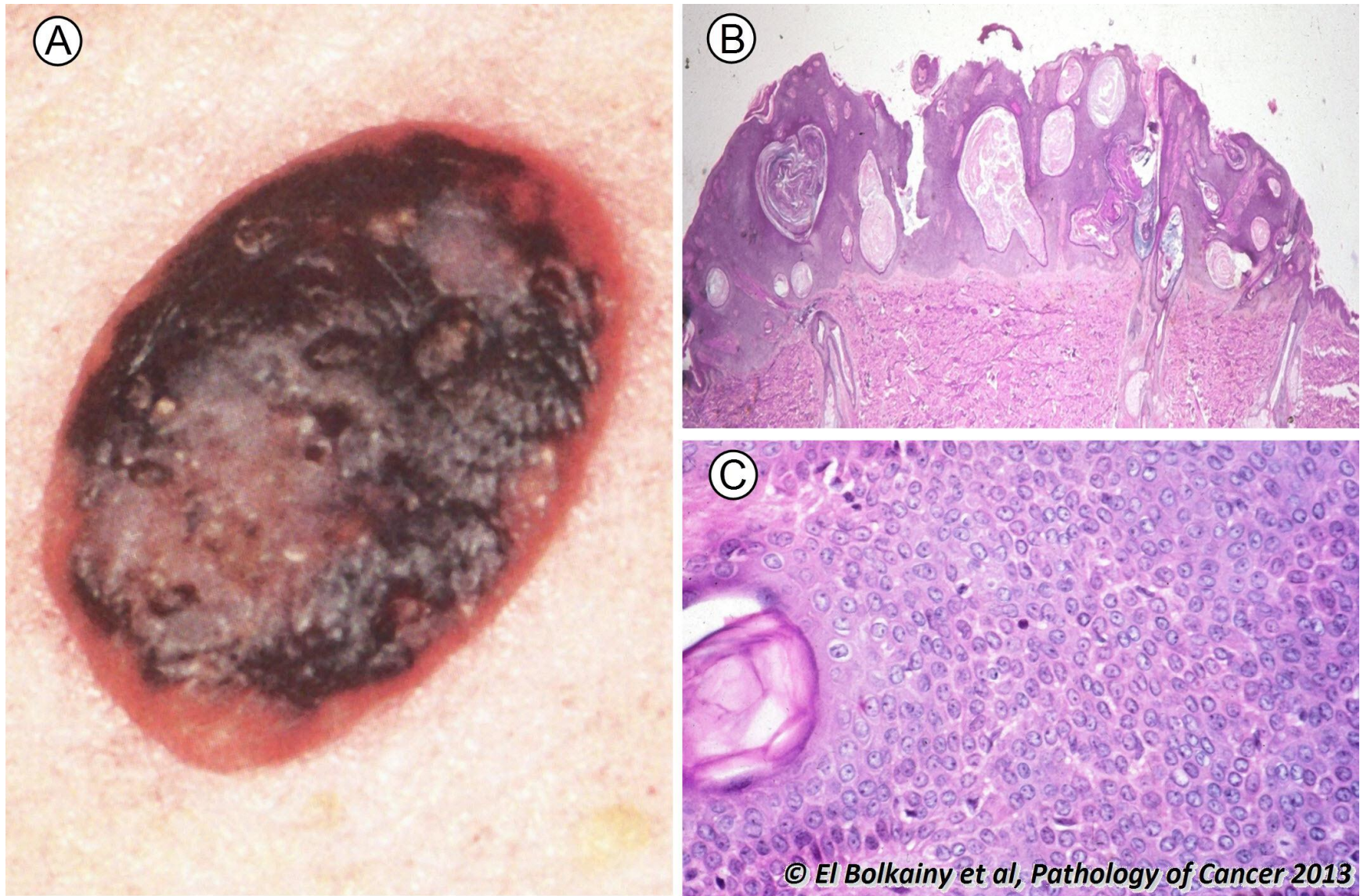
22.10 Adenoid variant of basal cell carcinoma, histology.



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**Picture 22-10** Adenoid variant of basal cell carcinoma, histology. Some of the epithelial groups contain a central lumen. Such a change does not influence biological behavior.

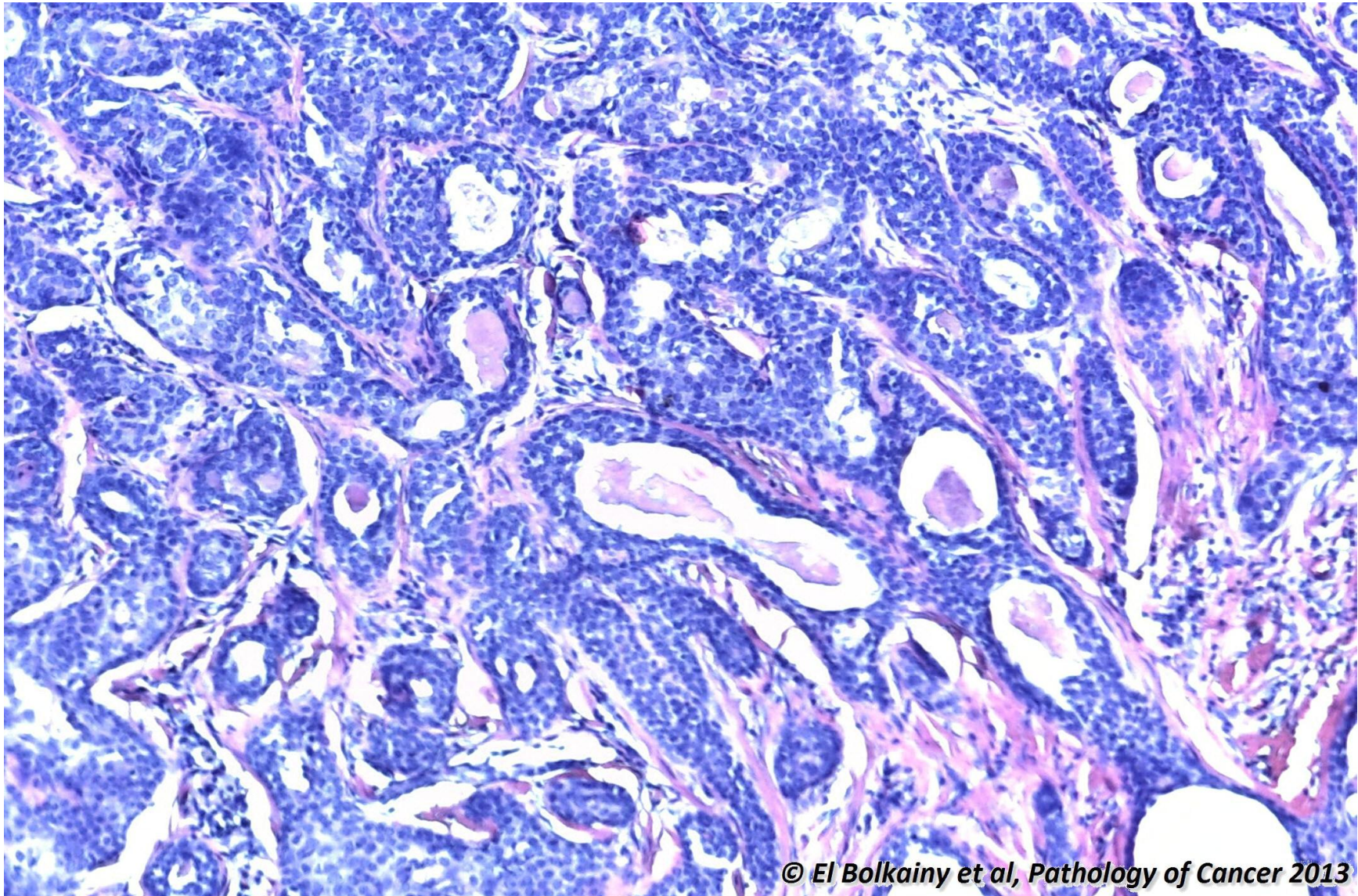
22.11 Seborrheic keratosis.



**Picture 22-11** Seborrheic keratosis. **A** Gross appearance, a sharply defined, dark keratotic lesion. **B** Histology, the superficial pattern is characteristic. There is hyperplasia of keratinocytes with surface hyperkeratosis and keratin cysts. **C** High power.

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22.12 Trichoblastoma, histology.



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**Picture  
22-12**

**Trichoblastoma, histology.** A rare benign follicular neoplasm, usually, simulates basal cell carcinoma. However, it is distinguished by absence of palisading retraction artifacts and presence of lobulation pattern and immunoreactivity of its stroma to CD34.

22.13 Solar keratosis.



**Picture 22-13**

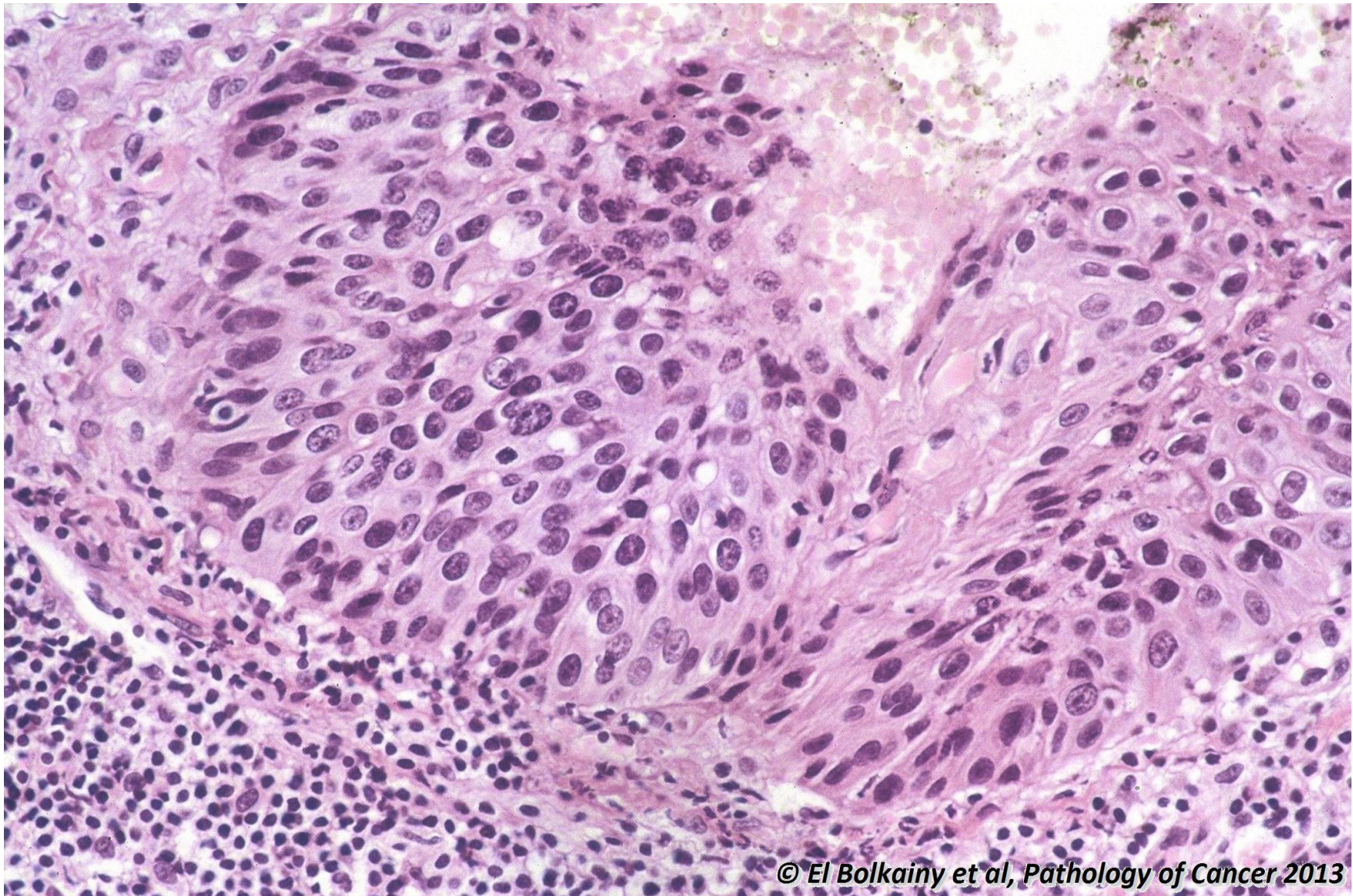
**Solar keratosis.** A and B Gross features. Multiple scaly erythematous lesions. C Histology, Hyperplasia and mild atypia of keratinocytes, alternating parakeratosis, elongation of rete ridges. There is actinic basophilic degeneration of dermal collagen (elastosis)(not shown).

22.14 Squamous carcinoma in situ (Bowen disease), low power histology.



**Picture 22-14** Squamous carcinoma in situ (Bowen disease), low power histology. Malignancy is confined to the epidermis and hair follicles, but basement membrane is intact and no stromal invasion.

22.15 Squamous cell carcinoma in situ (Bowen disease), high power histology.

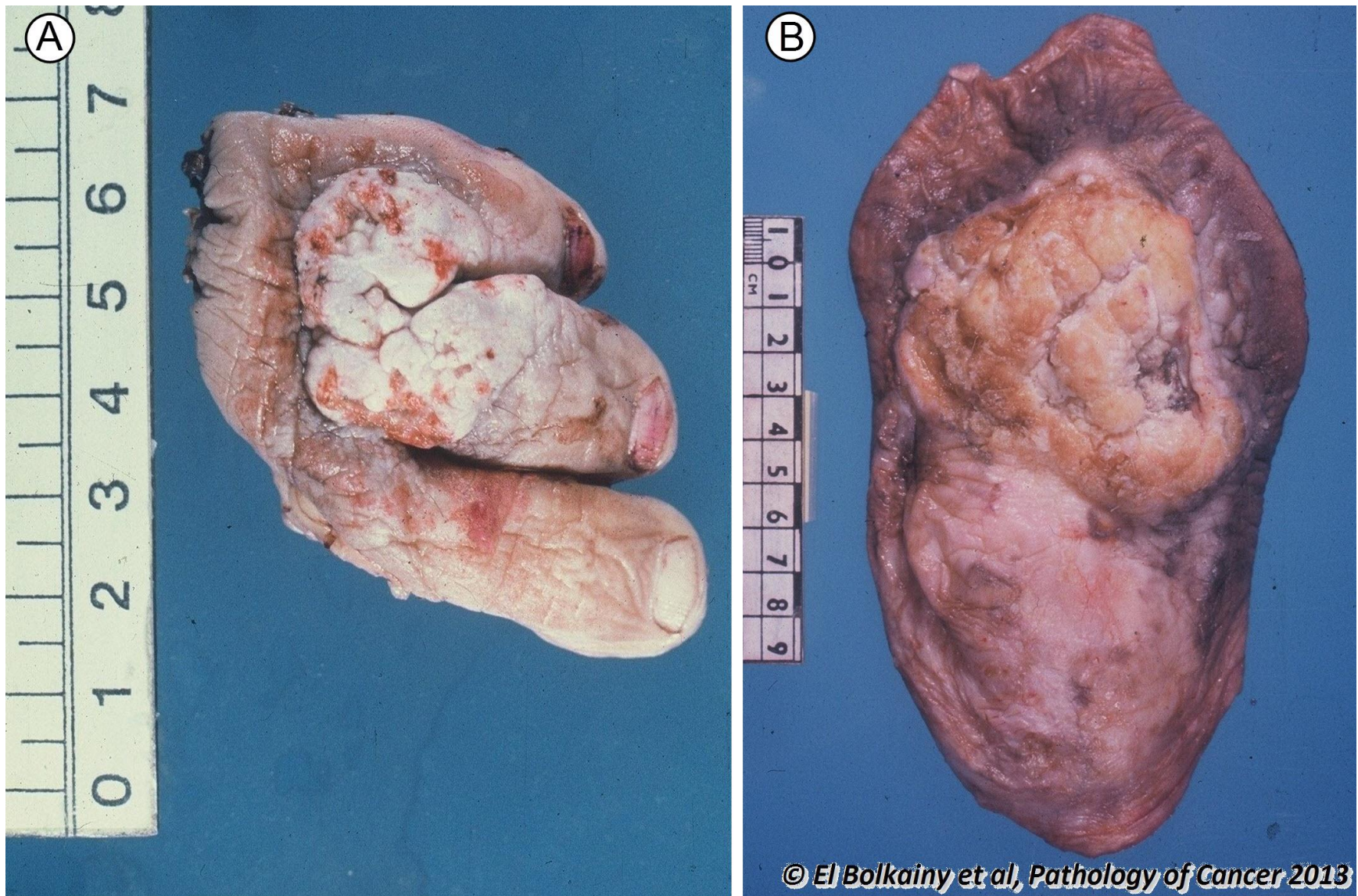


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**Picture 22-15** Squamous cell carcinoma in situ (Bowen disease), high power histology. There is increase of epidermal thickness by malignant keratinocytes with pleomorphic nuclei, frequent mitosis, confluent parakeratosis, loss of cell polarity, but basement membrane is intact.



22.16 Squamous cell carcinoma, skin of toes, gross features.



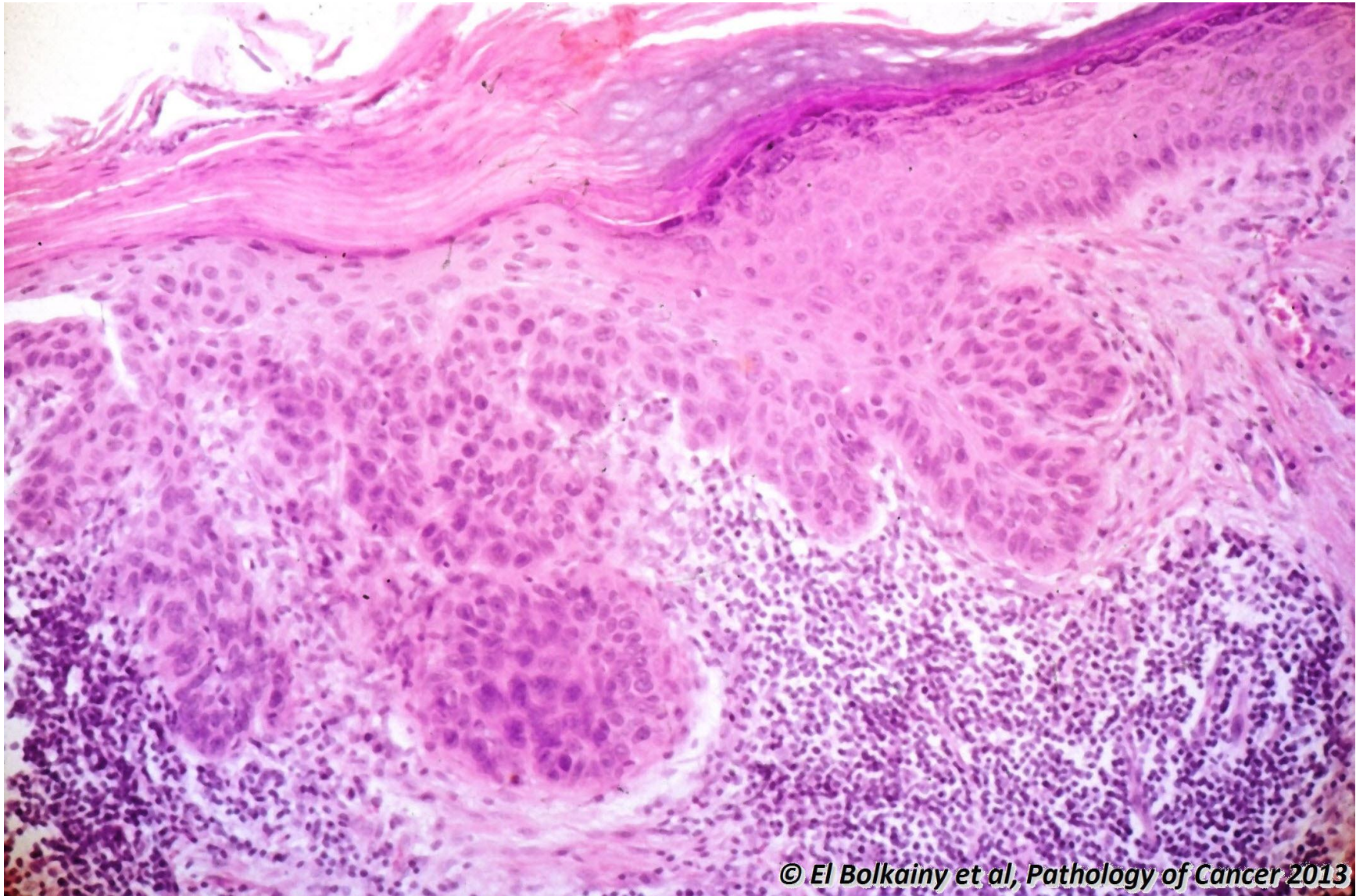
**Picture 22-16** Squamous cell carcinoma, skin of toes, gross features. The tumor appears whitish in color due to keratin content, with ulceration, everted edge and invasion of the stroma.

22.17 Squamous cell carcinoma of parotid region, gross features.



**Picture 22-17** Squamous cell carcinoma of parotid region, gross features. An ulcerated multinodular tumor with raised everted edge.

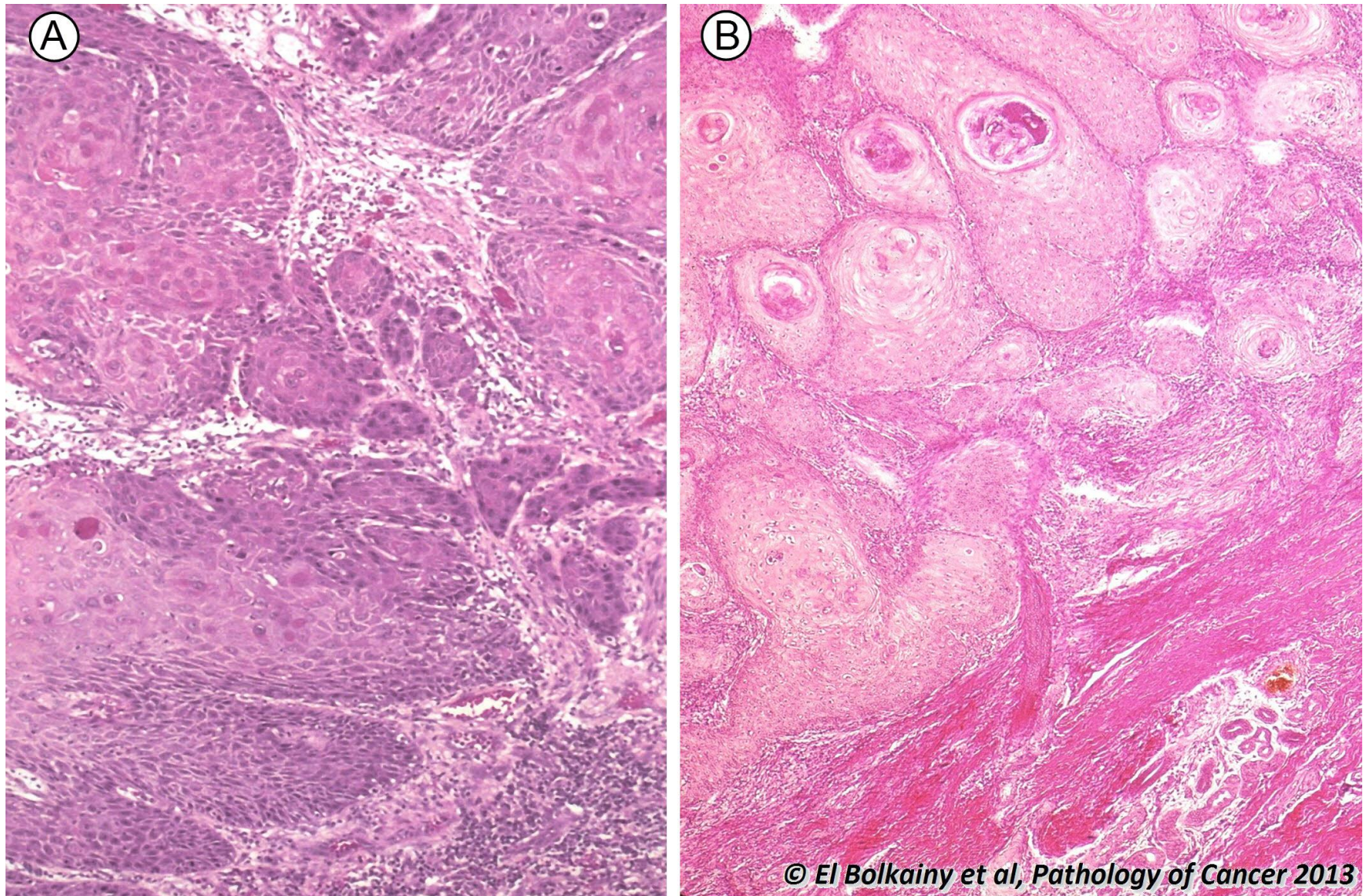
22.18 Microinvasive squamous cell carcinoma, histology.



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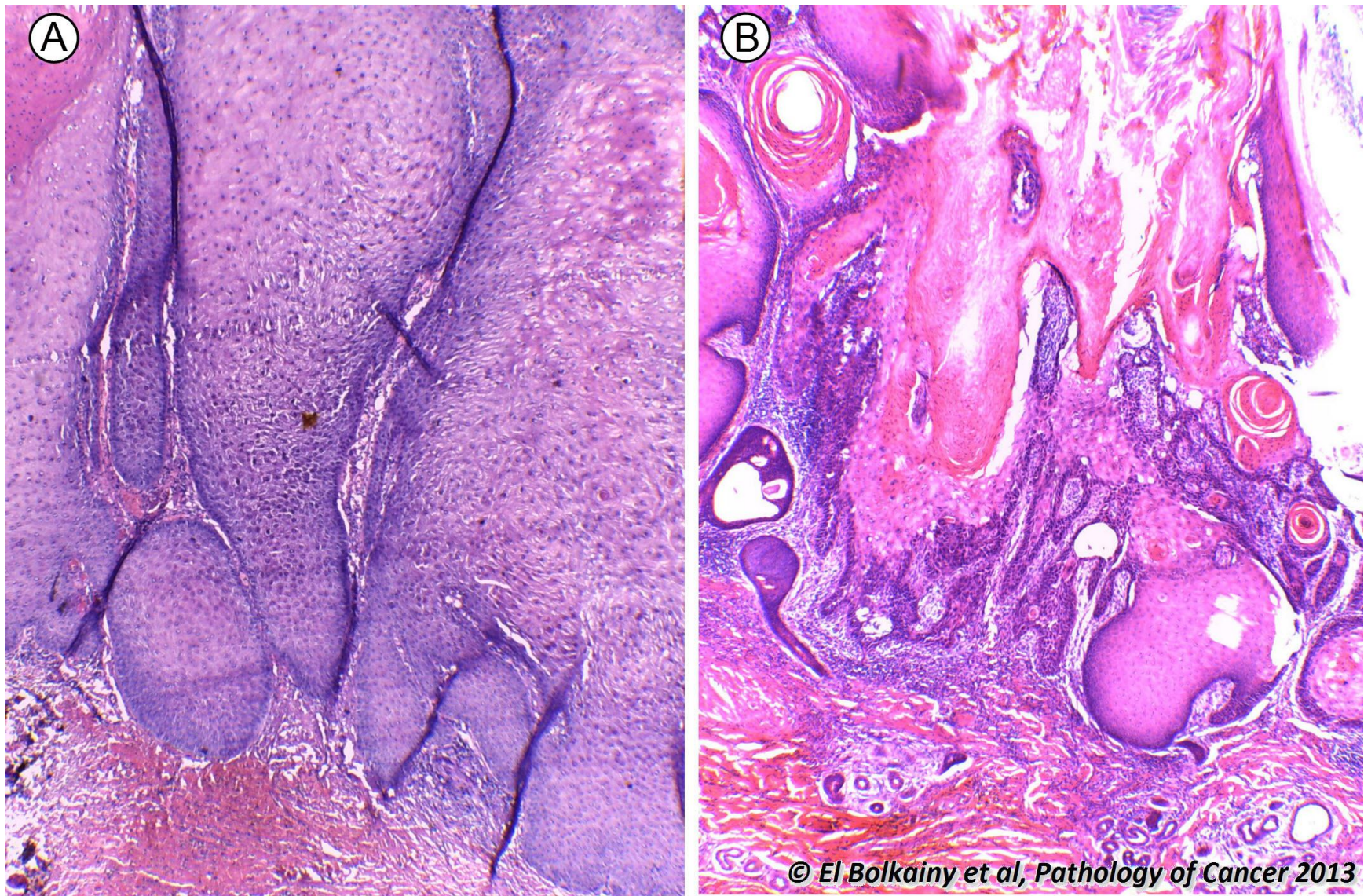
**Picture 22-18** Microinvasive squamous cell carcinoma, histology. There is invasion of basement membrane, but, very limited spread in upper dermis (< 1 mm).

22.19 Invasive squamous cell carcinoma, histology.



**Picture 22-19** Invasive squamous cell carcinoma, histology. **A** and **B** Malignant keratinocytes are arranged in irregular groups with pointed angles, eosinophilic cytoplasm with intercellular bridges and cell nests. There are cytologic features of anaplasia, active mitosis and invasion of stroma.

22.20 Verrucous squamous cell carcinoma, histology.



**Picture 22-20** Verrucous squamous cell carcinoma, histology. A and B The pattern is filamentous or peg-shaped with bulbous end with mild anaplasia and marked keratinization. It is a locally-malignant non-metastasizing carcinoma.

22.21 Keratoacanthoma, gross features.

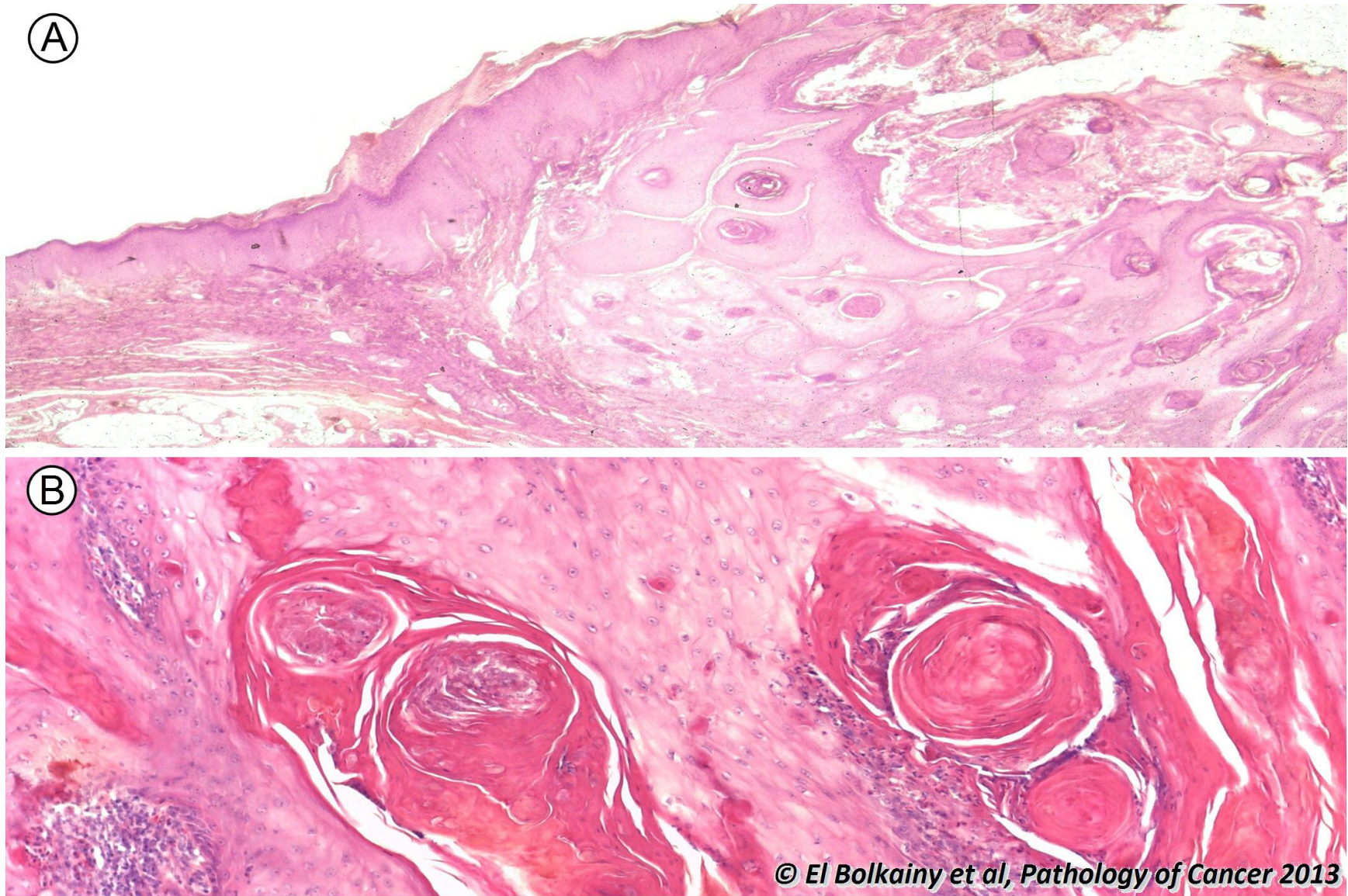


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**Picture  
22-21**

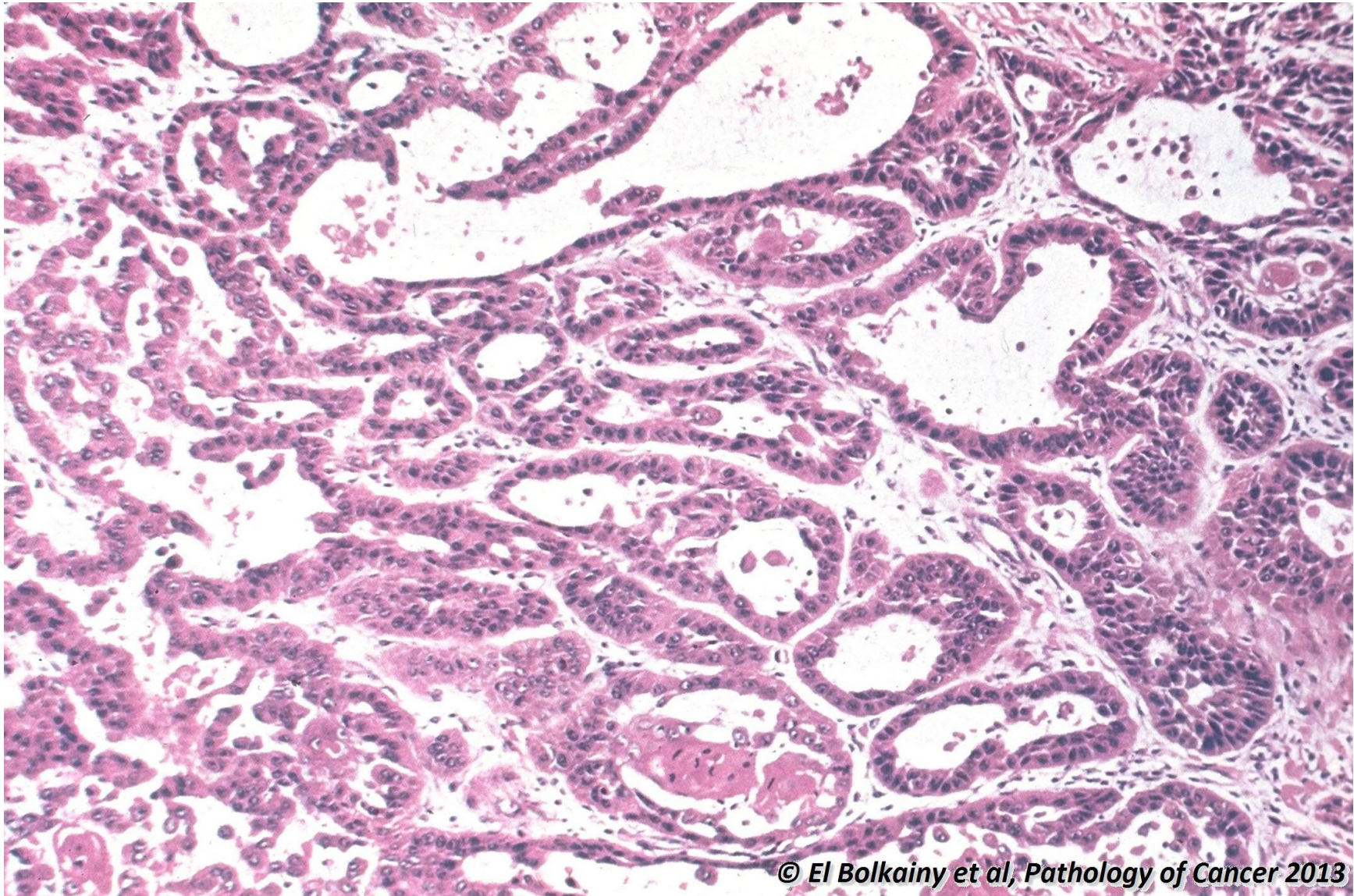
**Keratoacanthoma, gross features.** A nodular skin tumor with central ulcer (crater) filled with keratin.

22.22 Keratoacanthoma, histology.



**Picture 22-22** Keratoacanthoma, histology. **A** Low power, the pattern is exoendophytic with central crater filled with keratin and overhanging shoulders of normal epidermis. **B** Squamous cells are well-differentiated with pushing well-defined lower margin. It is considered a squamous neoplasm with self-limited and regressing growth.

22.23 Acantholytic squamous cell carcinoma, histology.

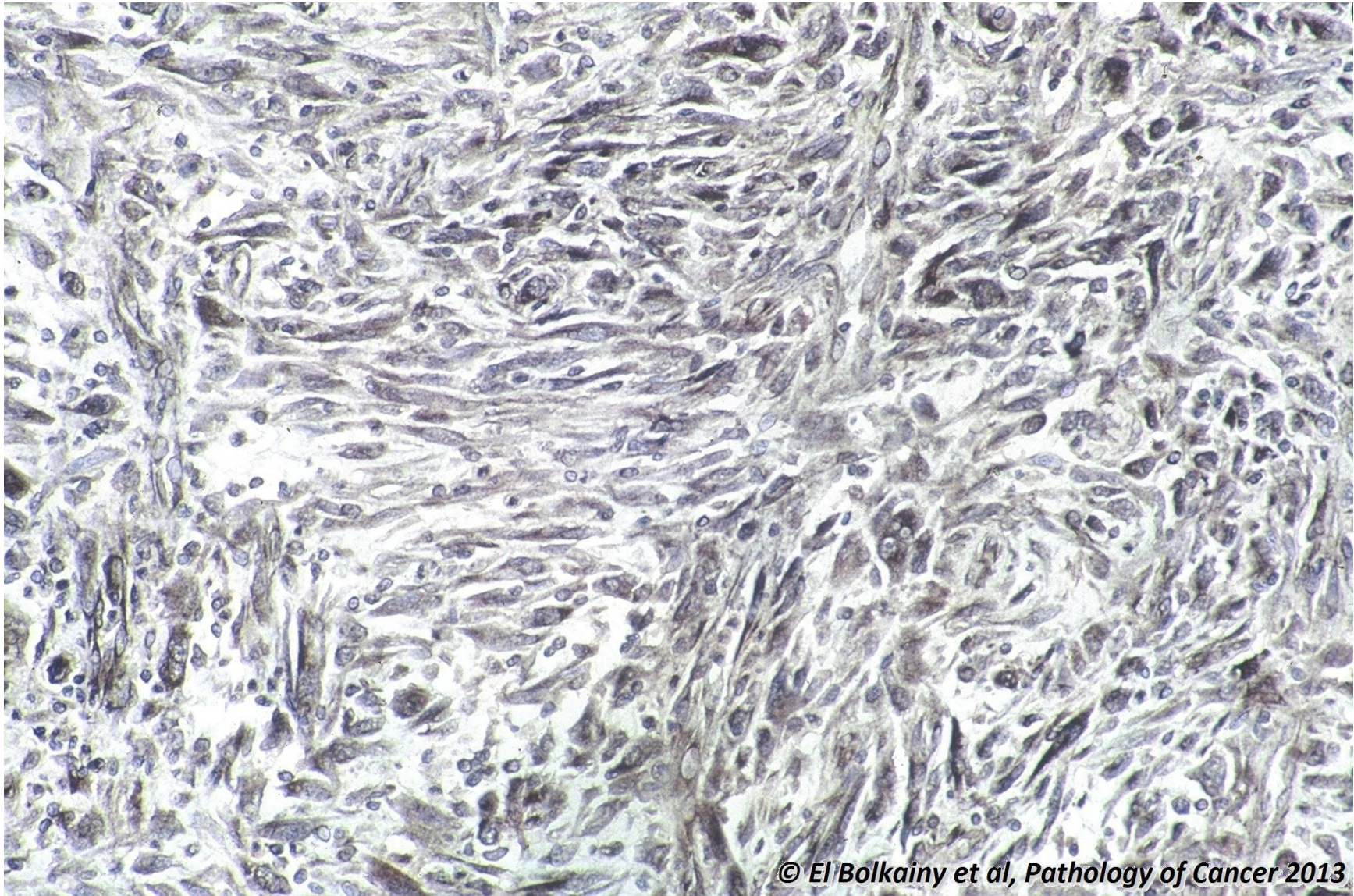


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**Picture 22-23** Acantholytic squamous cell carcinoma, histology. The central part of the neoplastic groups are lost due to degeneration (acantholysis) creating a gland-like lumen which may be misdiagnosed as adenocarcinoma.



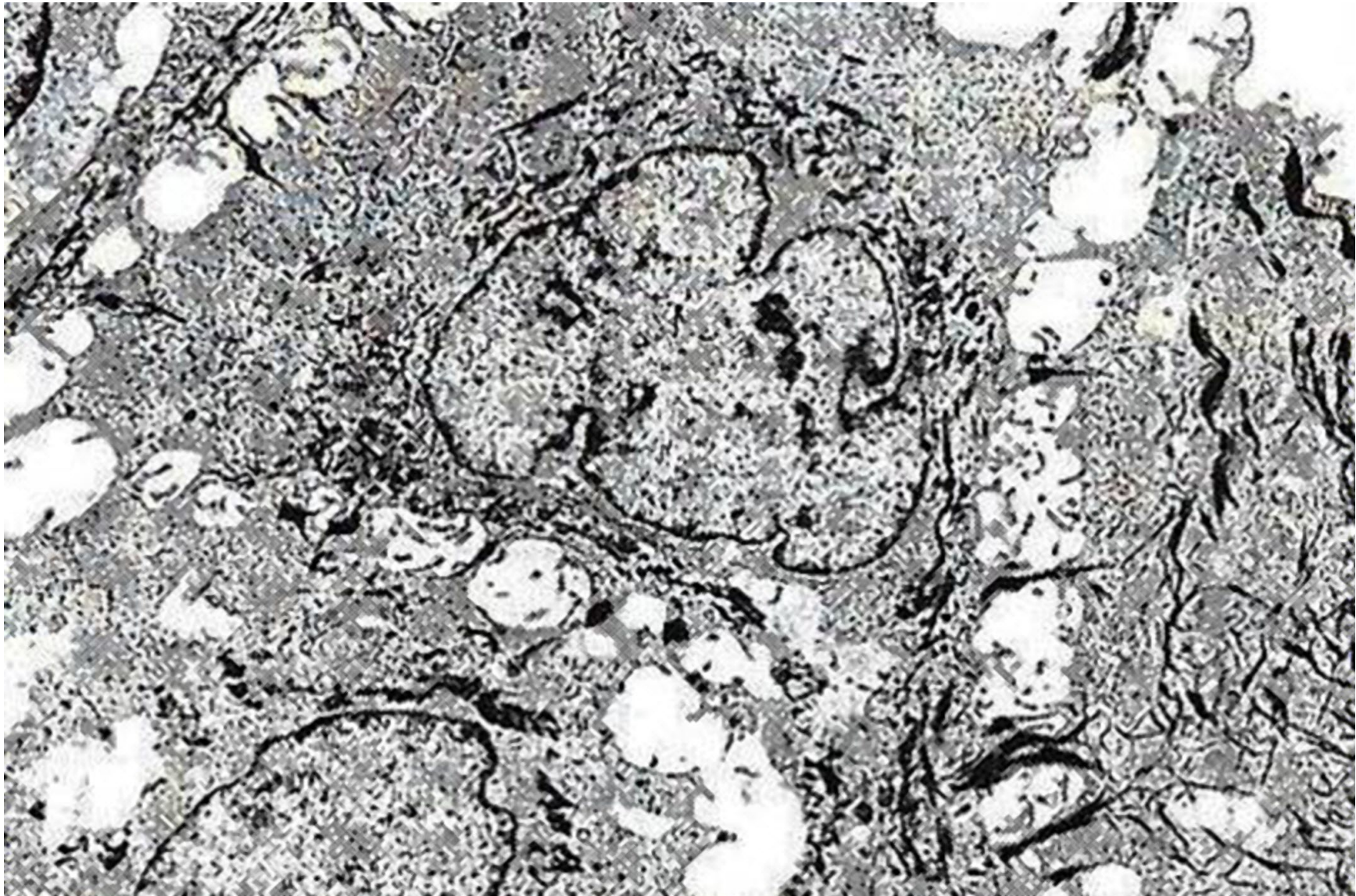
22.24 Sarcomatoid squamous cell carcinoma, immunohistochemistry.



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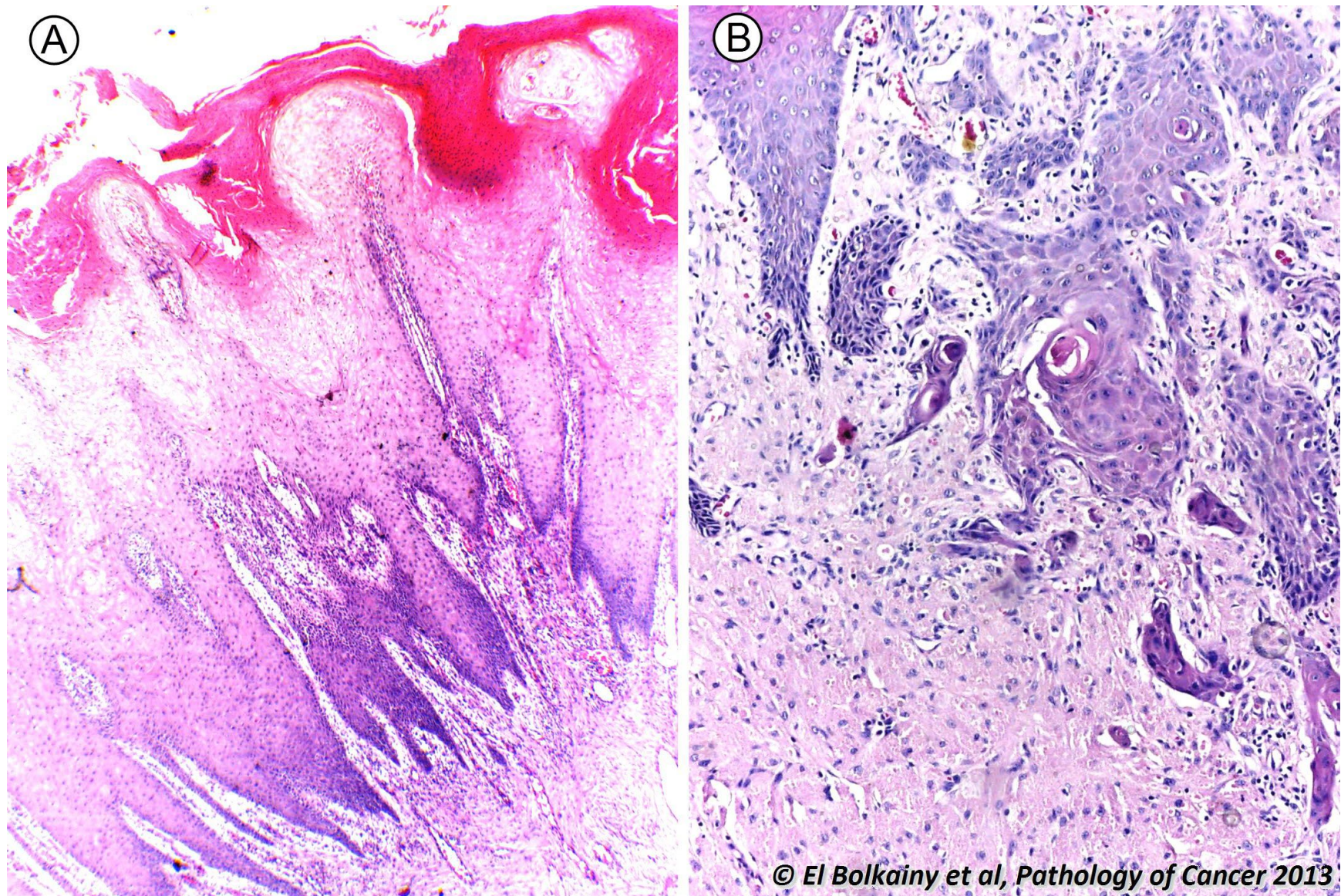
**Picture 22-24** Sarcomatoid squamous cell carcinoma, immunohistochemistry. This aggressive variant is characterized by spindle cells with marked anaplasia and mitosis. It is distinguished from sarcoma by IHC to pan CK.

22.25 Squamous cell carcinoma, electron microscopic features



**Picture 22-25** Squamous cell carcinoma, electron microscopic features showing multiple desmosomes attachment structures at cell membrane and cytoplasmic filaments (tonofilaments) attached to desmosomes. (Reproduced with permission, Fletcher CD, 2007).

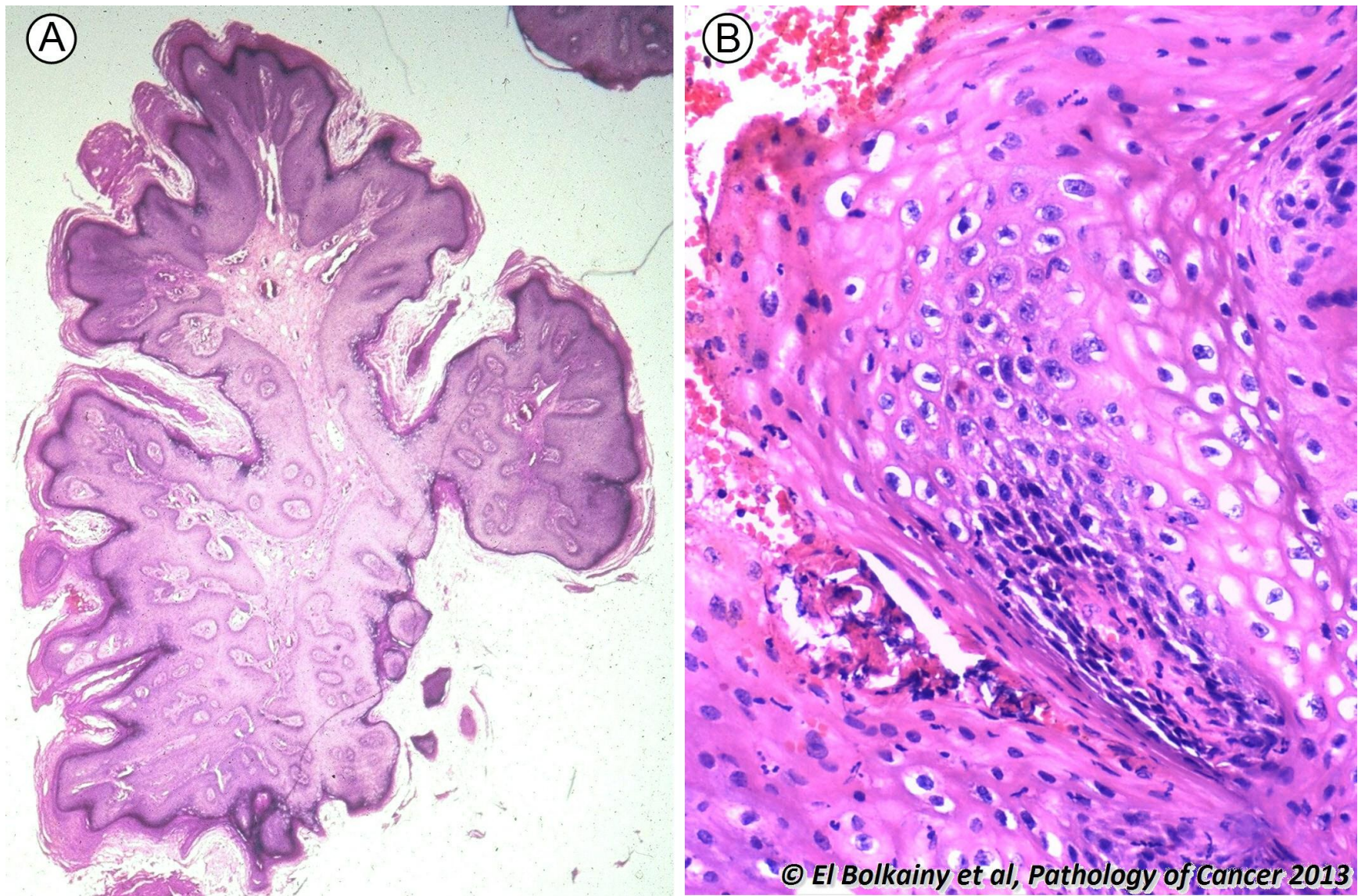
22.26 Pseudoepitheliomatous hyperplasia, histology.



**Picture 22-26** Pseudoepitheliomatous hyperplasia, histology. **A** It is seen associated with chronic leg ulcers. Note the thin elongated rete parallel ridges with cross-bridges. **B** Pseudo-carcinoma in association with granular cell tumor which is evident in lower part of the picture. It is due to secretion of growth factors by the tumor.

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22.27 Condyloma acuminatum.



**Picture 22-27** **Condyloma acuminatum.** **A** This genital wart, a result of HPV infection is characterized by a cauliflower papillary pattern. **B** A high power showing hyperplastic squamous epithelium with characteristic perinuclear vacuoles (koilocytosis) characteristic of viral infection.

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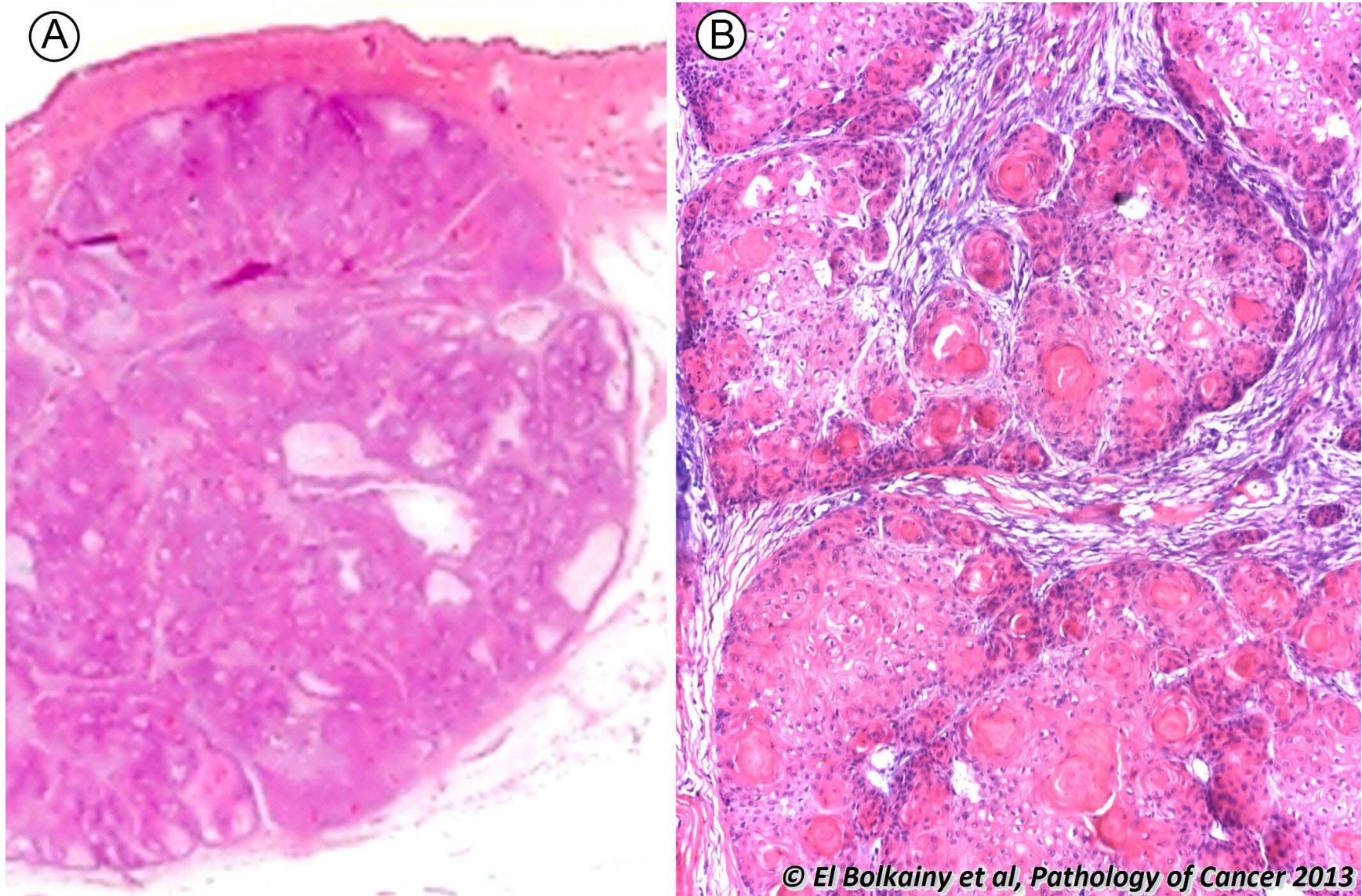
22.28 Verruca vulgaris, histology.



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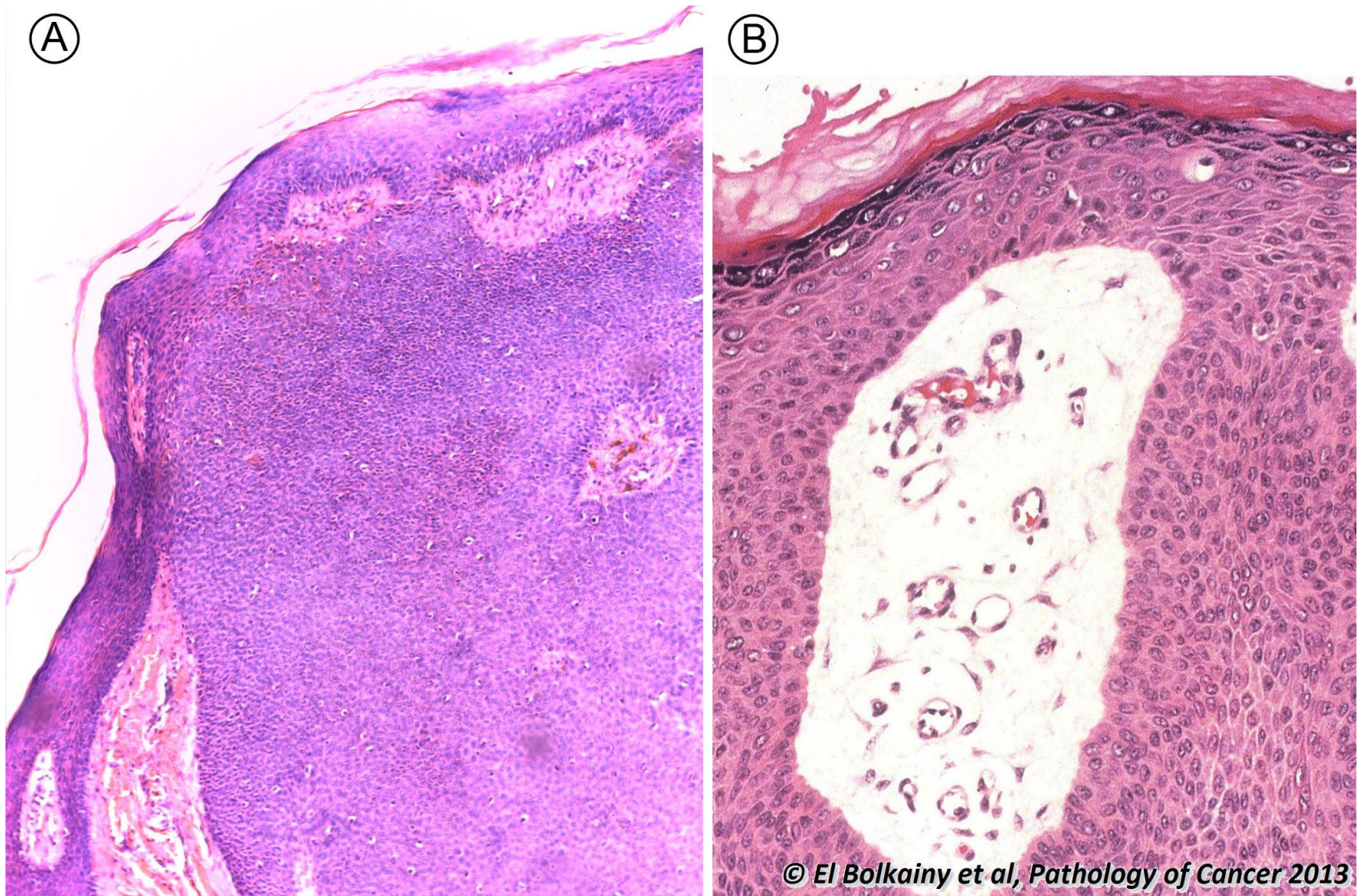
**Picture 22-28** Verruca vulgaris, histology. There is papillomatous proliferation of epidermis with parakeratosis. Cytoplasm shows perinuclear vacuole (koilocytosis), keratohyaline granules and nuclear pallor, all are features of HPV infection.

22.29 Proliferating pilar cyst, histology.



**Picture 22-29** Proliferating pilar cyst, histology. **A** Computer scan x5. A circumscribed solid tumor covered by intact skin. **B** High power. The tumor is formed of lobules of well-differentiated squamous epithelium lacking anaplasia and mitotic activity

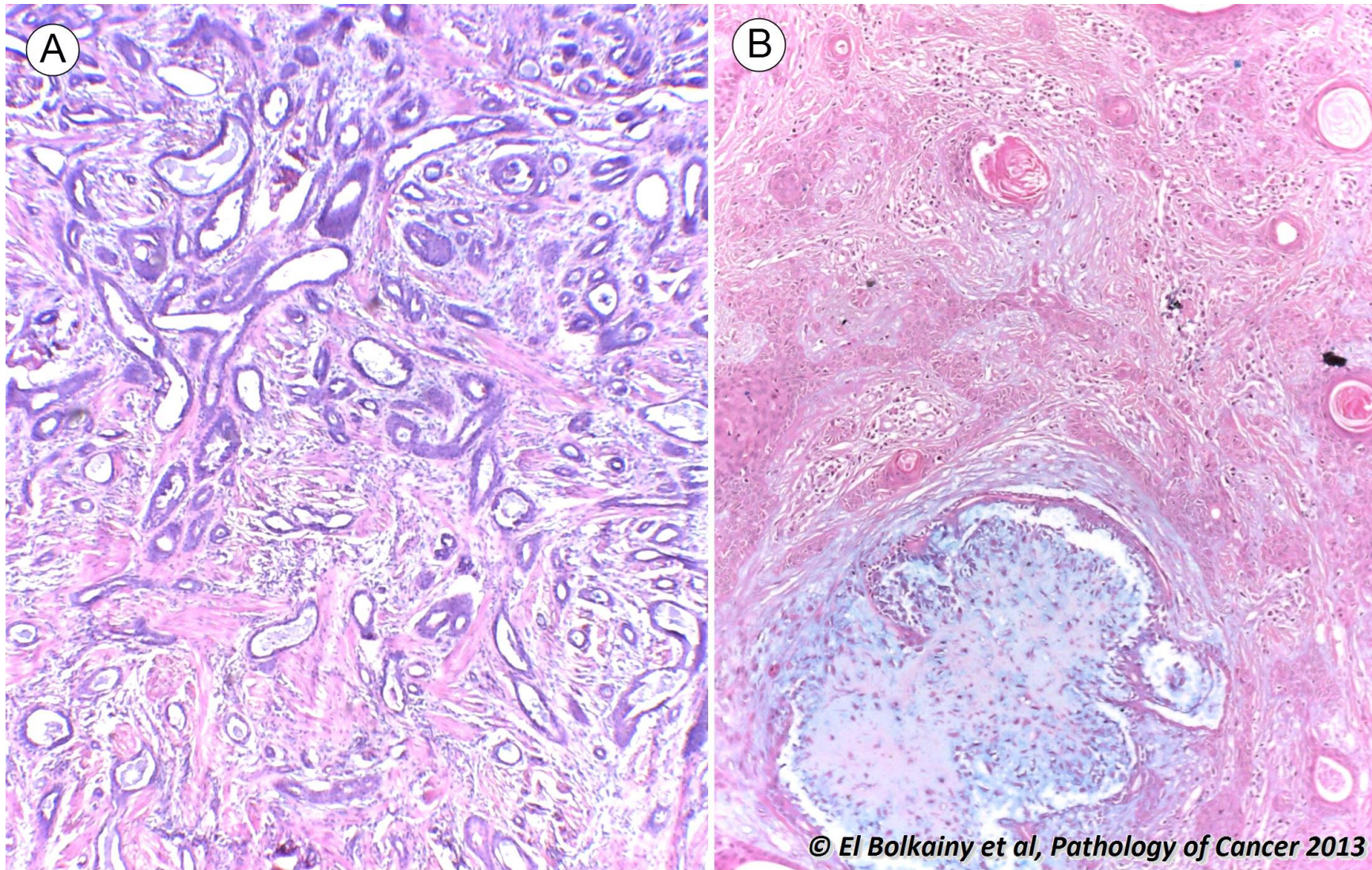
22.30 Poroma, histology.



**Picture 22-30** **Poroma, histology.** This benign tumor arises from intraepidermal portion of sweat gland ducts. **A** Low power. the tumor is solid with basaloid cells connected to surface epithelium. **B** High power. The tumor is merged with epidermal cells.

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22.31 Syringoma, histology.

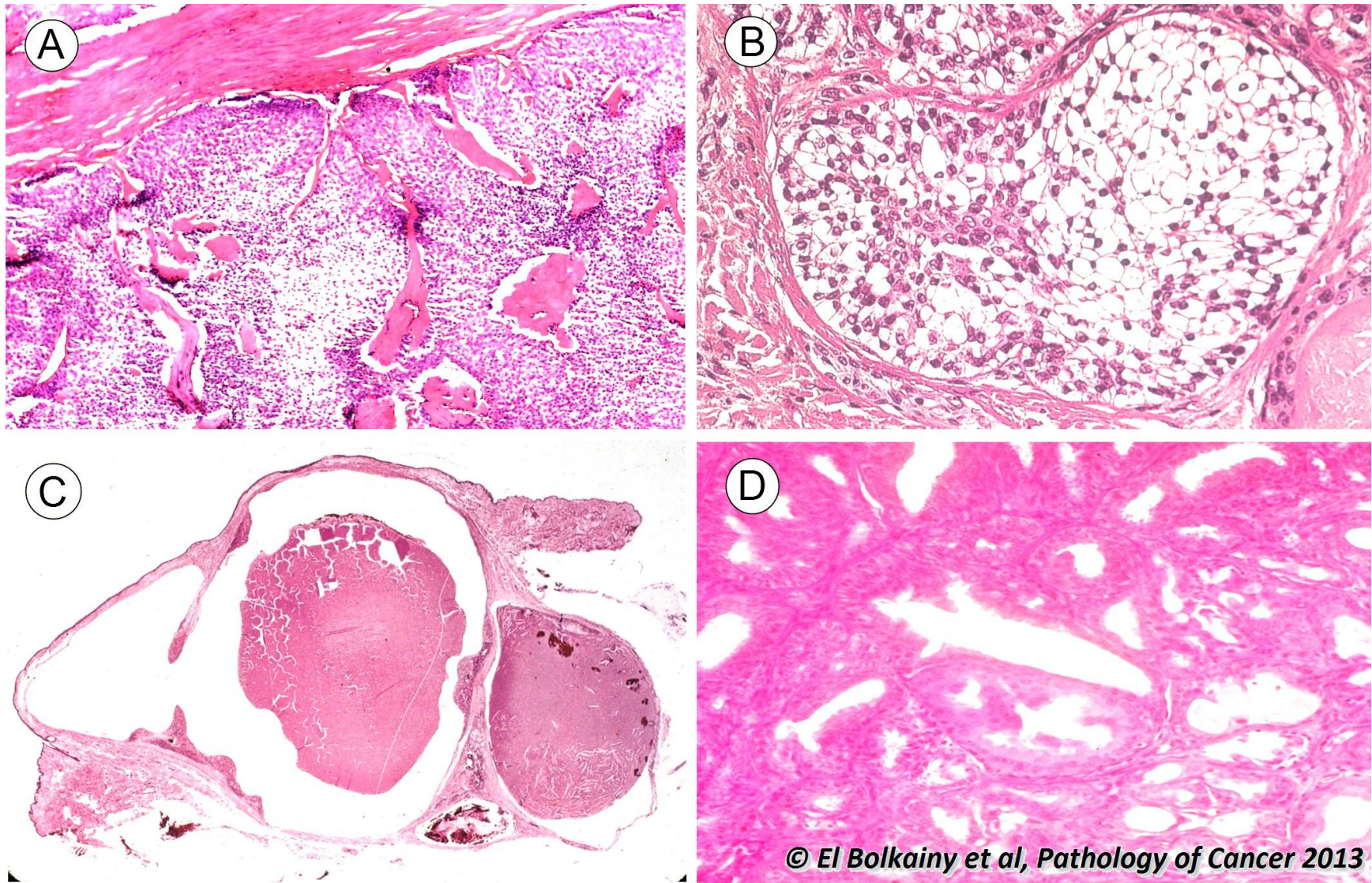


Picture  
22-31

**Syringoma, histology.** This benign tumor arises from the intradermal portion of sweat gland ducts. **A** Classic syringoma showing ductal structures with pointed ends lined by double-layered epithelial and myoepithelial cells. **B** Chondroid syringoma, simulating pleomorphic adenoma of salivary glands, showing epithelial and myoepithelial elements. These contribute to the formation of squamous and chondroid areas respectively.



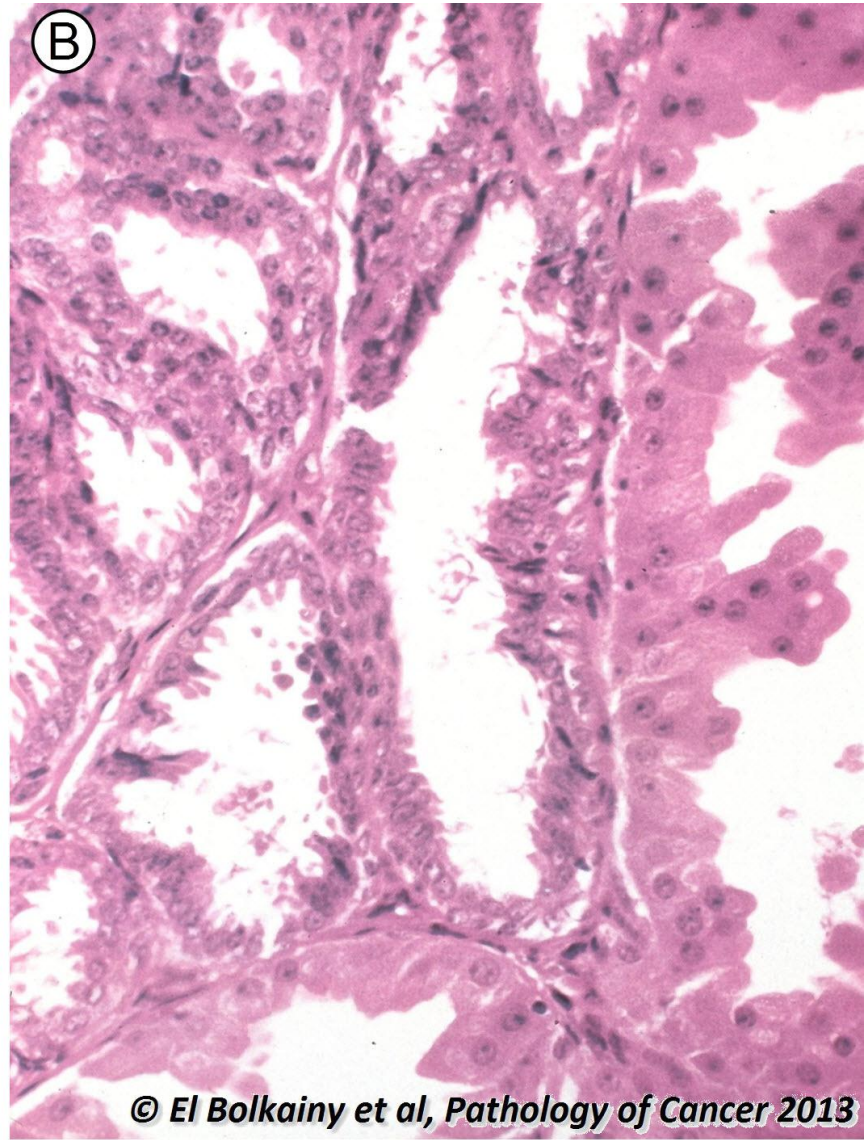
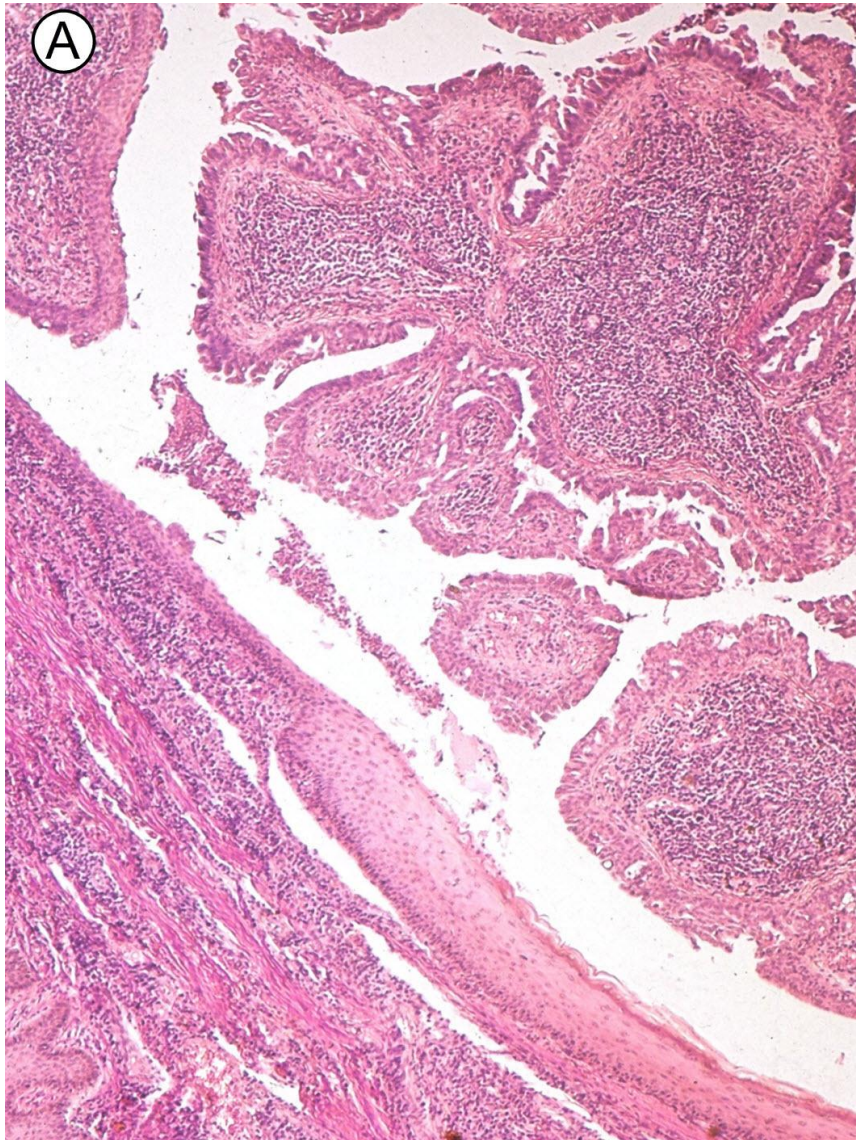
22.32 Hidradenoma, histology.



**Picture 22-32**

**Hidradenoma, histology.** A Benign tumor arising from sweat gland proper. It presents in one of 3 histological forms. **A and B** Solid, It appears as solid dermal nodule composed of multiple cell types ( polyhedral, spindle and clear). **C** Cystic, A well defined dermal lesion lined by cuboidal and flat epithelium. **D** Apocrine, shows eosinophilic cells with decapitation secretion. In all these forms, circumscription and basement membrane material are usually evident.

22.33 Syringocystadenoma papilliferum, histology.

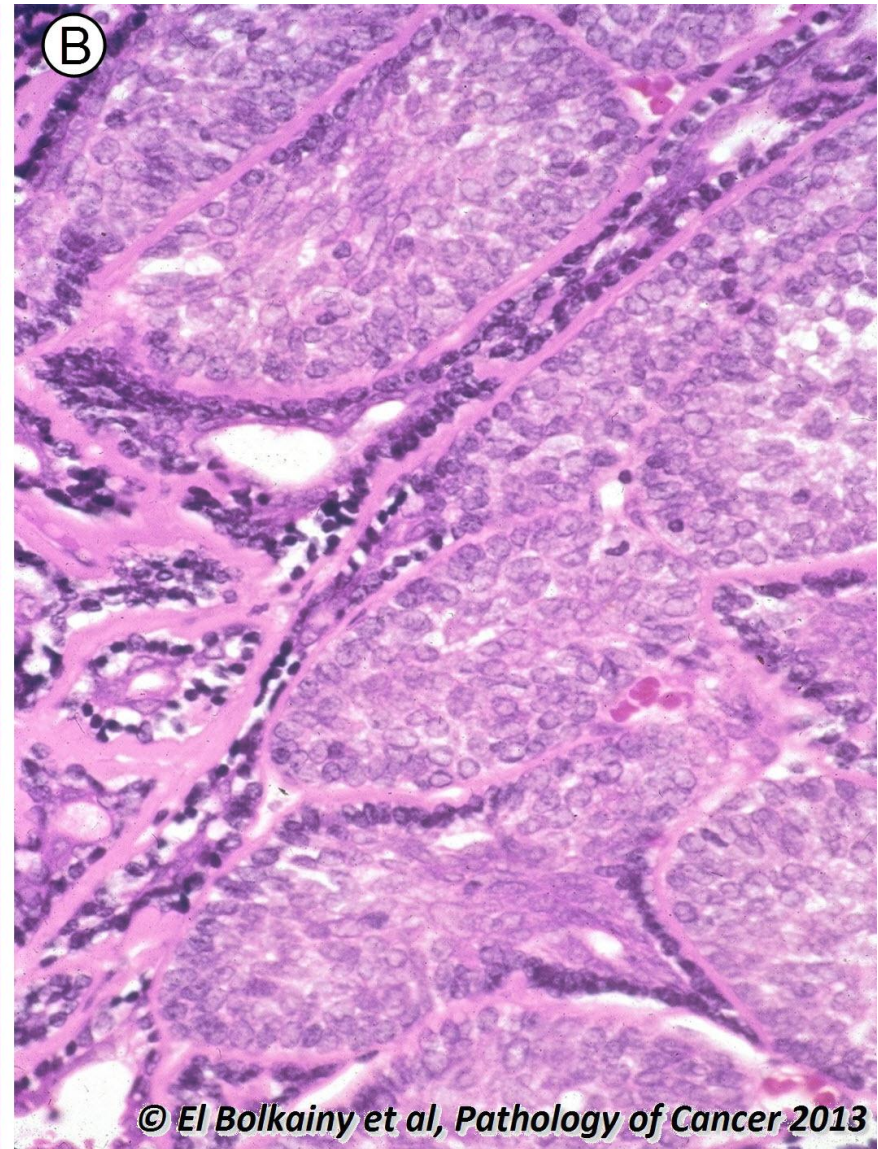
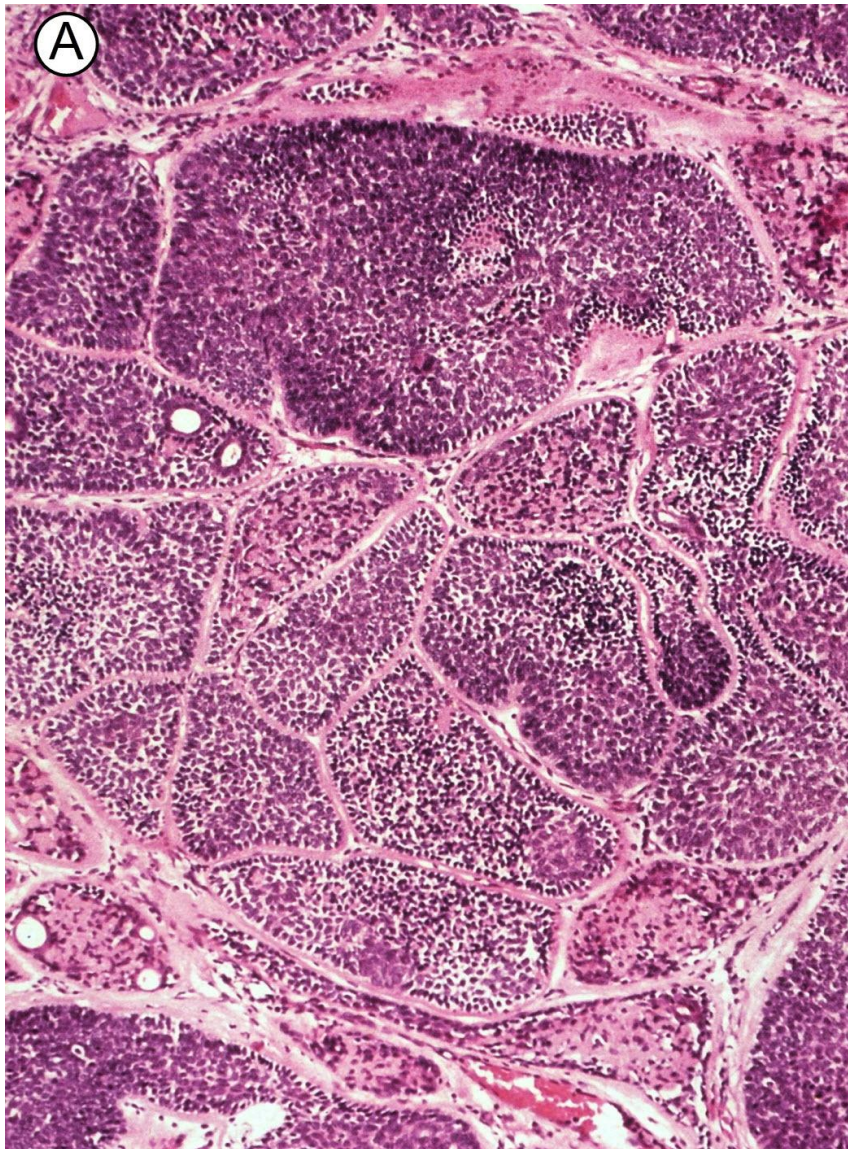


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**Picture 22-33**

**Syringocystadenoma papilliferum, histology.** A and B This tumor is a cystic papillary lesion located in epidermis. A similar tumor (papillary hidradenoma) is located in dermis. Both are lined by apocrine columnar epithelium with decapitation secretion.

22.34 Cylindroma (turban tumor), histology.



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**Picture 22-34** Cylindroma (turban tumor), histology. This benign sweat gland tumor is composed of islands of epithelial cells that fit together like peices of jigsaw puzzle and separate by hyaline sheath (basement membrane). The tumor is biphasic (epithelial and myoepithelial).

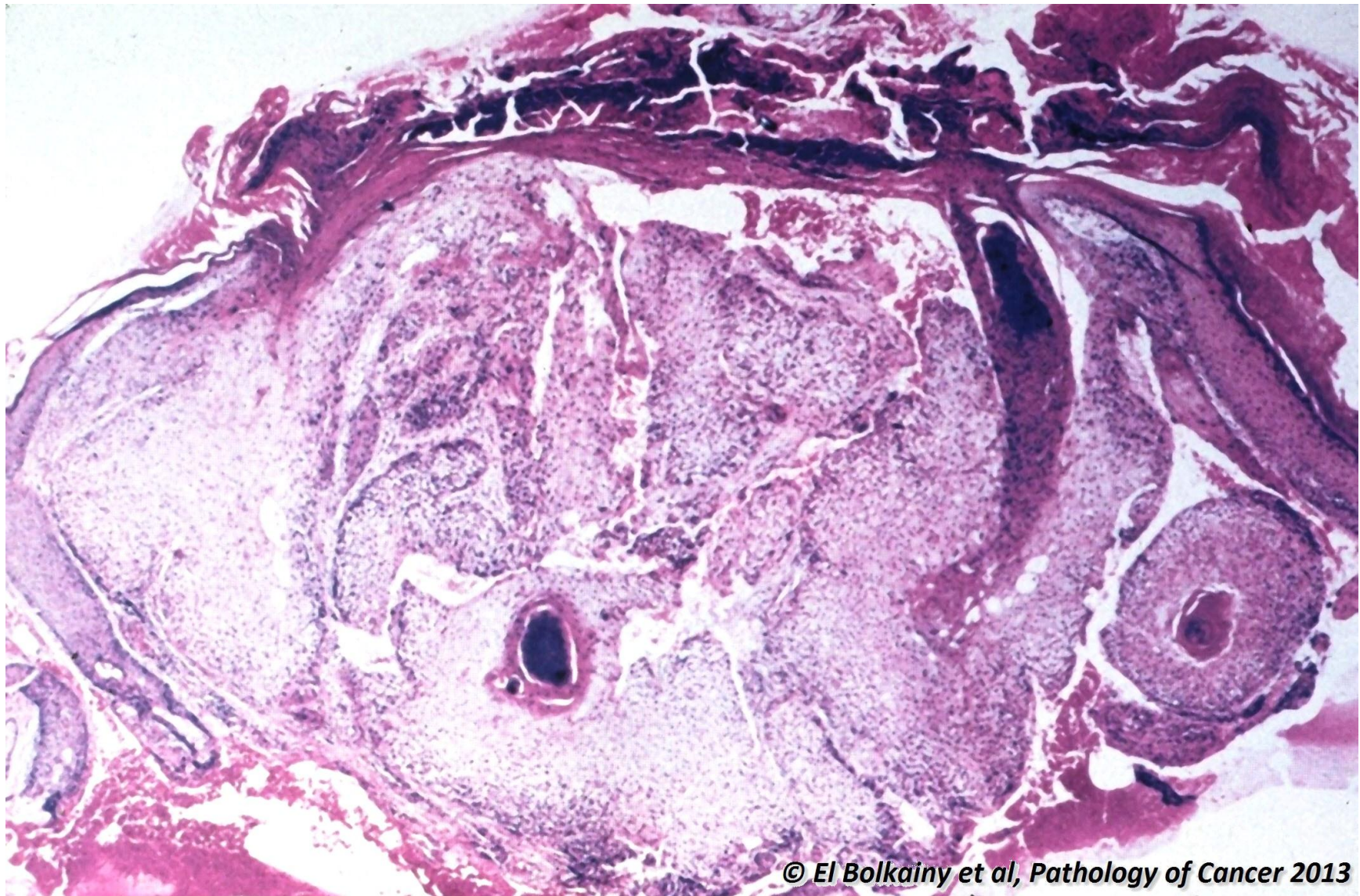
22.35 Sebaceous adenoma, histology.



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**Picture 22-35** Sebaceous adenoma, histology. A well-defined benign tumor attached to epidermis showing biphasic structure of peripheral keratinocytes and central sebaceous clear cells.

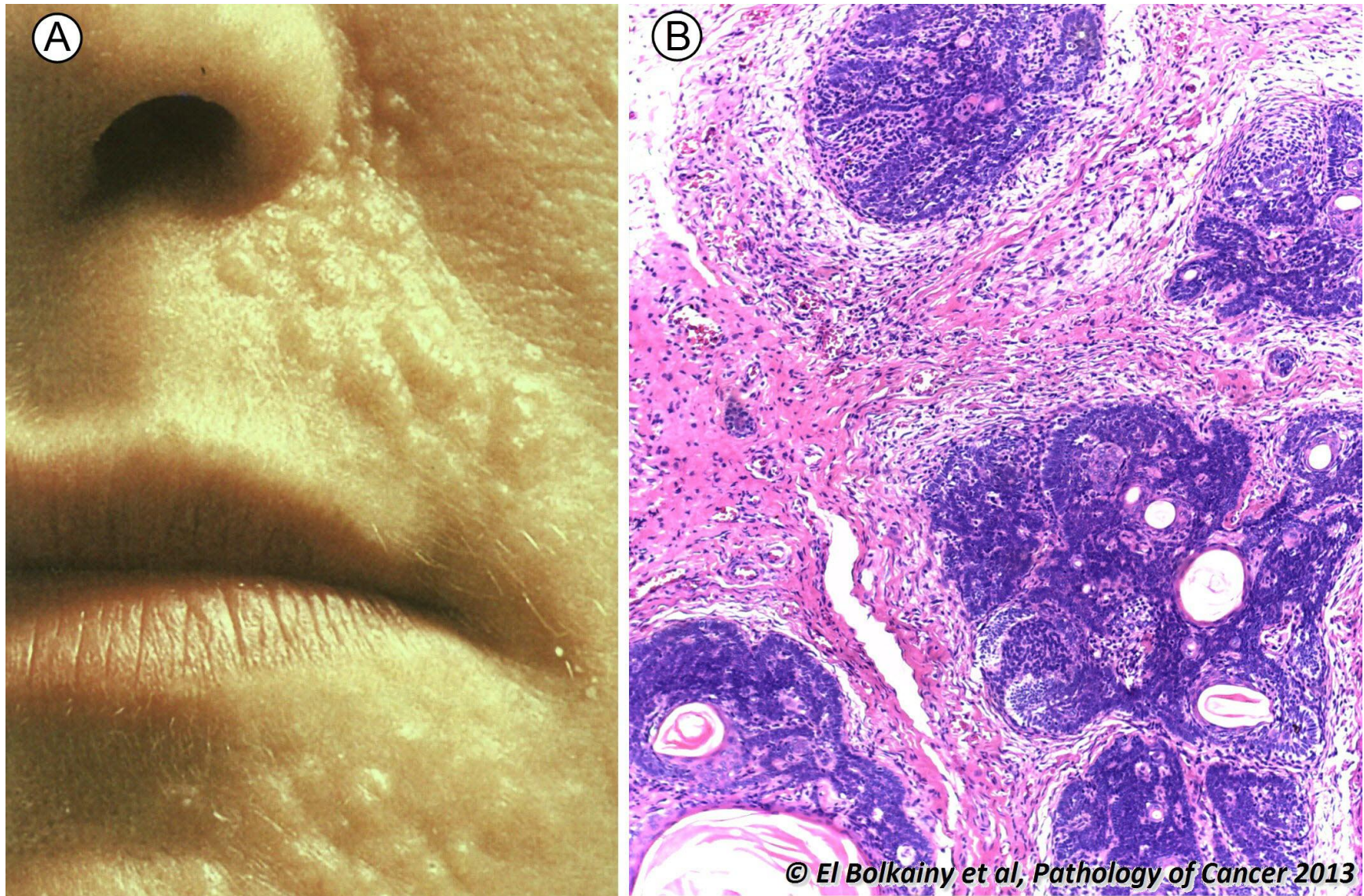
22.36 Trichilemmoma, histology.



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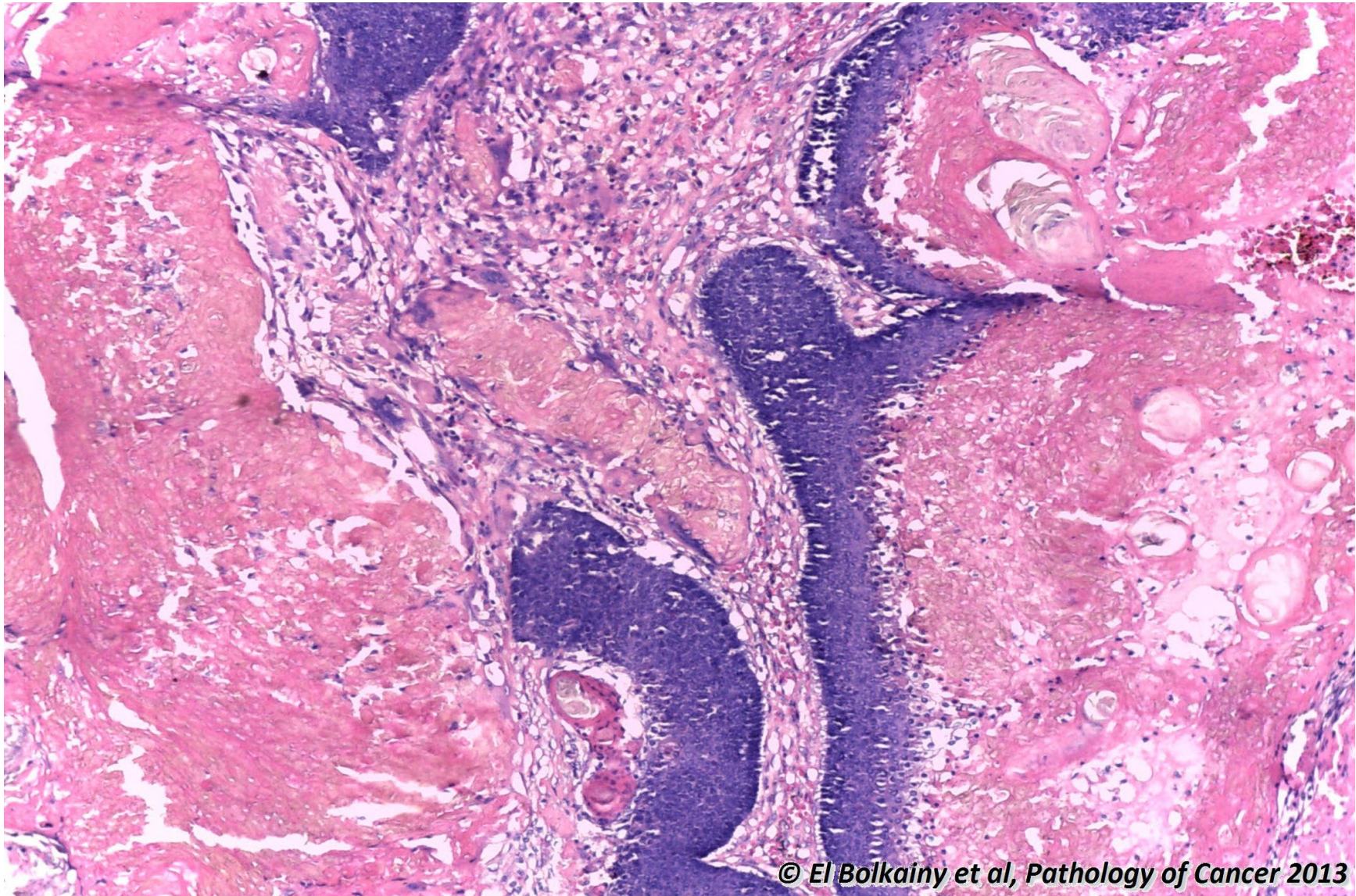
**Picture 22-36** Trichilemmoma, histology. A sharply defined proliferation of cells with clear cytoplasm resembling the outer root sheath of hair follicle. The tumor is closely related to the hair follicle rather than to the epidermis.

22.37 Trichoepithelioma.



**Picture 22-37** Trichoepithelioma. **A** Gross features, small multiple skin nodules. **B** Trichoepithelioma, basaloid cells containing keratinous cysts embedded in a cellular stroma with follicular differentiation in the form of bulbs and papillae.

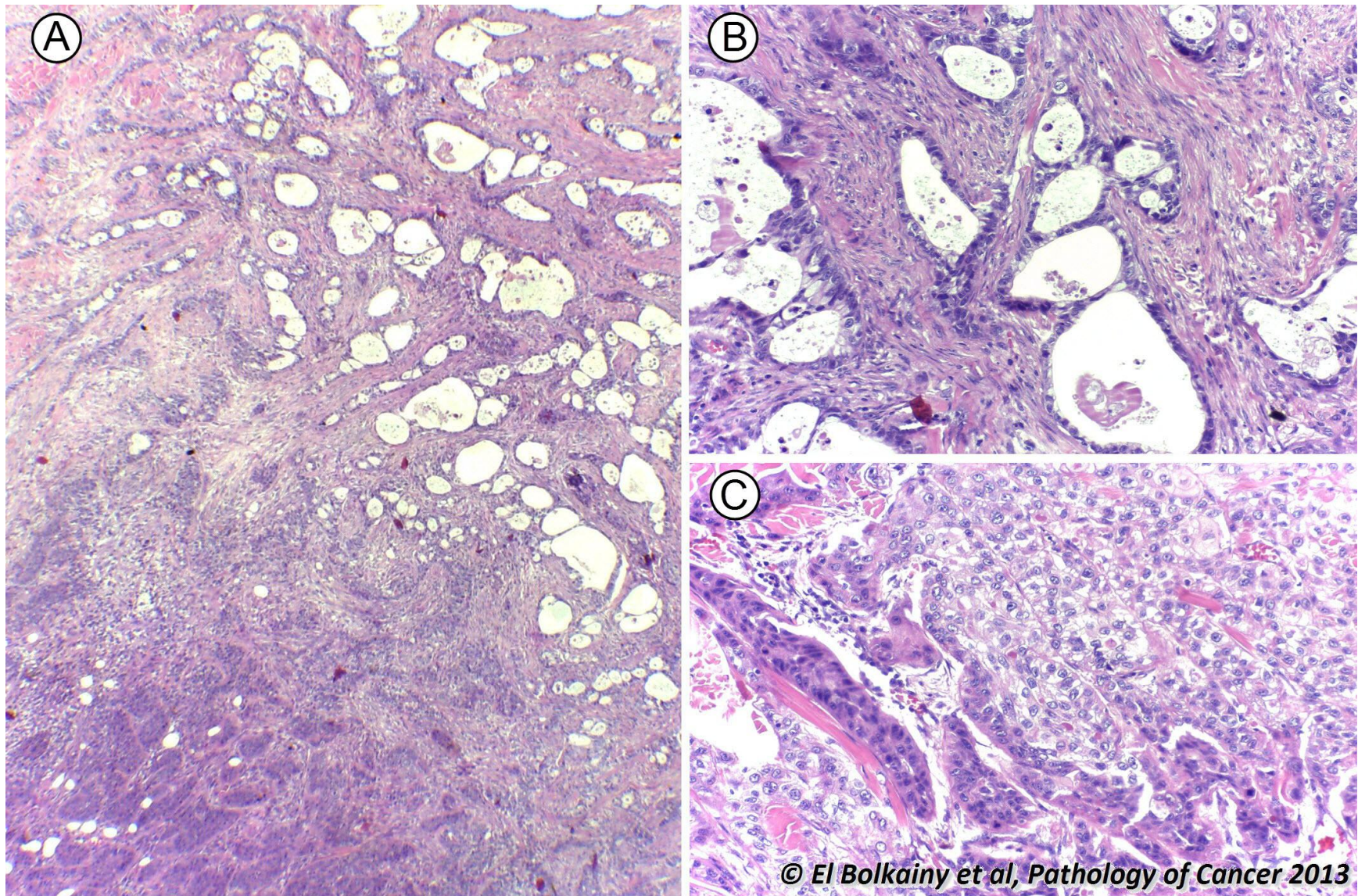
22.38 Pilomatricoma, histology.



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**Picture 22-38** Pilomatricoma, histology. **A** Low power showing a well-defined lesion covered by intact skin. **B** High power, shows characteristic combination of basaloid cells and ghost eosinophilic cells.

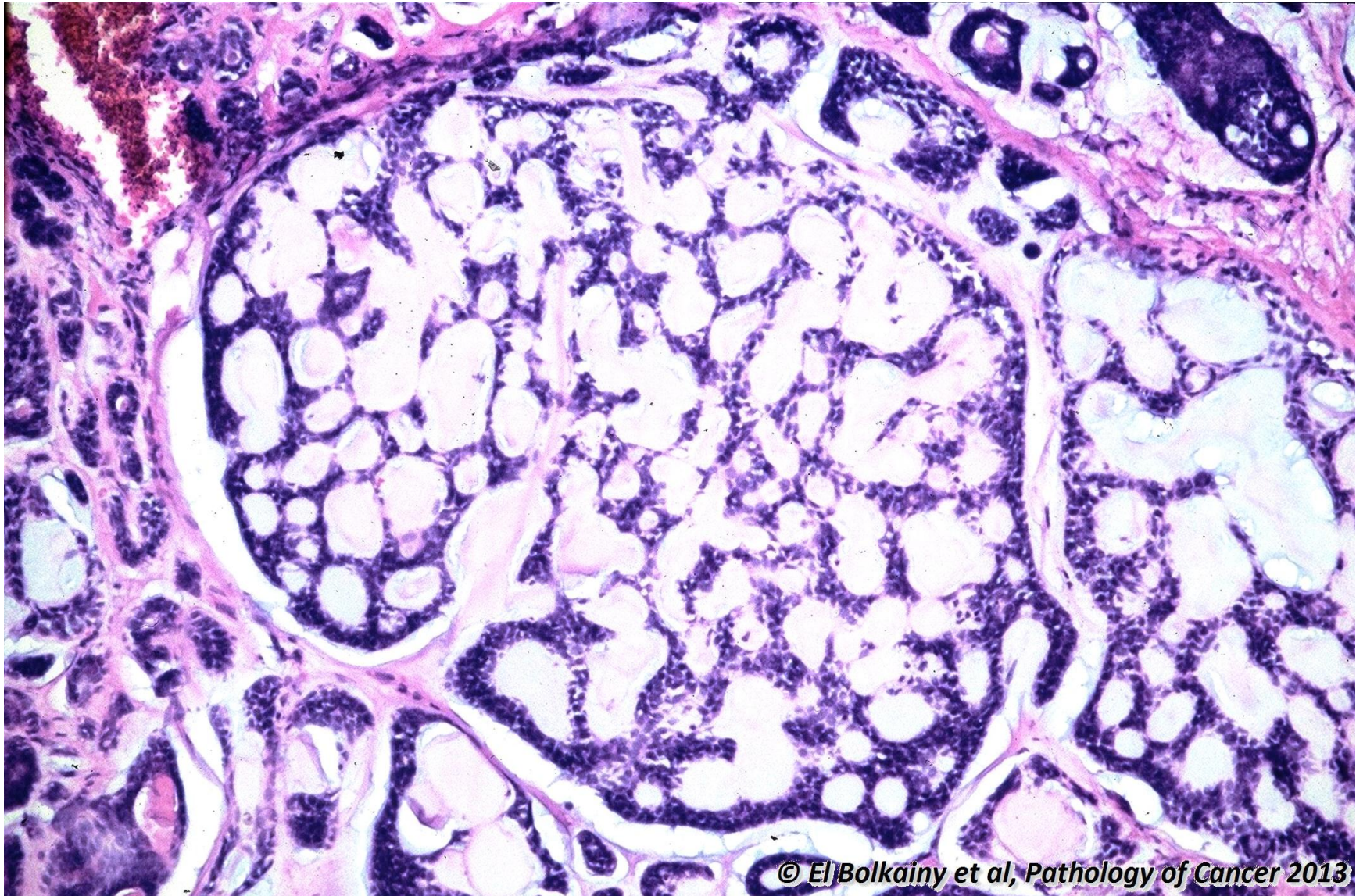
22.39 Microcystic adnexal carcinoma (sweat gland carcinoma), histology.



**Picture 22-39** Microcystic adnexal carcinoma (sweat gland carcinoma), histology. **A** Low power. This invasive tumor has multiphasic structure including glands of variable size, microcysts and areas of squamous differentiation. **B** High power. Glandular differentiation. **C** High power. Squamous differentiation.



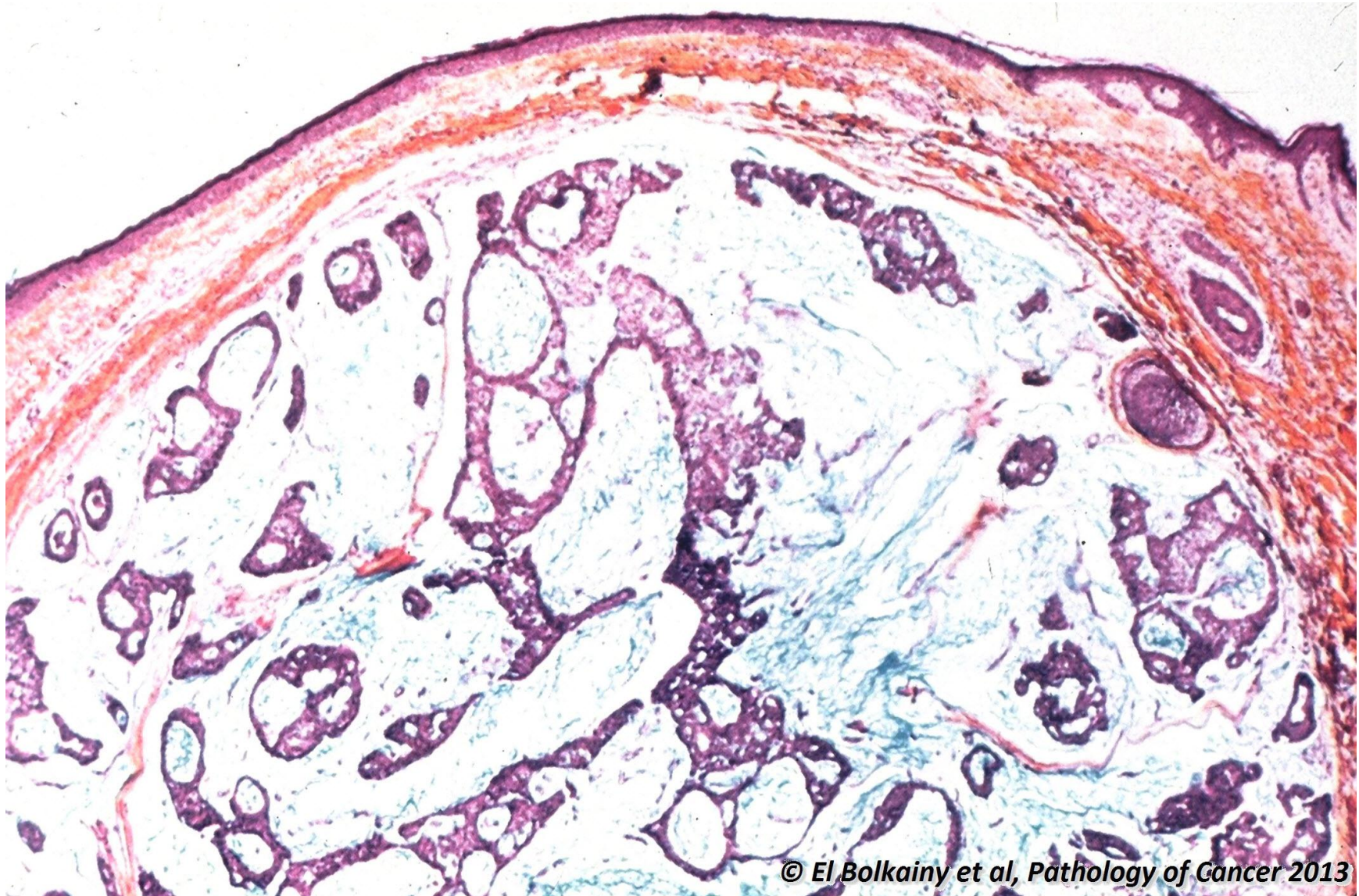
22.40 Adenoid cystic carcinoma of sweat glands, histology.



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**Picture 22-40** Adenoid cystic carcinoma of sweat glands, histology. Note the characteristic cylindromatous pattern with rounded spaces in tumor mass filled with eosinophilic basement membrane material. Perineural invasion is a prominent feature.

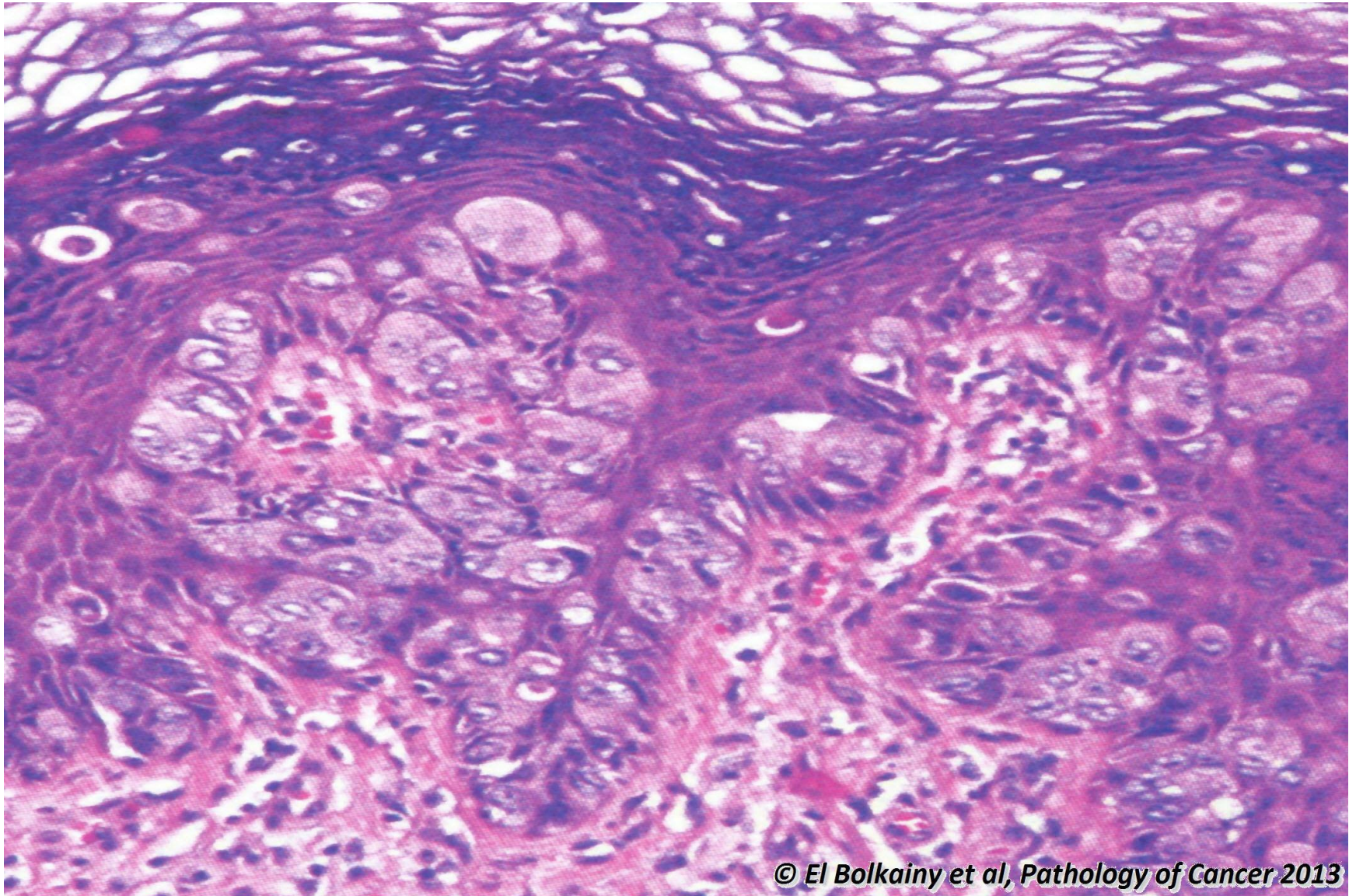
22.41 Mucinous adenocarcinoma of sweat glands, histology.



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**Picture 22-41** Mucinous adenocarcinoma of sweat glands, histology. There are islands of malignant cells in a pool of extracellular mucin.

22.42 Paget's disease of skin, histology.

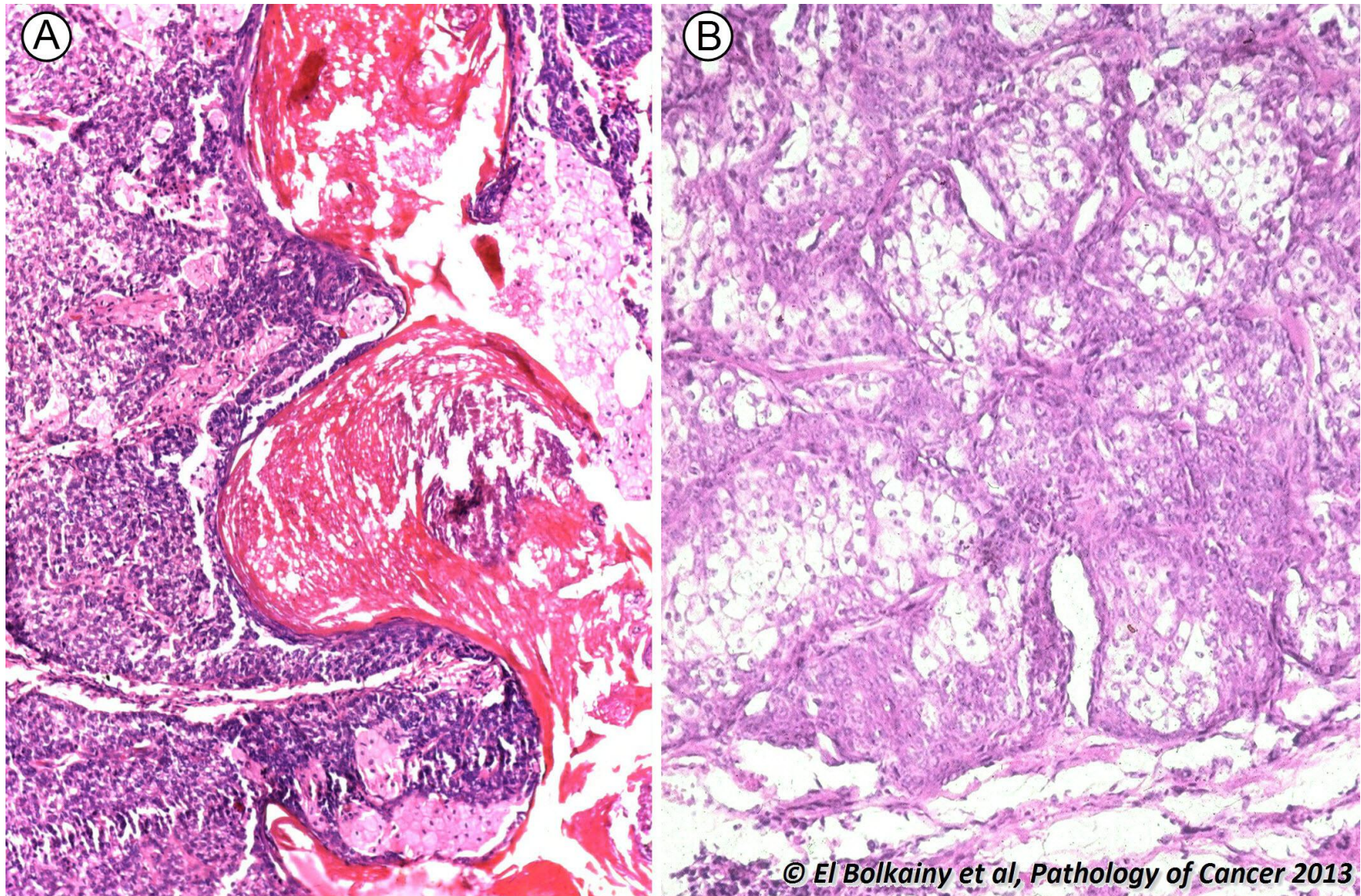


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**Picture  
22-42**

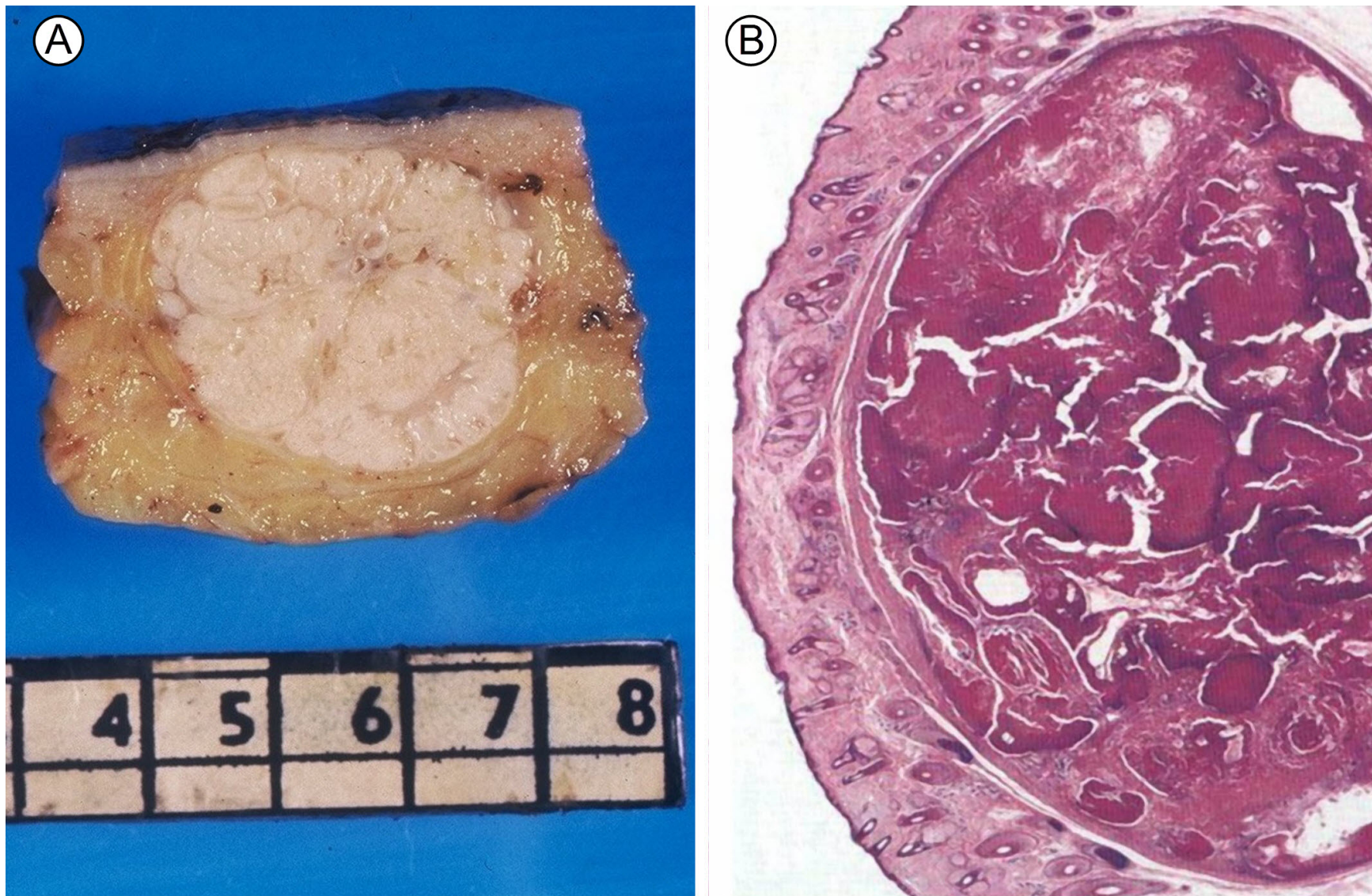
**Paget's disease of skin, histology.** Scattered malignant cells with clear cytoplasm (mucicarmine +) are evident in epidermis. The main differential diagnosis are melanoma and Bowen disease. Paget's disease is positive for CK 7 and CEA but melanoma and Bowen are negative for them. In addition, melanoma is positive for S-100 and Bowen disease for p63.

22.43 Sebaceous carcinoma, histology.



**Picture 22-43** Sebaceous carcinoma, histology. **A** Low power. **B** High power. Malignant tumor with biphasic structure (squamous and sebaceous) showing cellular anaplasia and mitotic activity, as well as, invasion of stroma.

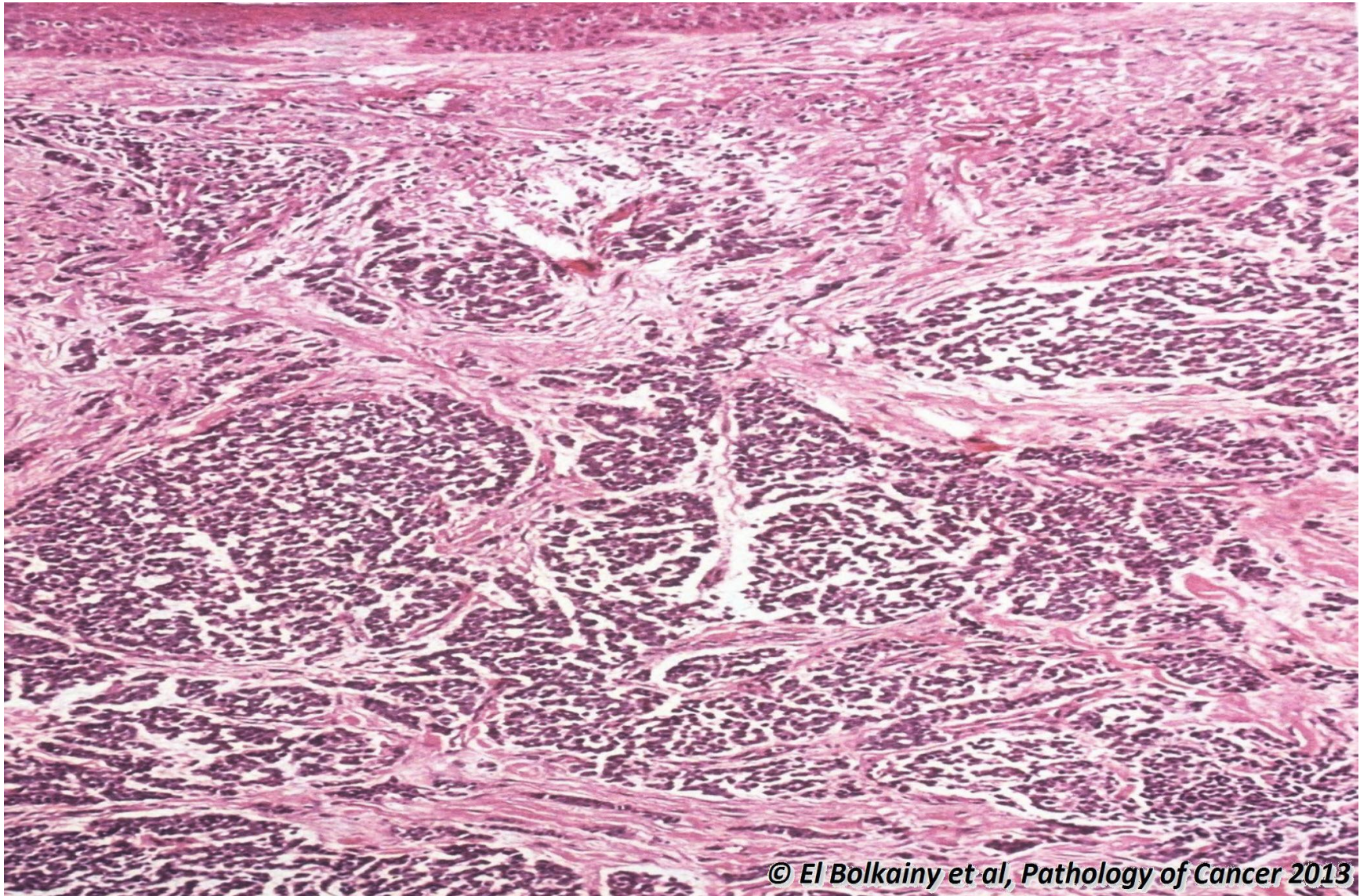
22.44 Proliferating pilar tumor, histology.



**Picture 22-44**

**Proliferating pilar tumor, histology.** **A** Gross features. A circumscribed solid tumor involving dermis and subcutaneous tissue covered by intact skin. **B** Well differentiated squamous cells with abrupt keratinization. Such tumor is considered of borderline malignancy especially if there is associated cytologic atypia or invasive margin. (Reproduced with permission, LeBoit et al, 2004, L.Requena, Figure 3-38).

22.45 Merkel cell carcinoma, histology.



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**Picture 22-45** **Merkle cell carcinoma, histology.** An ulcerating nodule associated with infiltrative pattern. Crowded cells with scanty cytoplasm, nuclei with finely dispersed chromatin and active mitosis. CK 20 immunoreactivity with characteristic dot-like paranuclear pattern. Merkle cell virus is evident in the tumor by PCR in 80 % of cases.

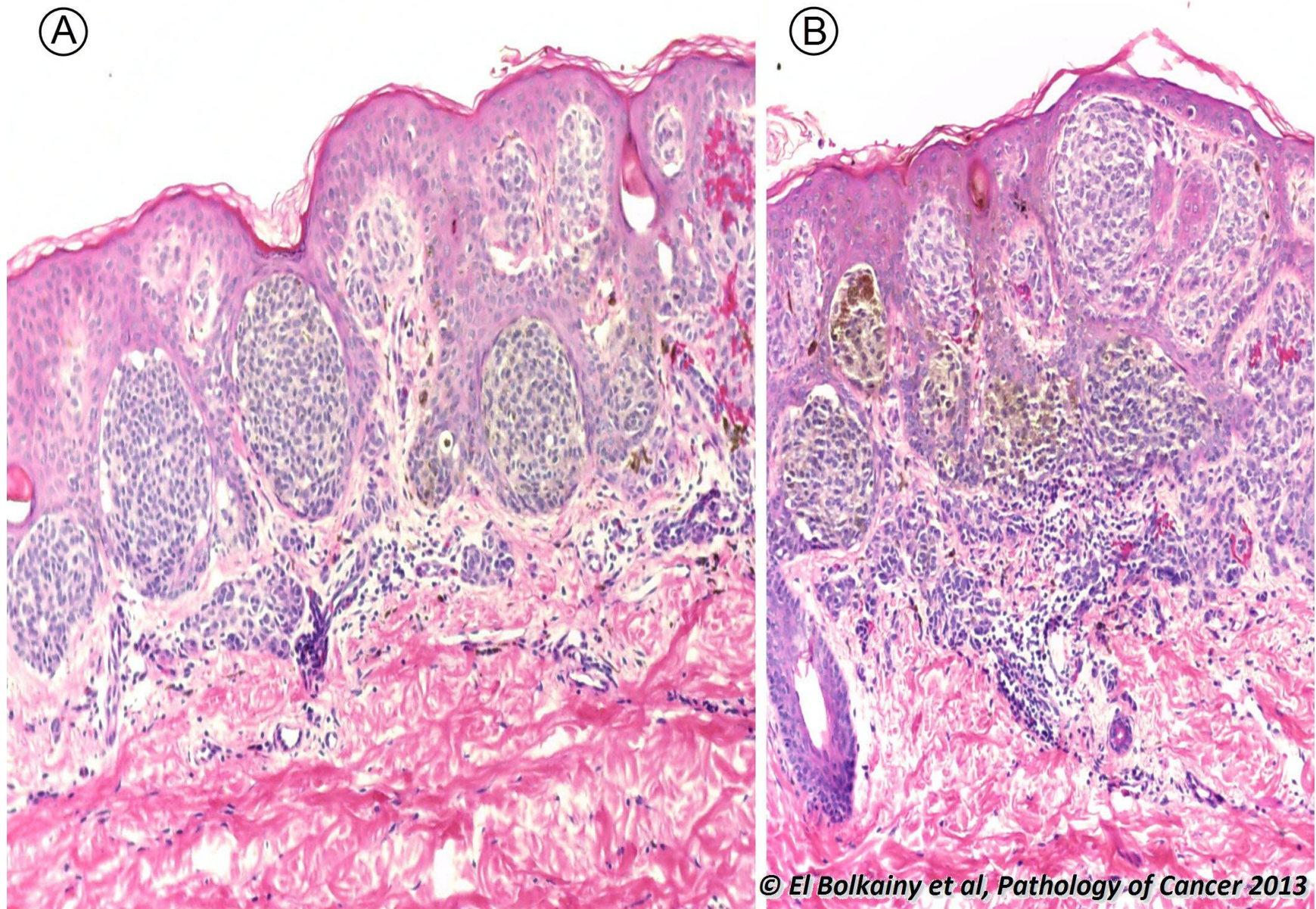
22.46 Dysplastic nevus syndrome, clinical picture.



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**Picture 22-46** Dysplastic nevus syndrome, clinical picture. Multiple large (> 5 mm) nevi of variable color and irregular periphery. This syndrome indicates increased risk to develop melanoma.

22.47 Dysplastic nevus syndrome, histology.



**Picture 22-47** Dysplastic nevus syndrome, histology. **A** and **B** There is fusion of junctional nests composed of atypical melanocytes with large hyperchromatic nuclei but no epidermal invasion.



22.48 Congenital giant hairy nevus, clinical picture.



**Picture 22-48**

**Congenital giant hairy nevus, clinical picture. A** A benign congenital nevus with no malignant transformation.

**B** A child with congenital nevus complicated by malignant melanoma (large nodule); a quite rare malignancy in children except in this setting.

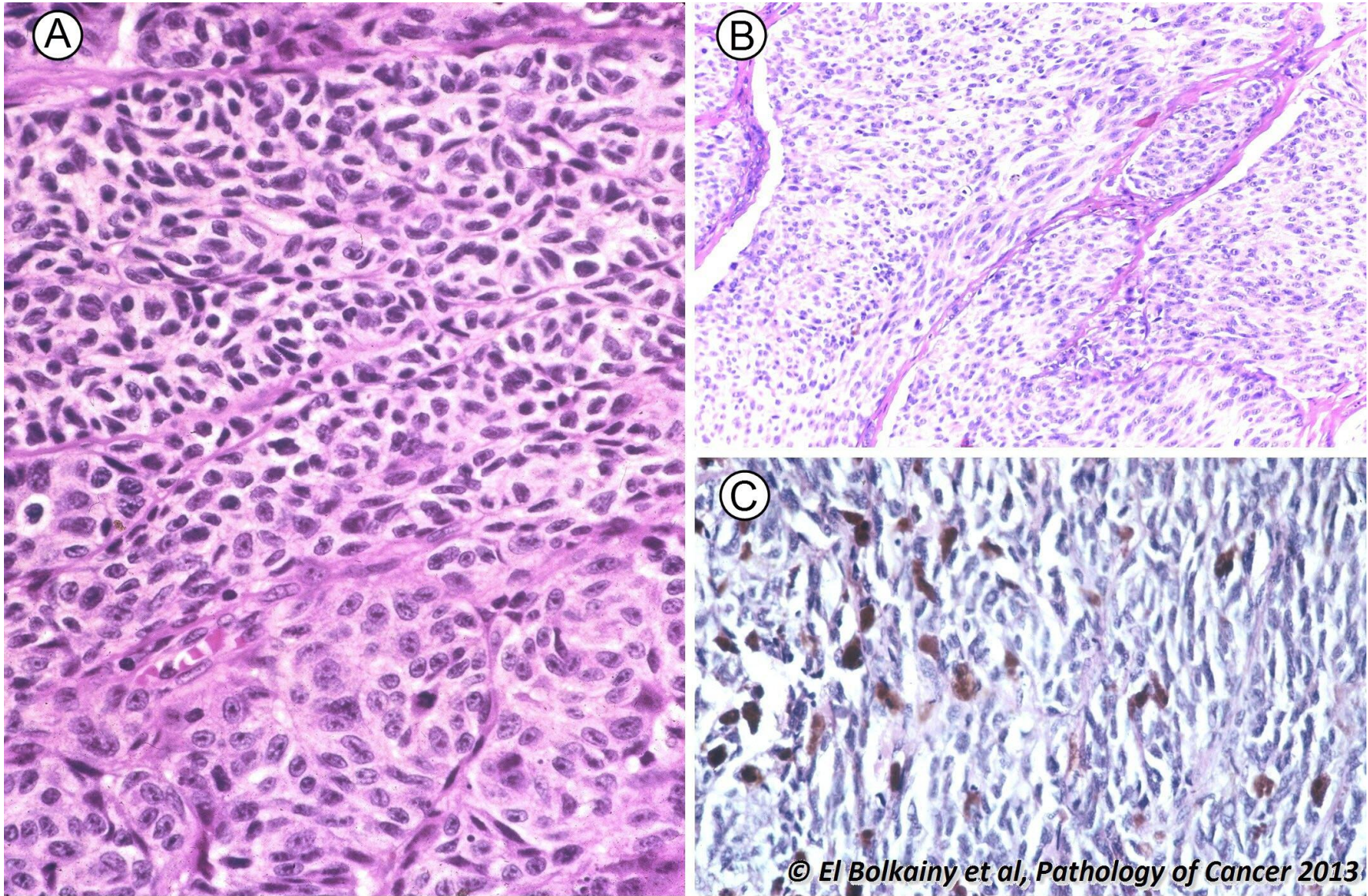
22.49 Malignant melanoma, gross features.



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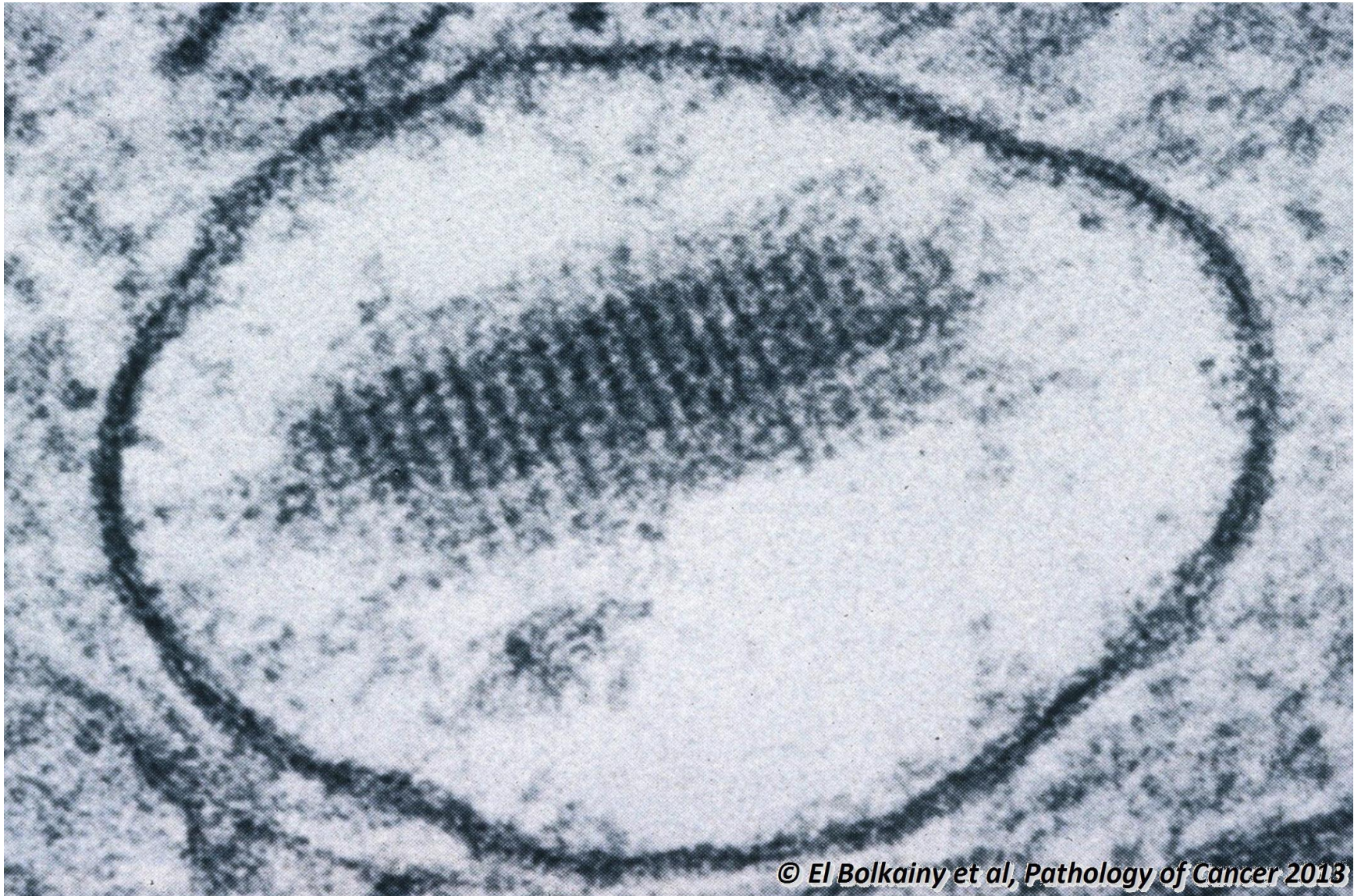
**Picture 22-49** Malignant melanoma, gross features. Note asymmetry in shape, depigmented areas, irregular borders and variation in surface texture (flat and nodular areas).

22.50 Malignant melanoma, histology.



**Picture 22-50** Malignant melanoma, histology. Note variation of cell shape (small, epithelioid and spindle cells), prominent nucleoli, active mitosis and melanin pigment located in cytoplasm of tumor cells.  
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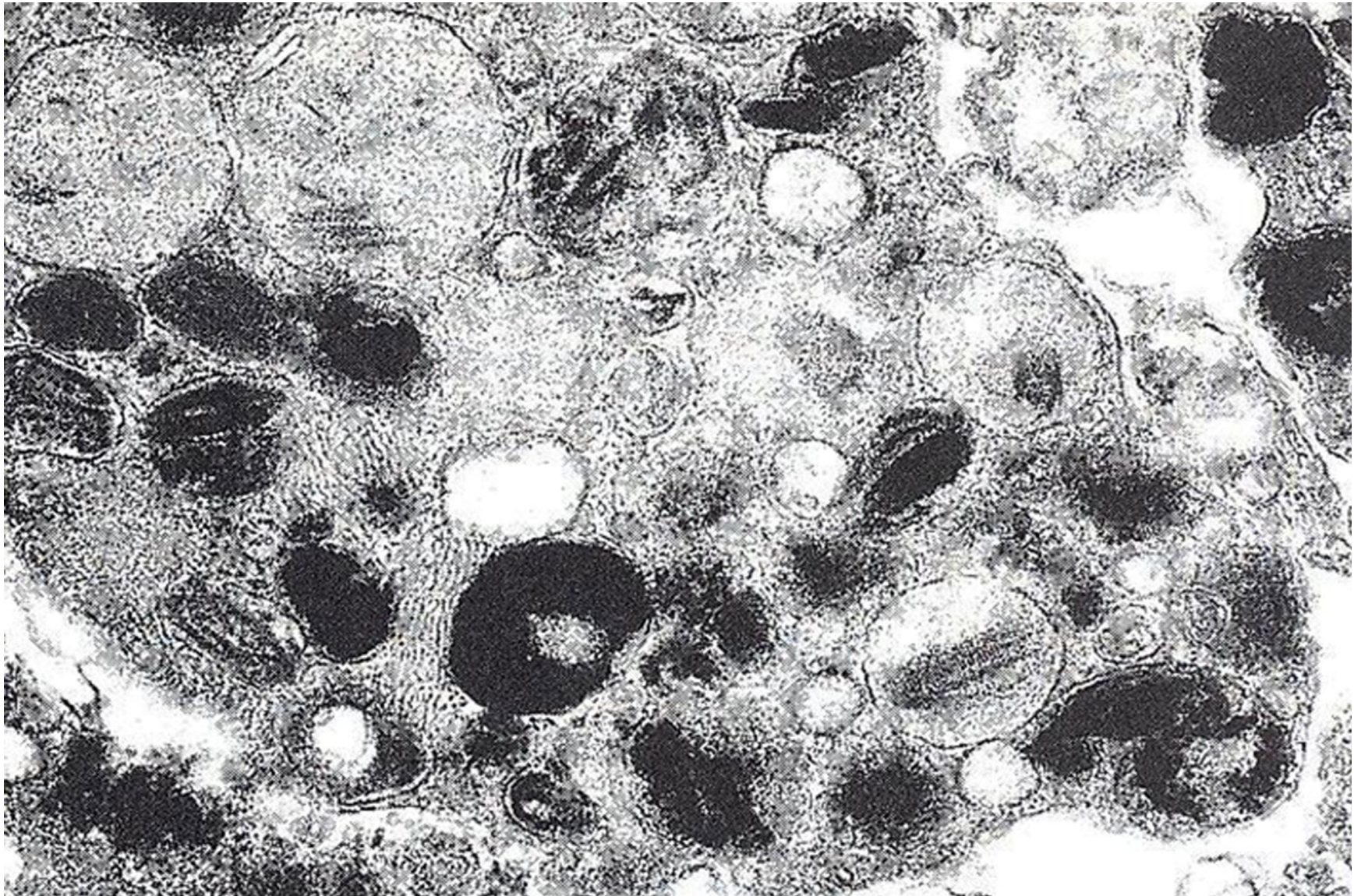
22.51 Amelanotic melanoma, electron microscopy.



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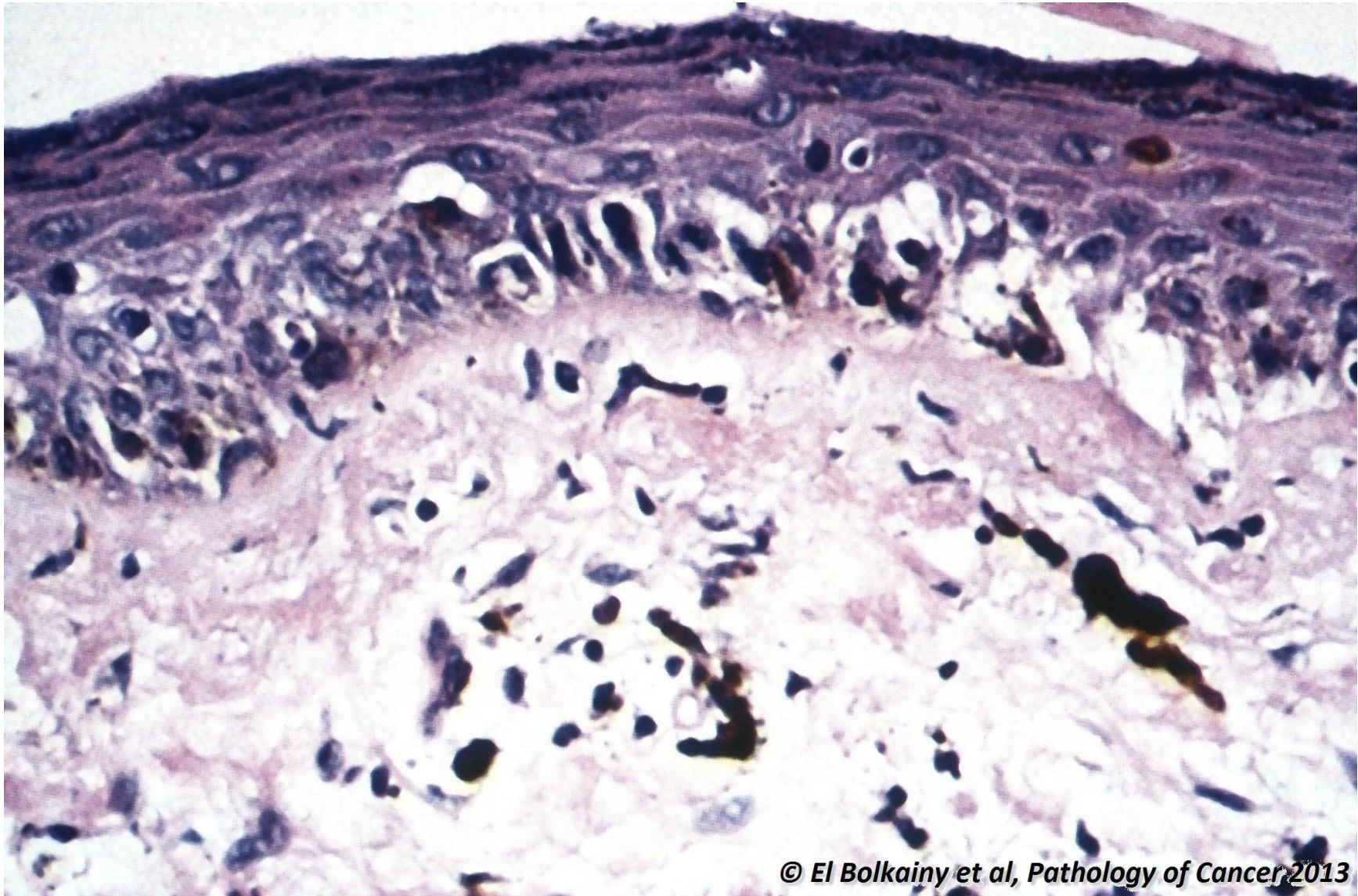
**Picture 22-51** Amelanotic melanoma, electron microscopy. It is confirmed by presence of premelanosomes (stage II) with striated structure and lack of pigment.

22.52 Malignant melanoma, electron microscopic features,



**Picture 22-52** Malignant melanoma, electron microscopic features, showing well formed melanosomes (stage IV). Note the lack of desmosomes at cell membrane explaining the dissociated pattern of melanoma cells. (Reproduced with permission, Fletcher CD, 2007).

22.53 Lentigo maligna, histology.

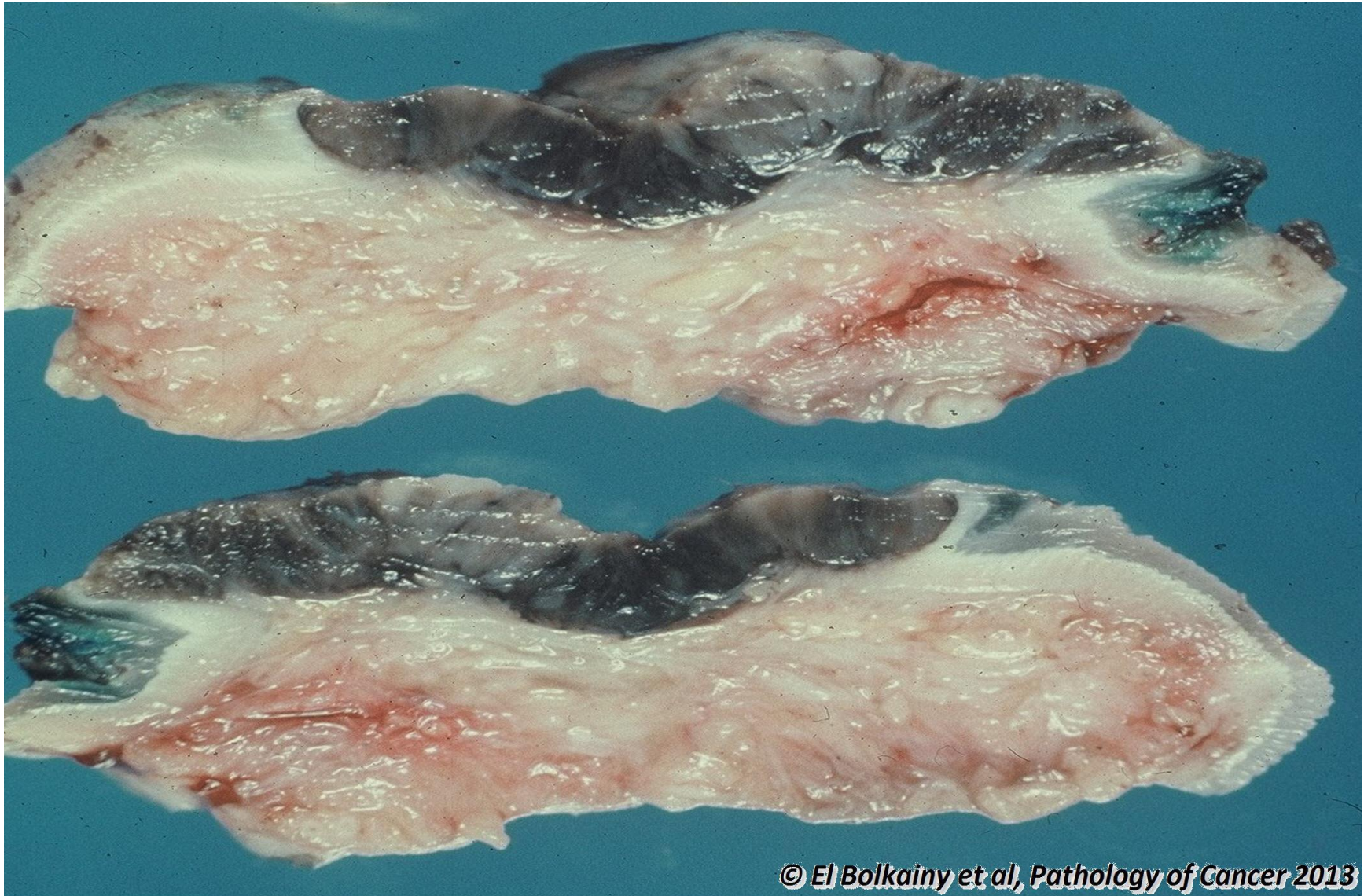


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**Picture  
22-53**

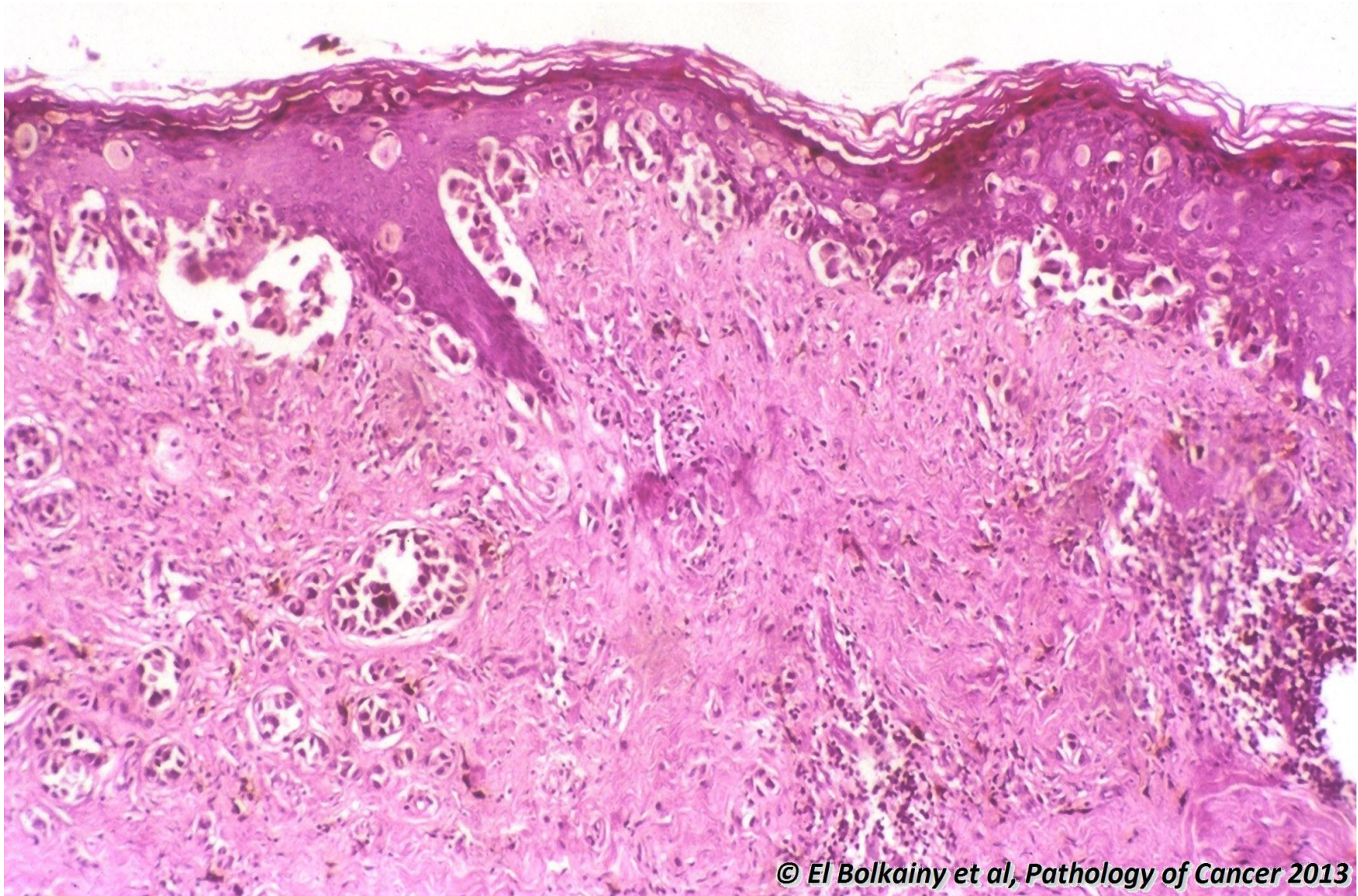
**Lentigo maligna, histology.** This represents melanoma in situ. Malignant melanocytes are limited to basal layer, but no invasion of stroma. Pigmented cells in the dermis are histiocytes with phagocytosed melanin pigment (melanophages).

22.54 Superficial spreading melanoma, gross features.



**Picture 22-54** Superficial spreading melanoma, gross features. Tissue section of a pigmented melanoma with a radial rather than vertical growth. Clearance of tissue around such large tumor should be at least 3 cm.

22.55 Malignant melanoma, superficial spreading type, histology.



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**Picture 22-55** Malignant melanoma, superficial spreading type, histology. There is invasion of upper dermis and the associated intraepidermal components are both junctional nests, as well as, intraepidermal pagetoid pattern.



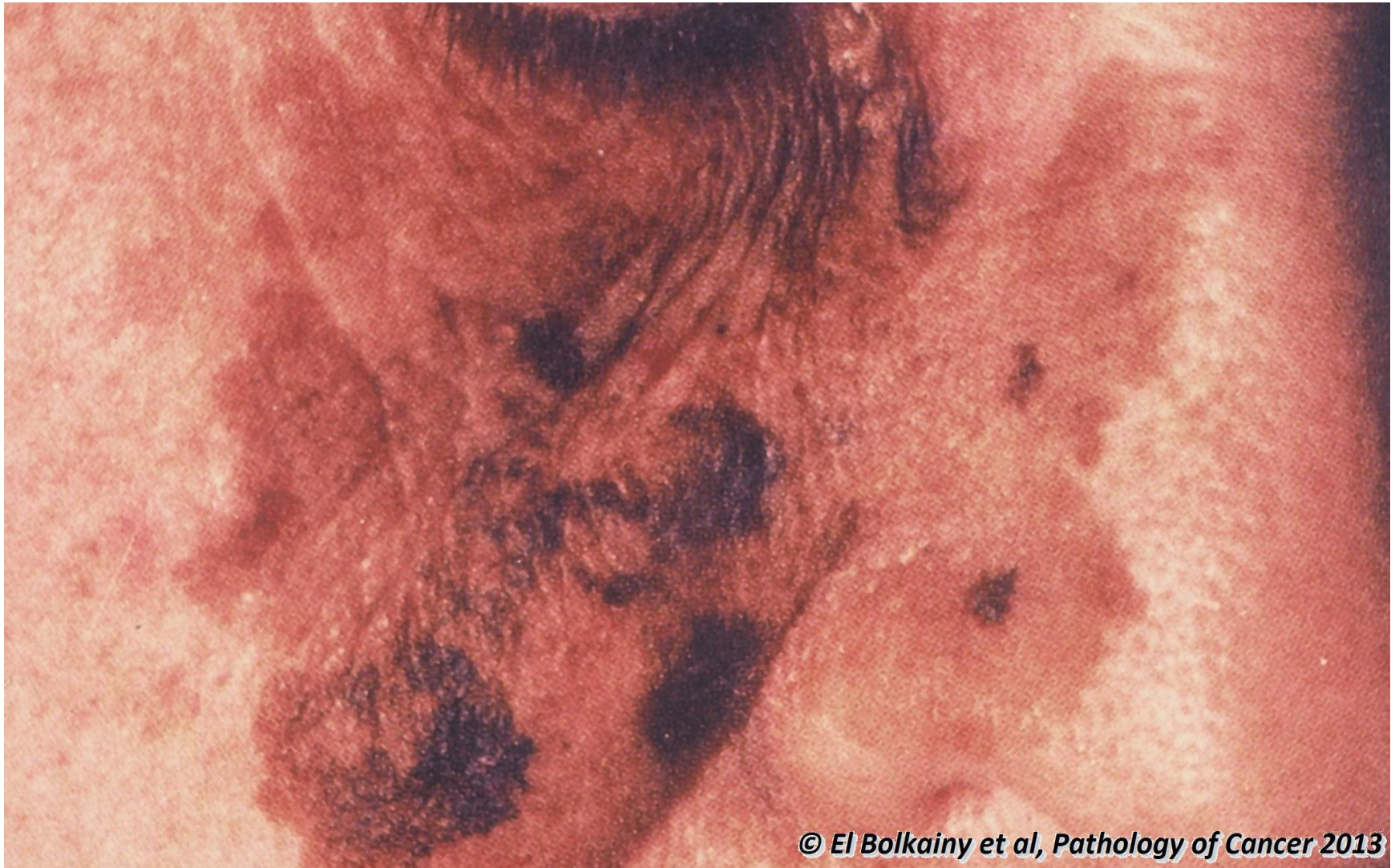
22.56 Nodular melanoma, histology.



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**Picture 22-56** Nodular melanoma, histology. Advanced tumor reaching subcutaneous fat, no evidence of maturation in the deep part of the tumor compared to its superficial part. No associated intraepidermal melanoma component.

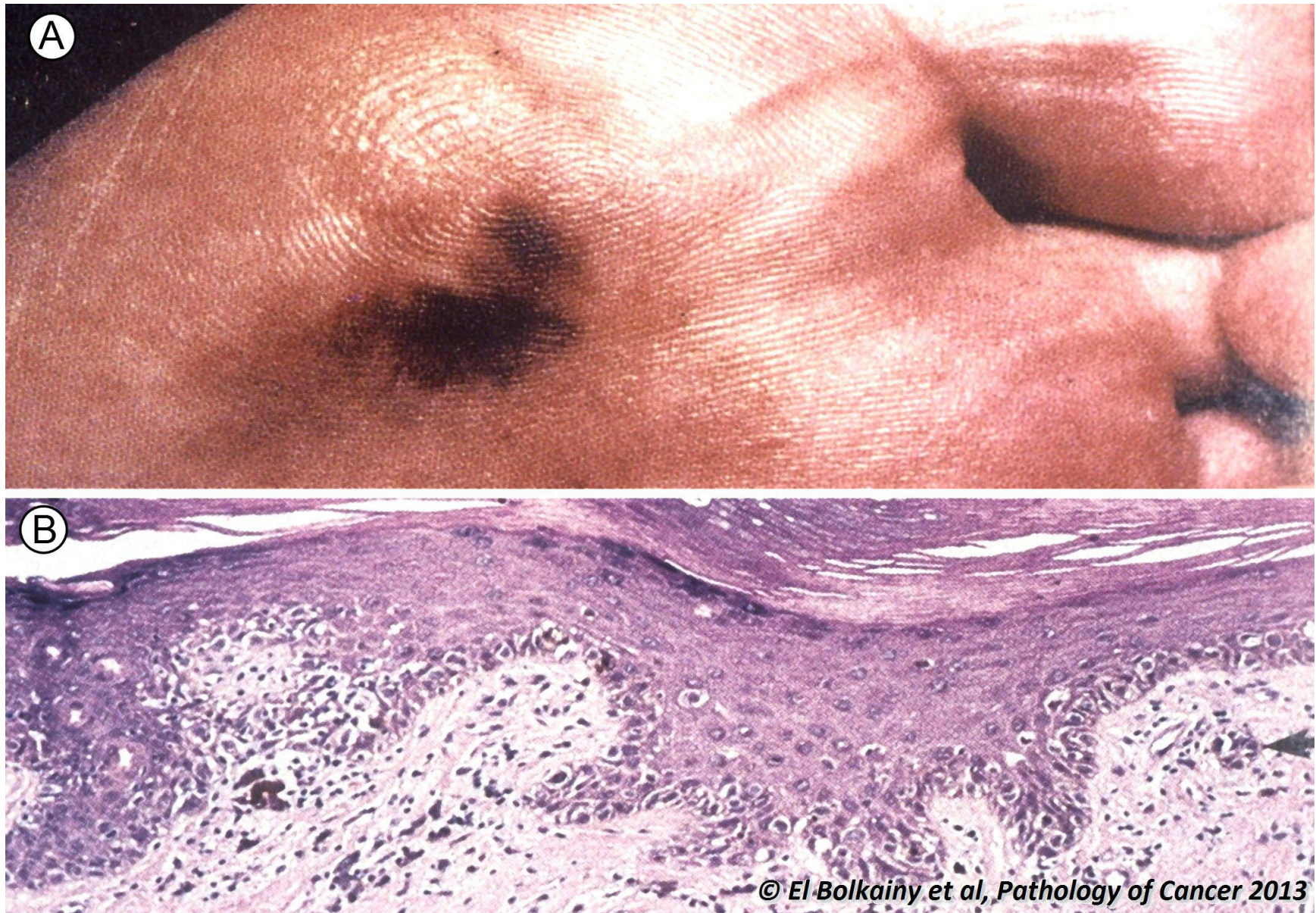
22.57 Lentigo maligna melanoma, clinical picture.



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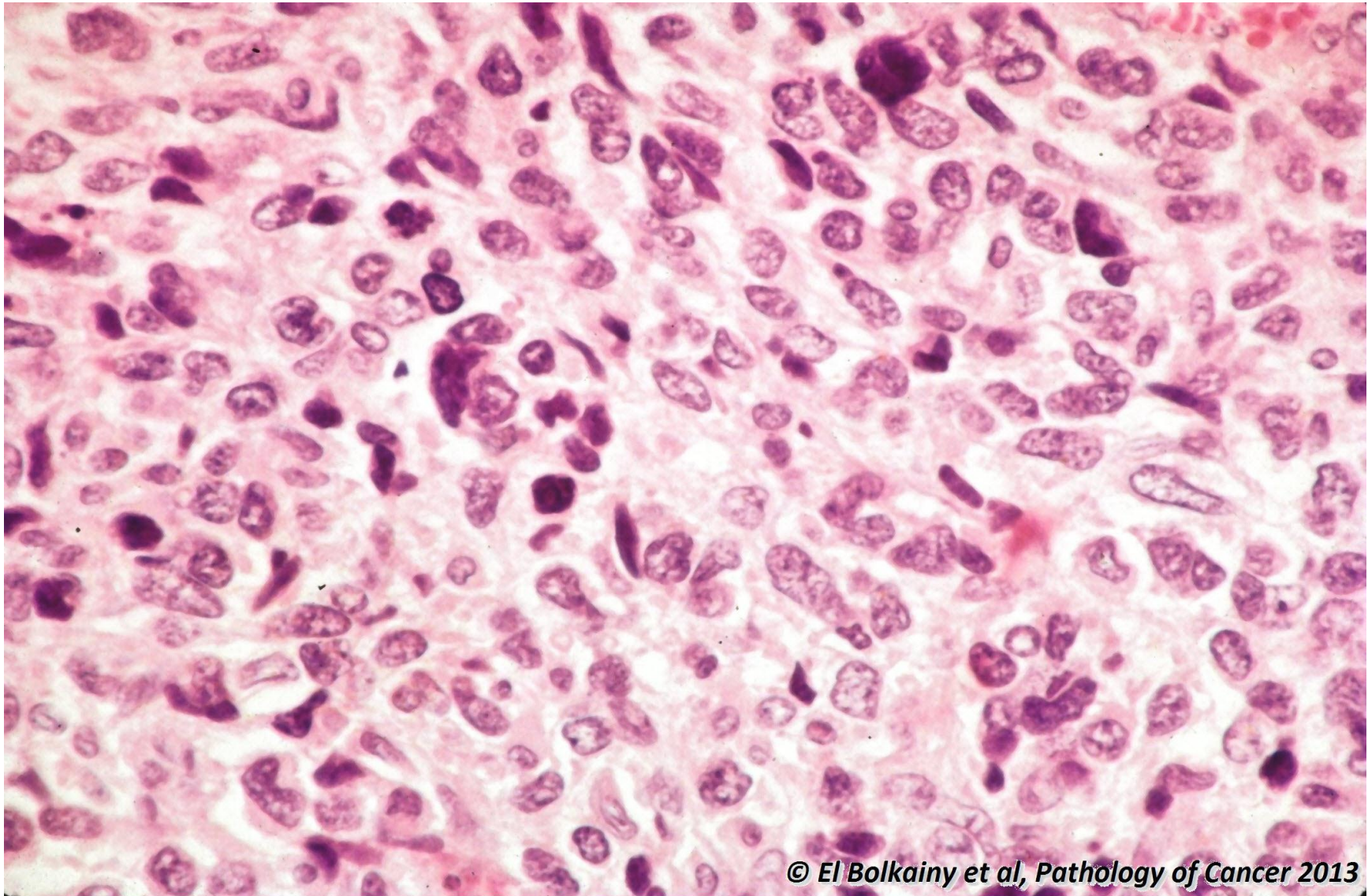
**Picture 22-57** Lentigo maligna melanoma, clinical picture. The nodules in the lesion are malignant melanoma complicating lentigo maligna ( the flat brownish lesion).

22.58 Acral melanoma.



**Picture 22-58** Acral melanoma. **A** Gross features, a pigmented macule at sole of the foot. **B** Malignant melanocytes invading the stroma, associated with intraepidermal lentiginous melanoma component.

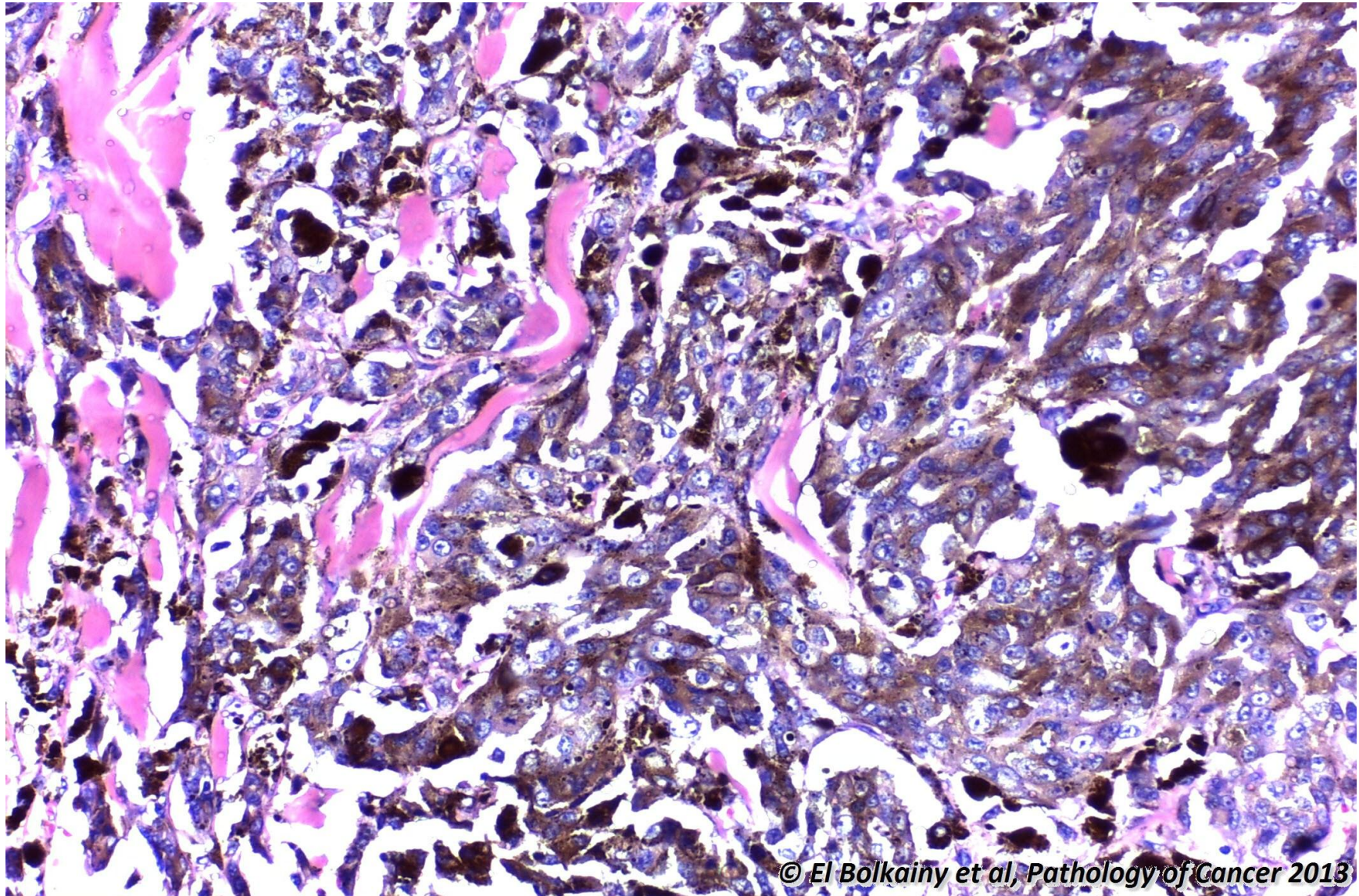
22.59 Amelanotic melanoma, histology.



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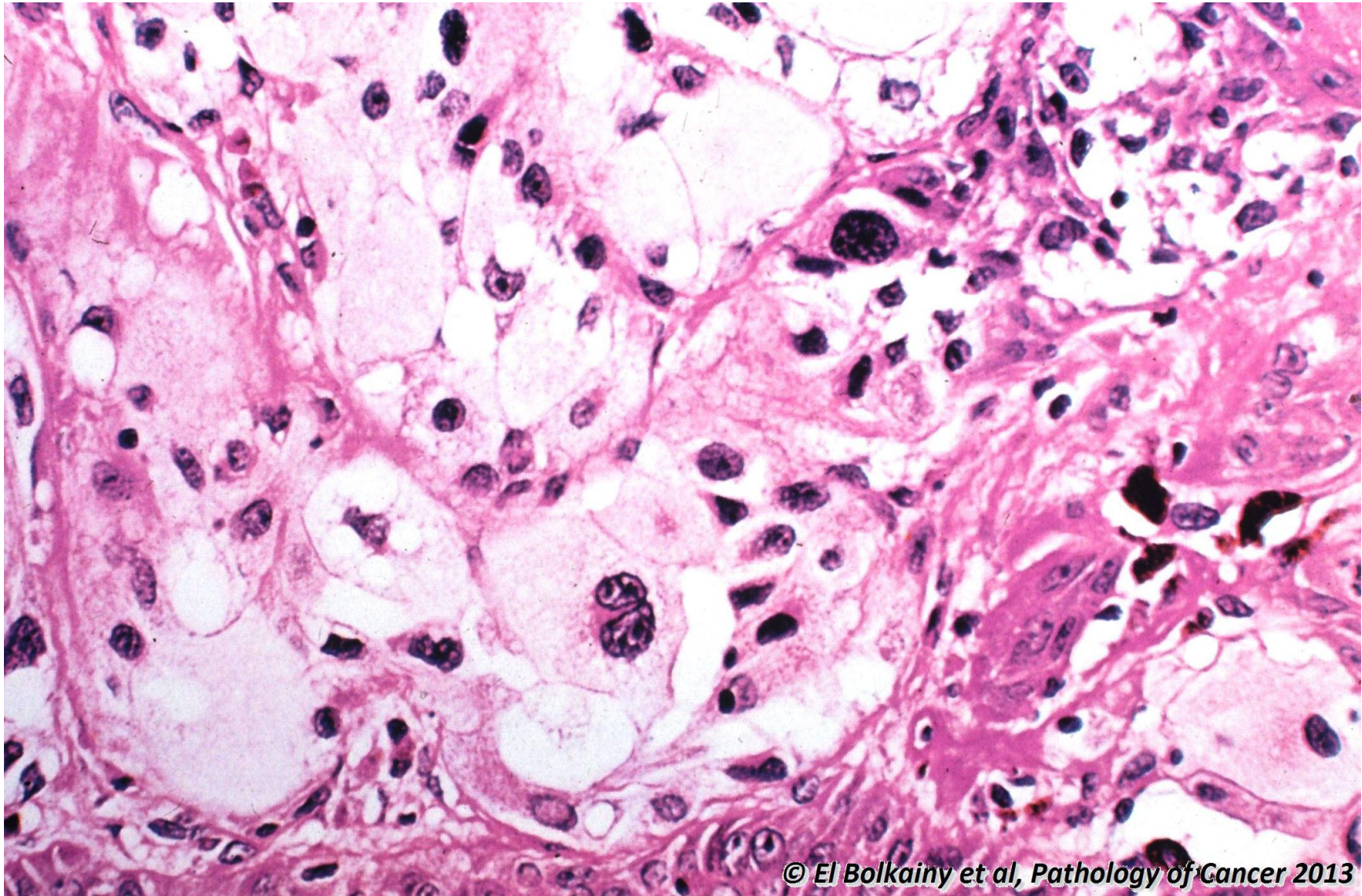
**Picture 22-59** Amelanotic melanoma, histology. In absence of melanin, the presence of discohesive epithelioid, spindle and giant cells should raise the suspicion of melanoma. Immunophenotyping is confirmatory (positive S-100, HMB-45 and Melan-A).

22.60 Malignant blue nevus, histology.



**Picture 22-60** Malignant blue nevus, histology. Cellular spindle cell melanoma in deep dermis. The nuclei are pleomorphic with prominent nucleoli and mitotic activity . No evidence of cell maturation in deep parts of tumor.

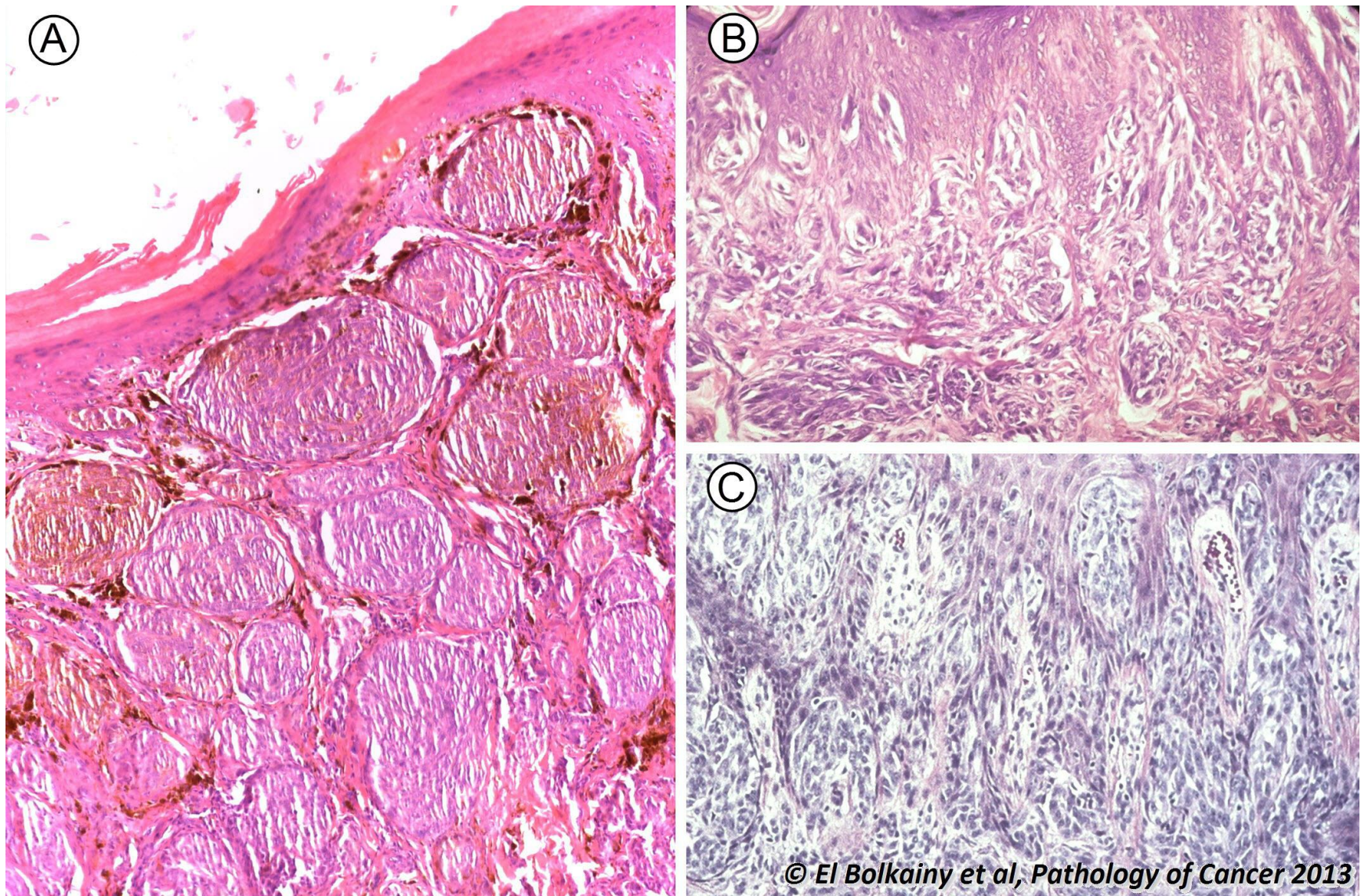
22.61 Balloon cell melanoma, histology.



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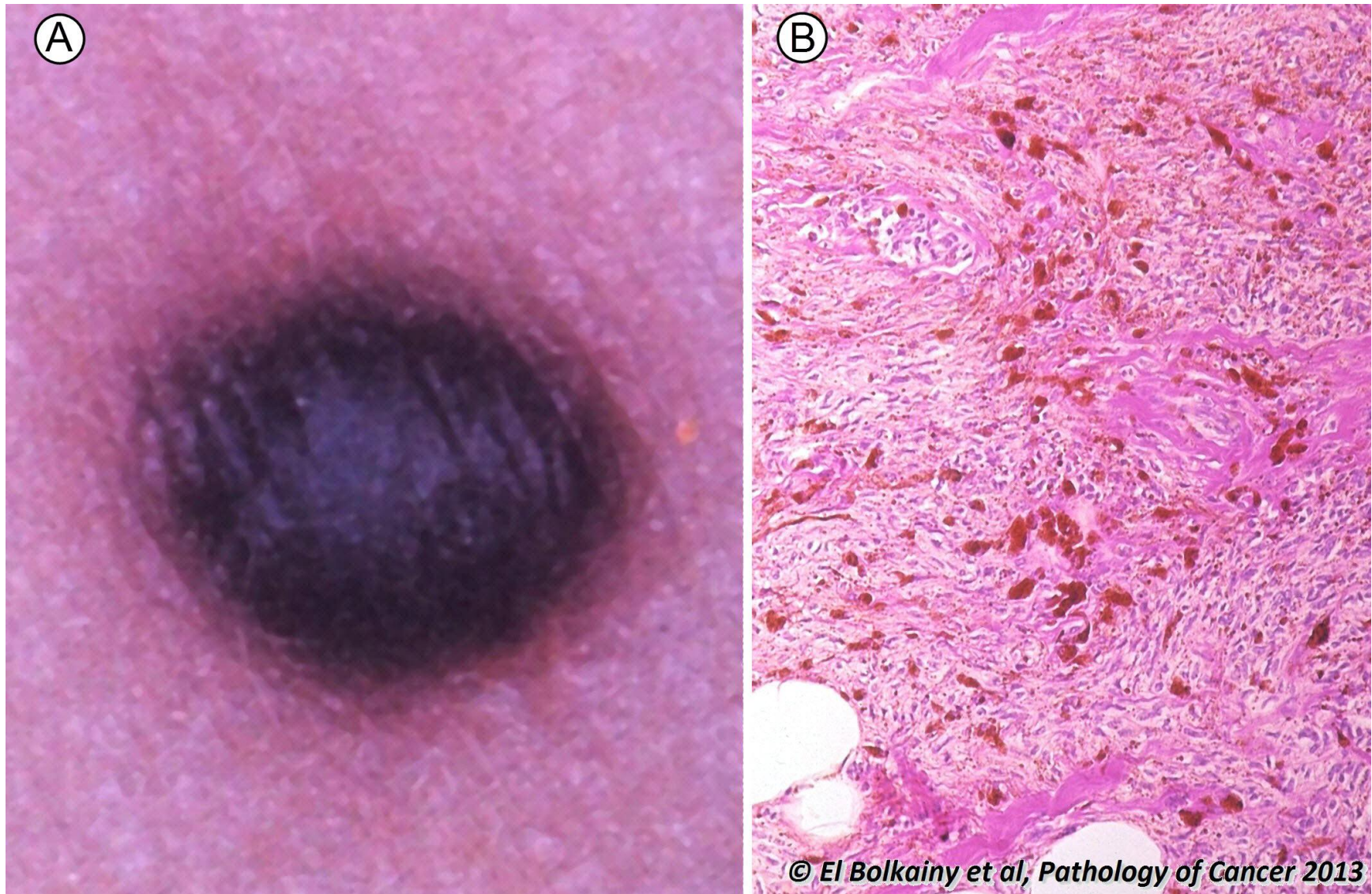
**Picture 22-61** Balloon cell melanoma, histology. Large cells with abundant clear cytoplasm. Immunophenotyping will distinguish this rare melanoma variant from sarcoma or clear carcinoma.

22.62 Juvenile spindle cell nevus of Spitz, histology.



**Picture 22-62** Juvenile spindle cell nevus of Spitz, histology. There is marked junctional activity composed of spindle cells and giant cells. It commonly affects children, an age group rarely affected by melanoma.

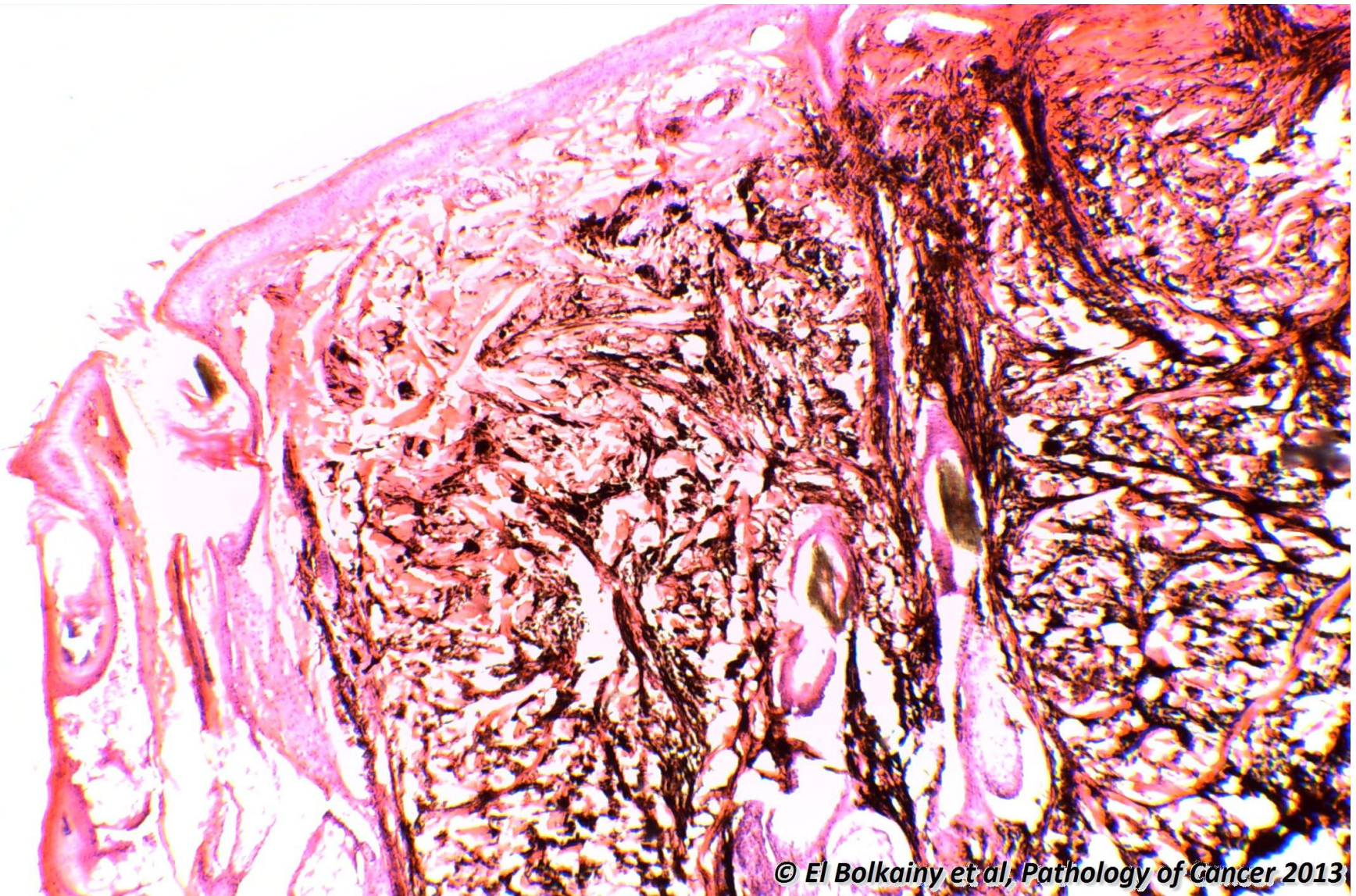
22.63 Blue nevus.



**Picture 22-63** Blue nevus. **A** Grossly, It appears as a small bluish papule. **B** Histology, It is composed of heavily pigmented spindle cells, located deep in dermis, with associated marked fibrosis.



22.64 Blue nevus, histology.

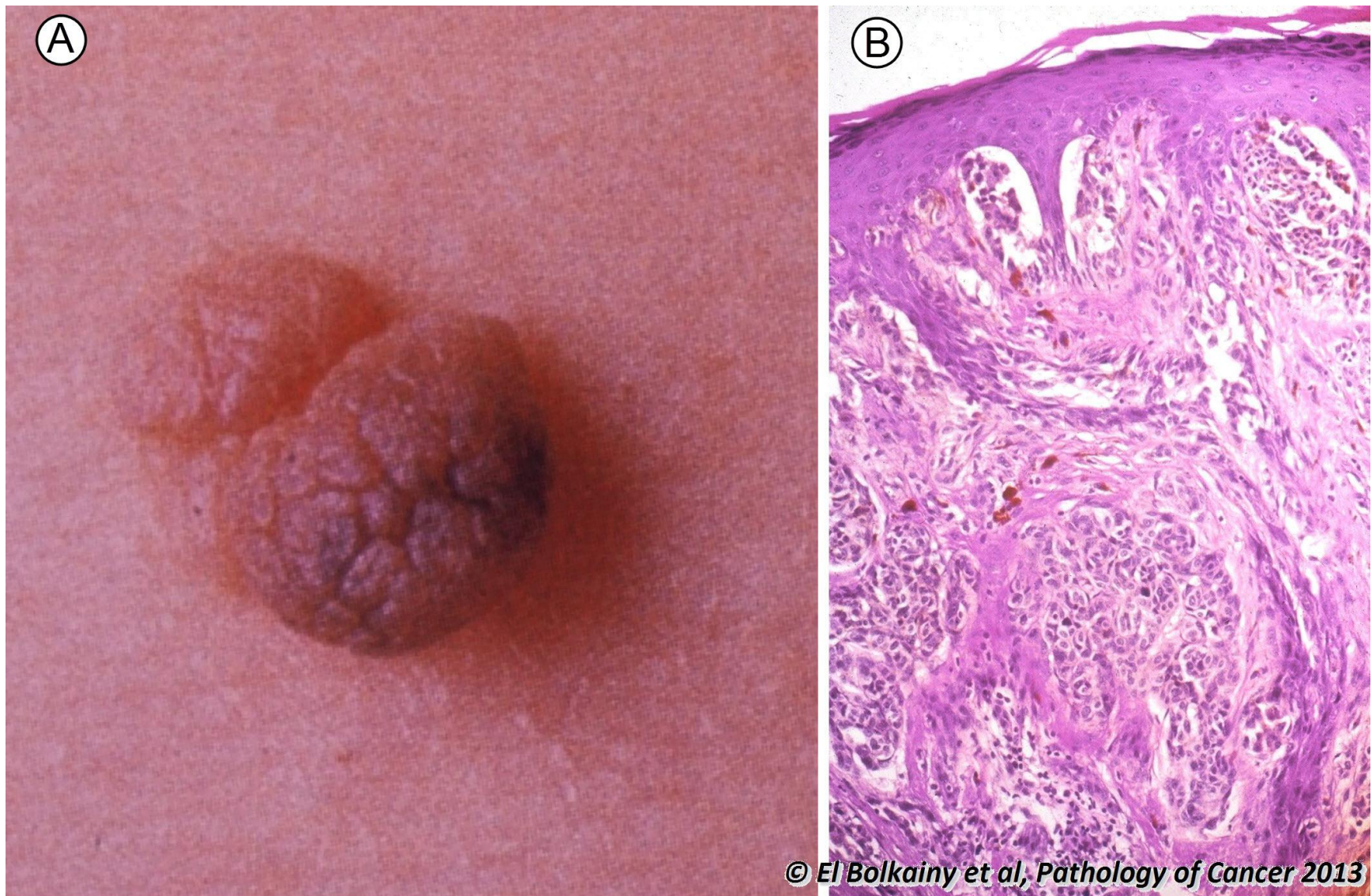


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**Picture  
22-64**

Blue nevus, histology. Heavily pigmented spindle and dendritic cells with abundant stromal fibrosis.

22.65 Compound nevus.



**Picture  
22-65**

**Compound nevus.** **A** Gross, symmetrical pigmented nodule of homogeneous color. **B** Histology, the presence of nests of melanocytes in junctional area, as well as, in dermis. The stroma is rich and there is evidence of maturation in deep cells, changing from large superficial epithelioid cells to deep spindle cells with neurilemmal features.

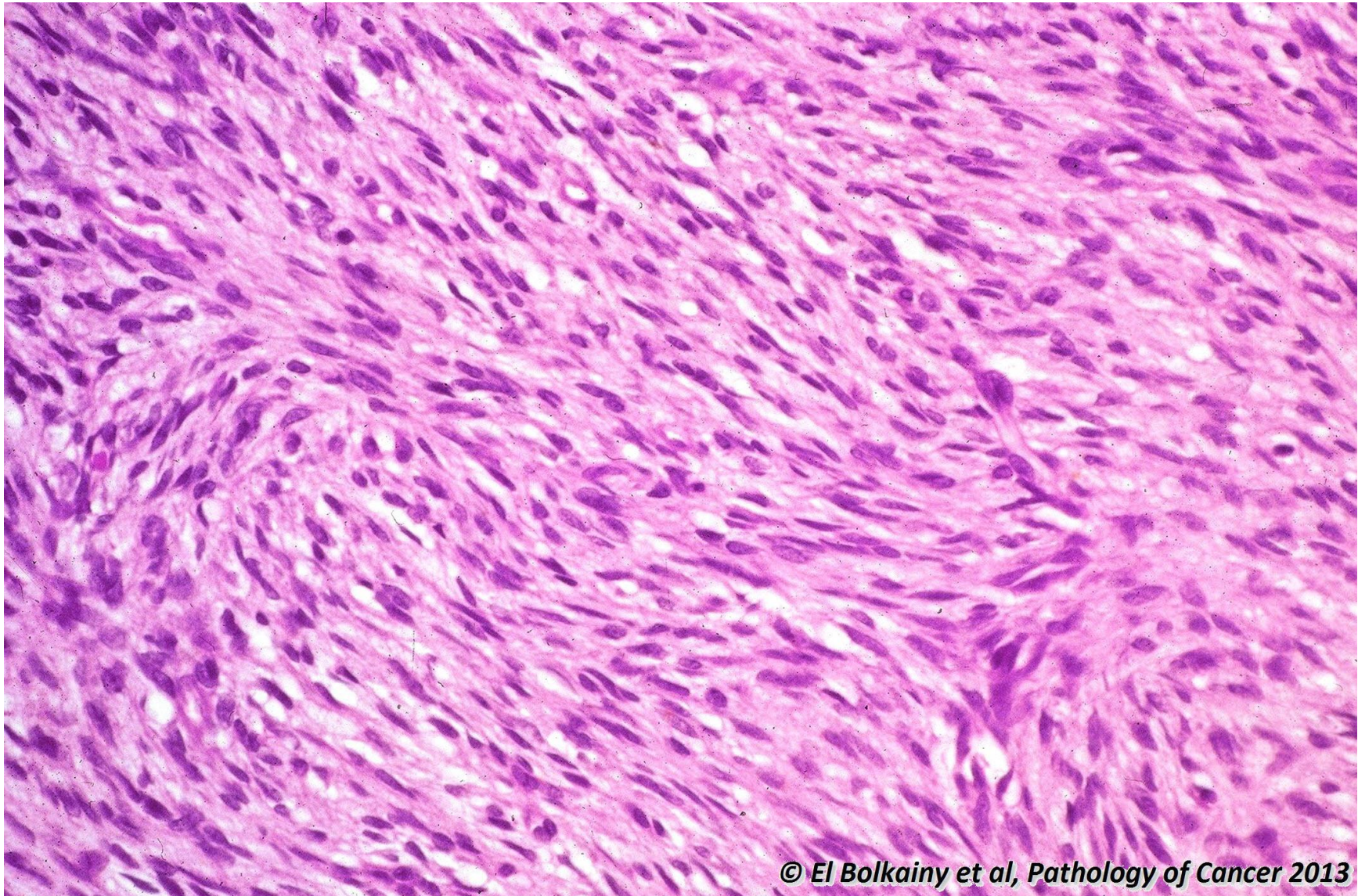
22.66 Dermatofibrosarcoma protuberans, gross features.



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**Picture 22-66** Dermatofibrosarcoma protuberans, gross features. The tumor is fibrotic trabeculated, superficial and adherent to skin which is thin and stretched, difficult to separate.

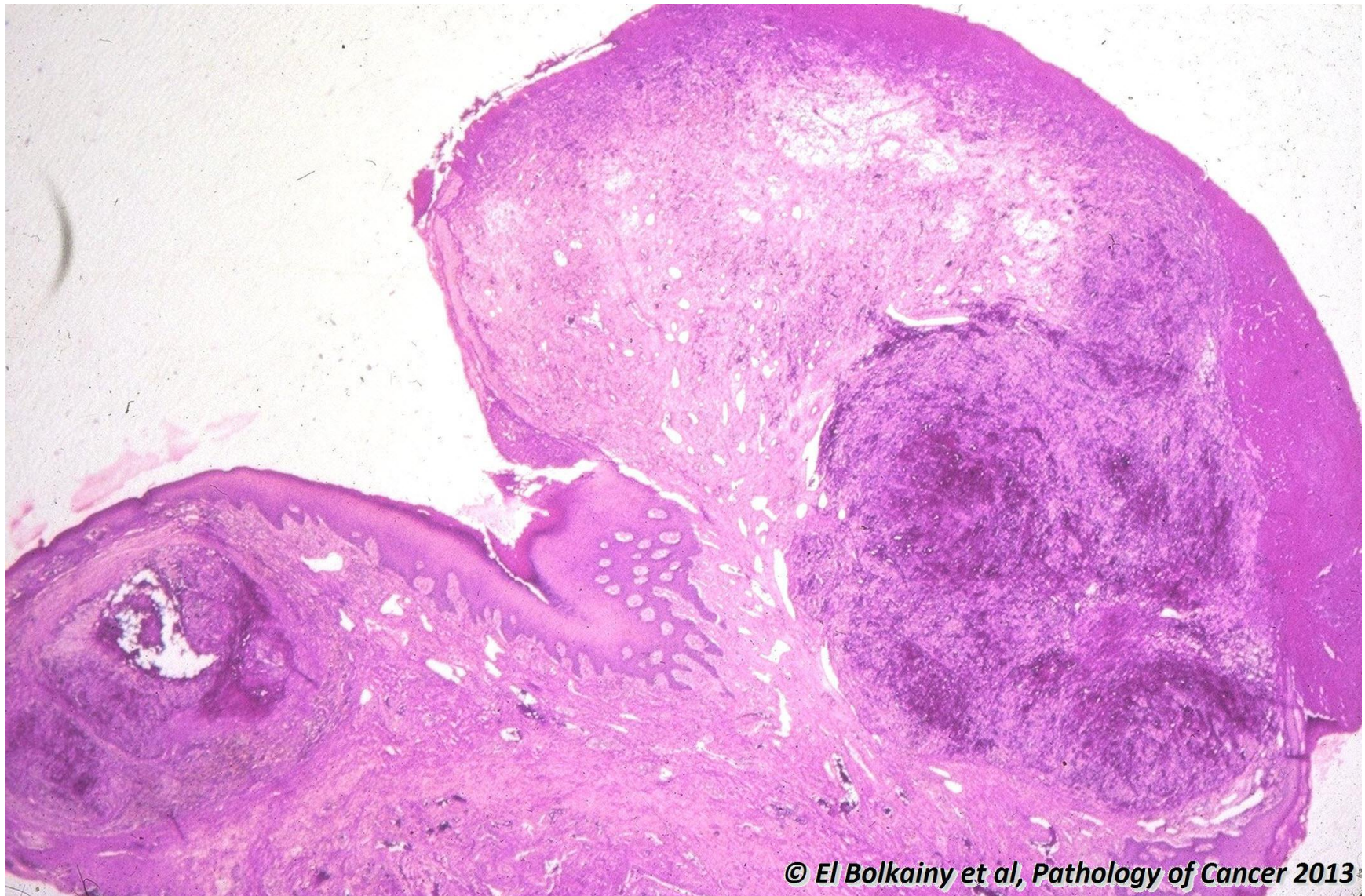
22.67 Dermatofibrosarcoma protuberans, histology.



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**Picture 22-67** Dermatofibrosarcoma protuberans, histology. Cellular fibroblasts with storiform pattern, with infiltrating border reaching subcutaneous fat. The fibroblasts are positive to CD34.

22.68 Kaposi sarcoma, histology, low power.

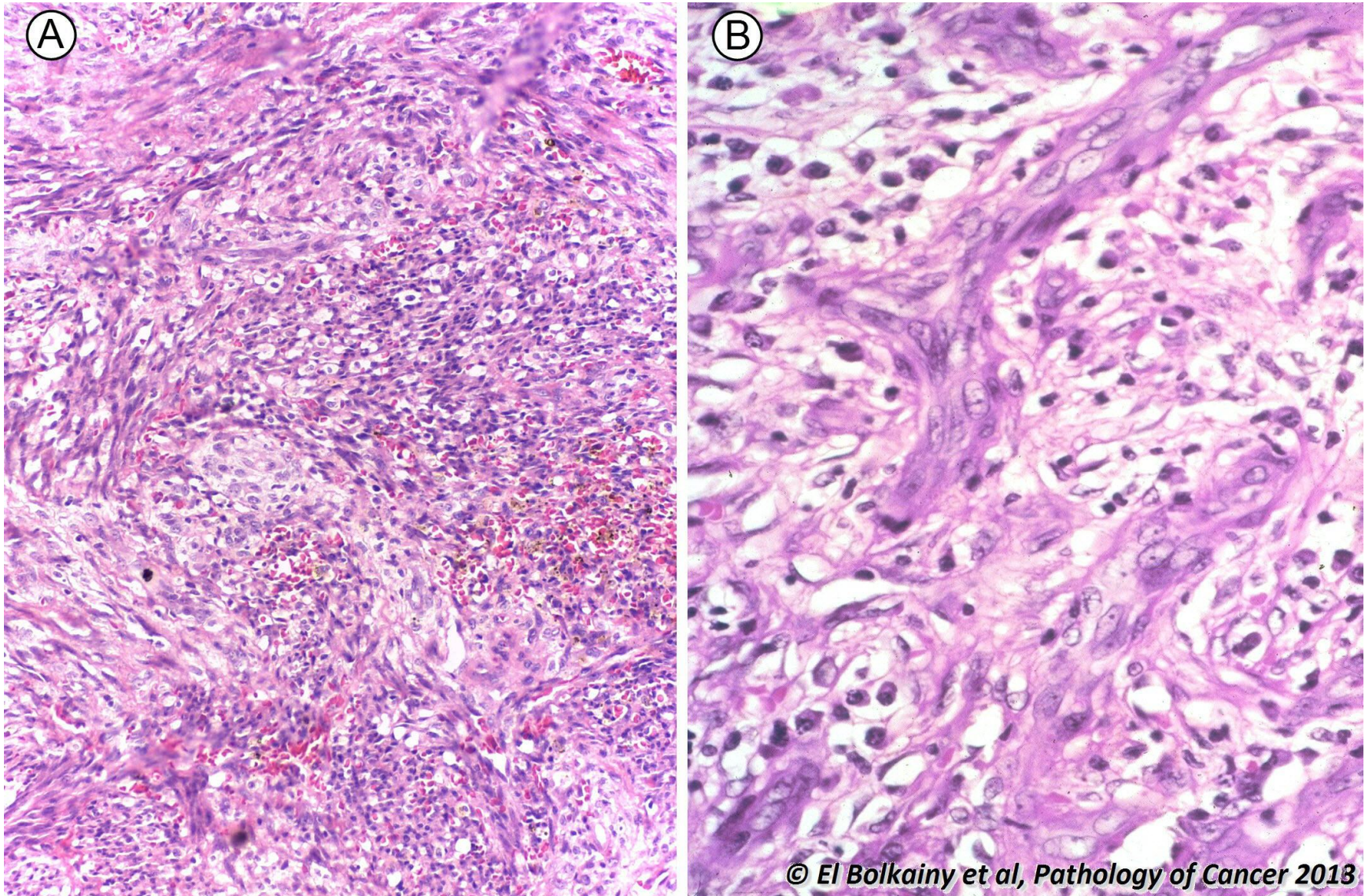


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**Picture  
22-68**

**Kaposi sarcoma, histology, low power.** Note the multiple rounded lesions in the dermis of variable but mostly small size.

22.69 Kaposi sarcoma, histology, high power.



**Picture 22-69** Kaposi sarcoma, histology, high power. Note proliferating spindle cells around slit-like vascular spaces, extravasated red blood cells, hemosiderin deposits in stroma and macrophages. Spindle cells are immunoreactive to CD34.

22.70 Classic Kaposi sarcoma, clinical picture.



**Picture 22-70** Classic Kaposi sarcoma, clinical picture. The patients are elderly and the lesion (scaly plaques and nodules) affects mainly lower extremities.

22.71 Kaposi sarcoma in AIDS patient.



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**Picture 22-71** Kaposi sarcoma in AIDS patient. In contradistinction with classic Kaposi, the patient is younger, plaques and nodules affect skin of head and neck as well as oral mucosa rather than than lower extremity.



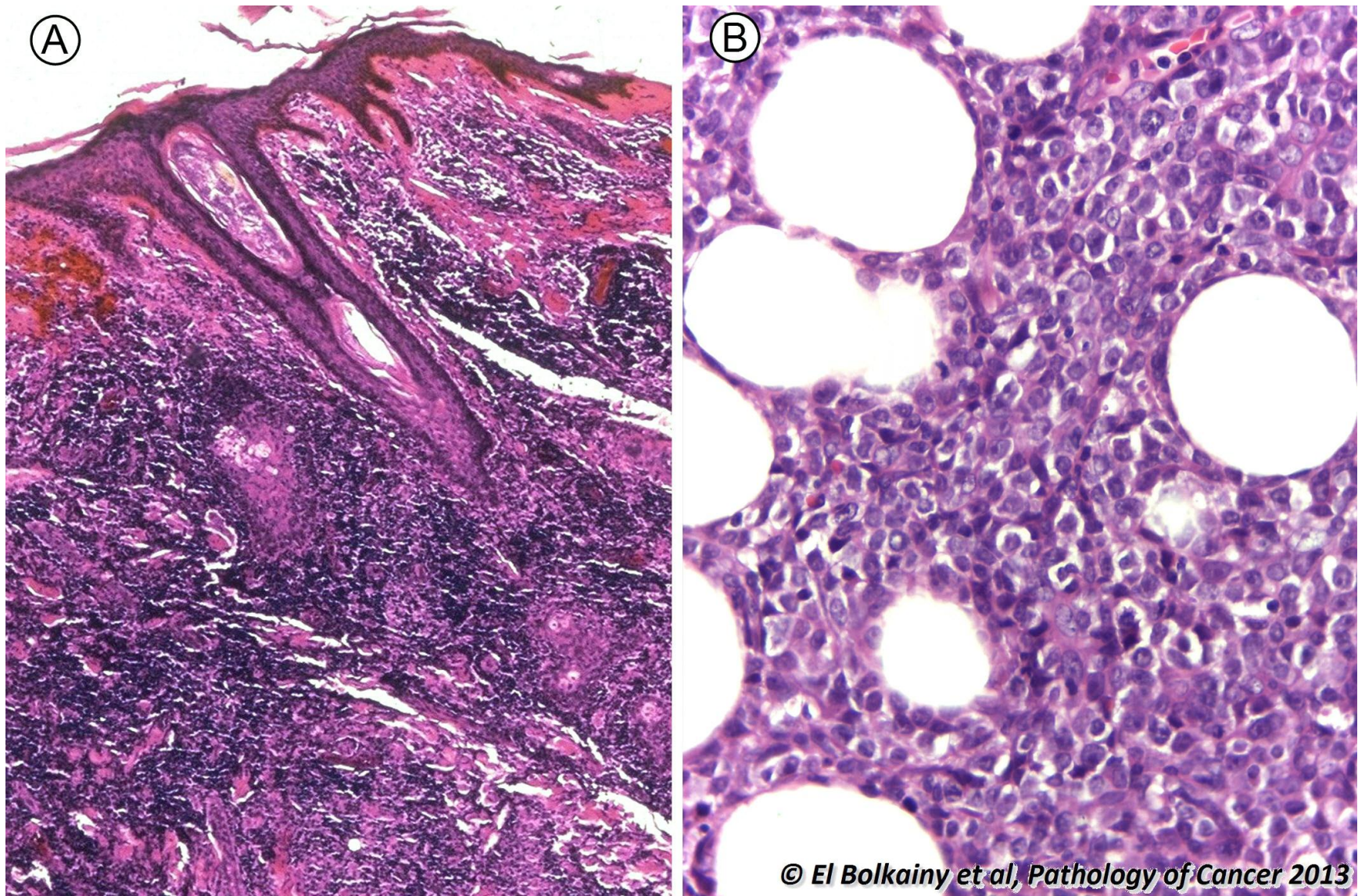
22.72 Mycosis fungoides.



**Picture 22-72** Mycosis fungoides. **A** Gross, multiple erythematous patches, plaques and nodules. **B** Histology, atypical T-lymphocytes (CD 3+) infiltrate in upper dermis with epidermal infiltration (epidemotropism). **C** High power.

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22.73 Primary cutaneous B-cell lymphoma, histology.

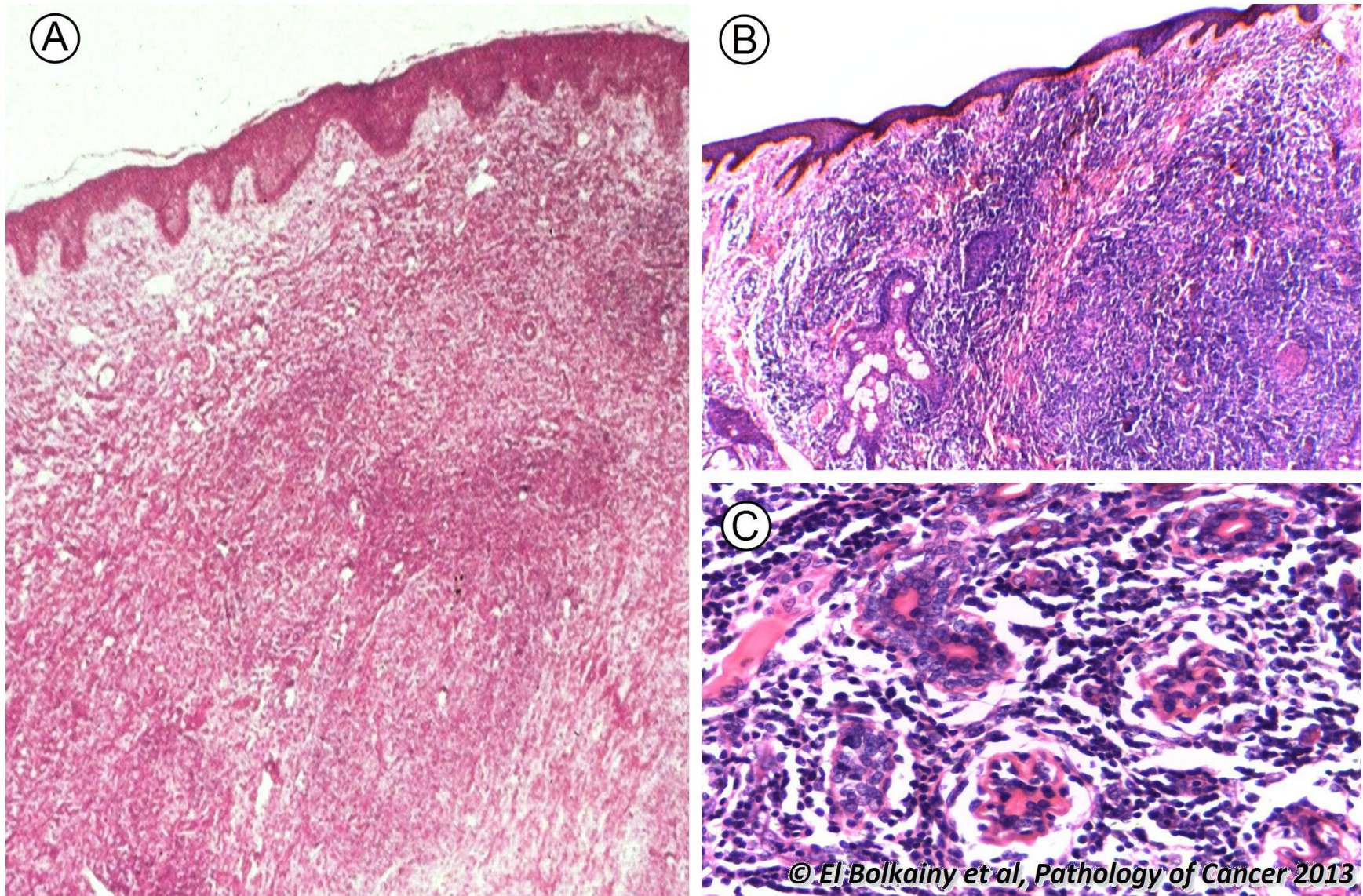


**Picture 22-73**

**Primary cutaneous B-cell lymphoma, histology.** **A** The infiltrate lacks epidemotropism and > 80% of cells are B cells (CD20+). The size of lymphocytes may be small (marginal zone type or MALT) or large (diffuse large B-cell lymphoma). **B** A monomorphic large atypical lymphoid cells with absence of fibrosis and new vessel formation.

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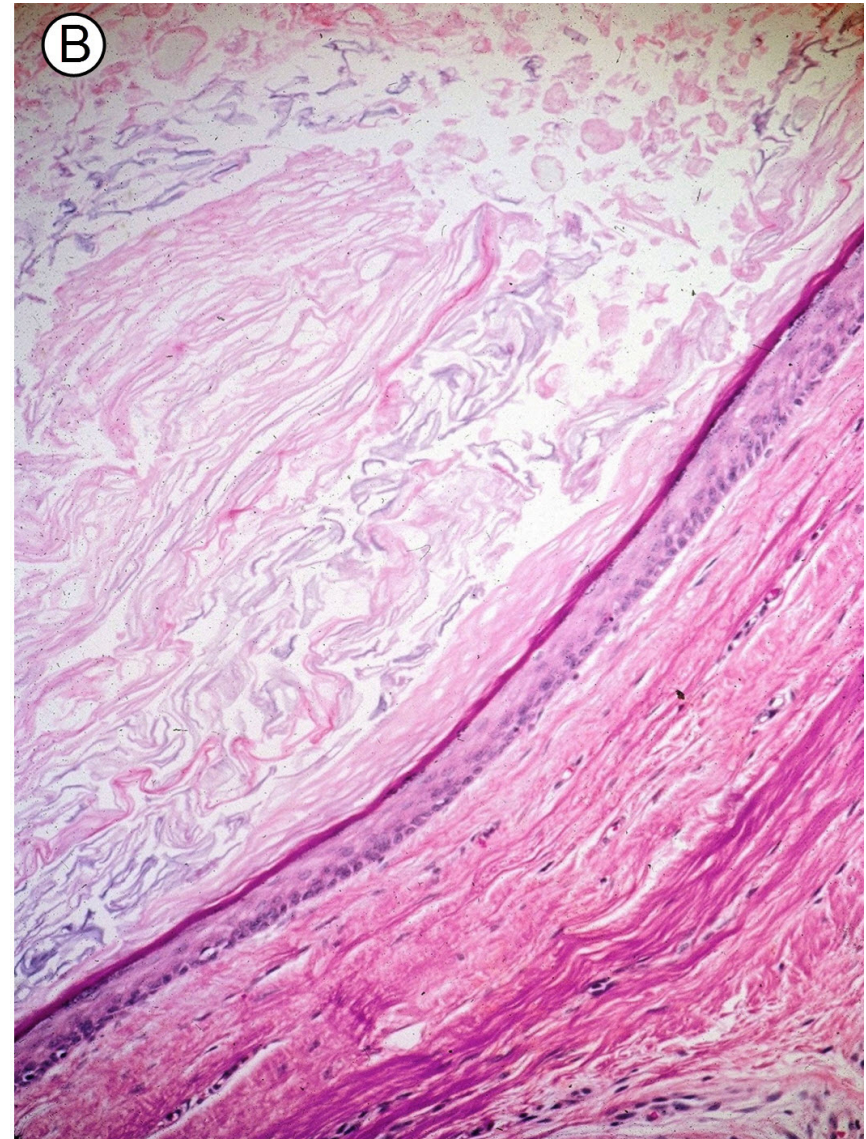
22.74 Reactive lymphoid hyperplasia, histology.



**Picture 22-74** **Reactive lymphoid hyperplasia, histology.** A Low power, note the germinal centers. B Low power and C High power. The infiltrate is polymorphous (lymphocytes, plasma cells and histiocytes), biphenotypic (a mixture of B and T phenotypes), vascular stroma and associated fibrosis. Lymphocytes are polyclonal.

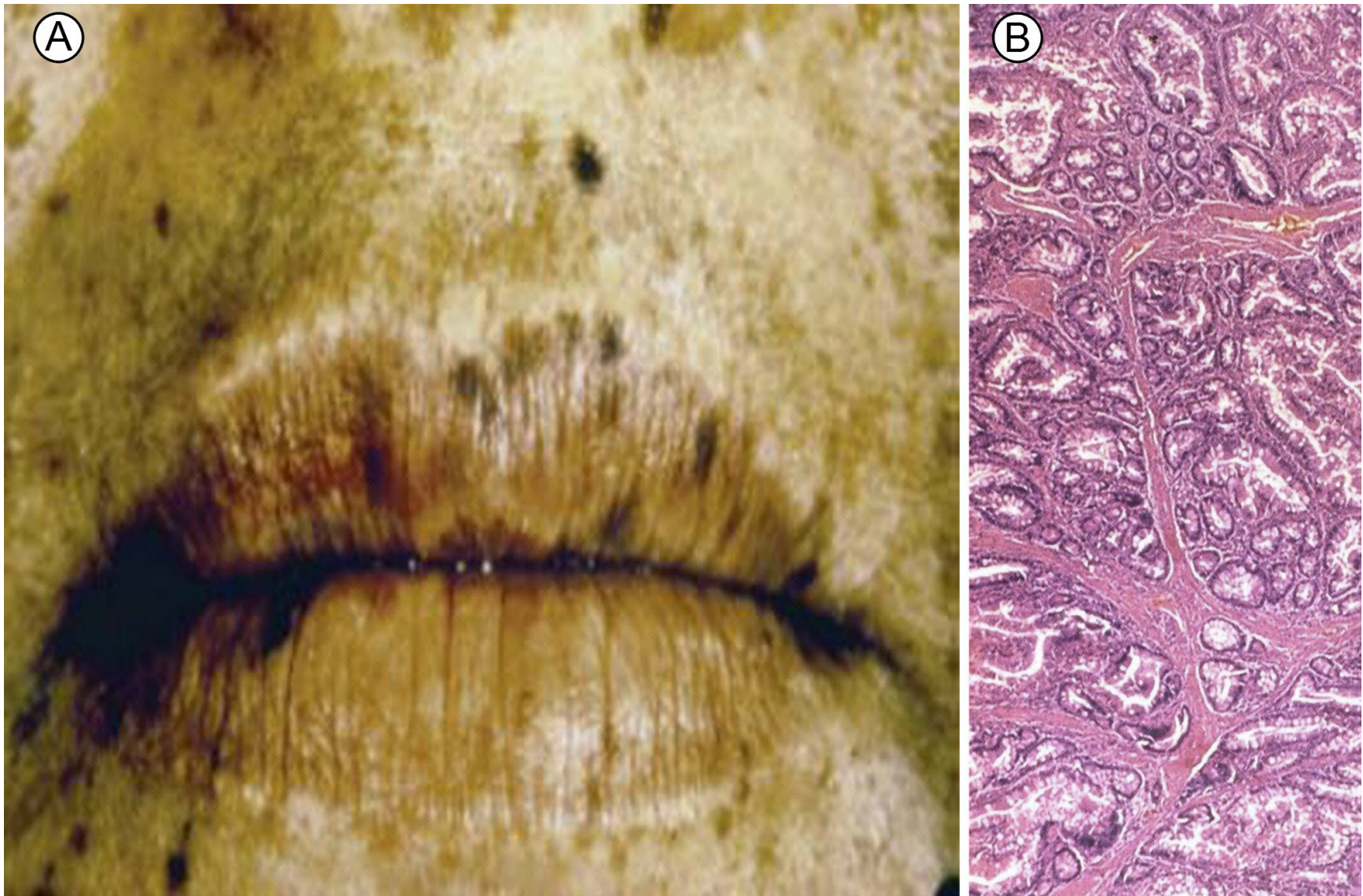
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22.75 Gardner syndrome.



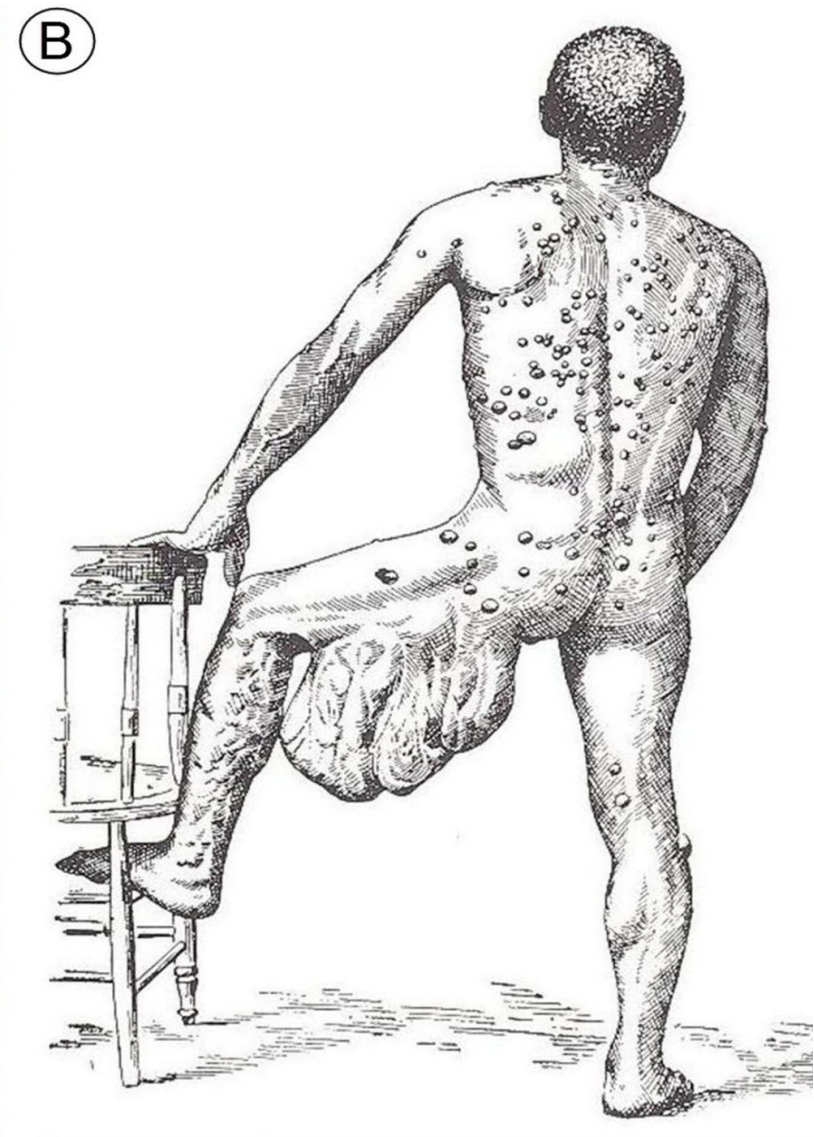
**Picture 22-75** Gardner syndrome. **A** Clinical picture of cutaneous epidermal cysts. **B** Histology of cutaneous cysts. Associated intestinal adenomas with risk of colonic carcinoma at a young age. Other tumors are desmoid and osteomas. (Reproduced with permission, Thiers et al, 2009).

22.76 Peutz-Jeghers syndrome.



**Picture 22-76** Peutz-Jeghers syndrome. **A** Clinical picture, hyperpigmented freckle-like macules of perioral skin and oral mucosa. **B** Associated gastrointestinal polyps with risk of malignancy in small intestine. (Reproduced with permission, Thiers et al, 2009).

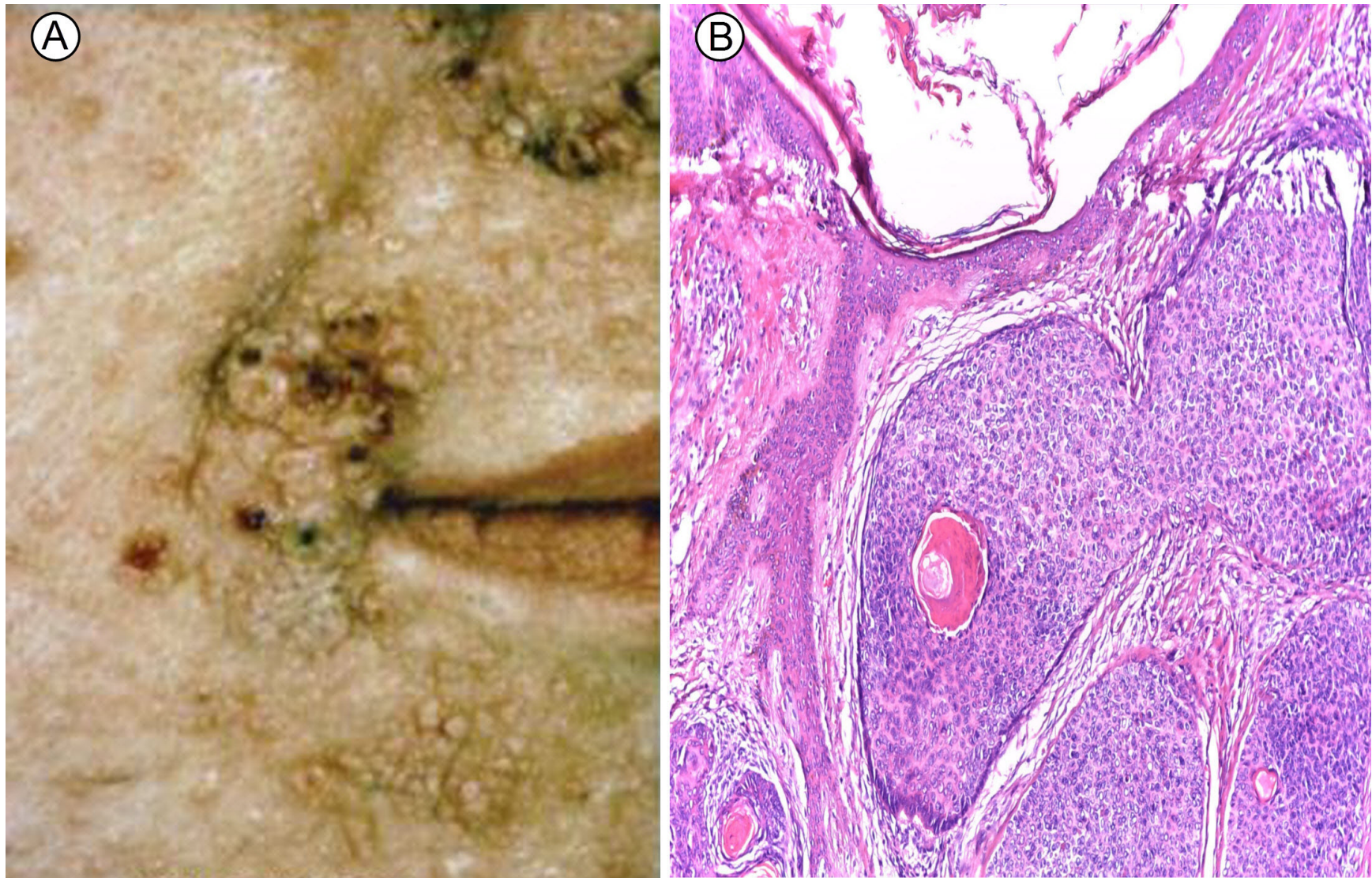
22.77 Neurofibromatosis Type I (von Recklinghausen disease).



**Picture  
22-77**

**Neurofibromatosis Type I (von Recklinghausen disease).** **A** Gross, multiple neurofibromas of back and café au lait macules. **B** Malignant change in sciatic nerve. In MEN-2 B syndrome perioral neuromas are associated with thyroid medullary carcinoma. (Reproduced with permission, Thiers et al, 2009).

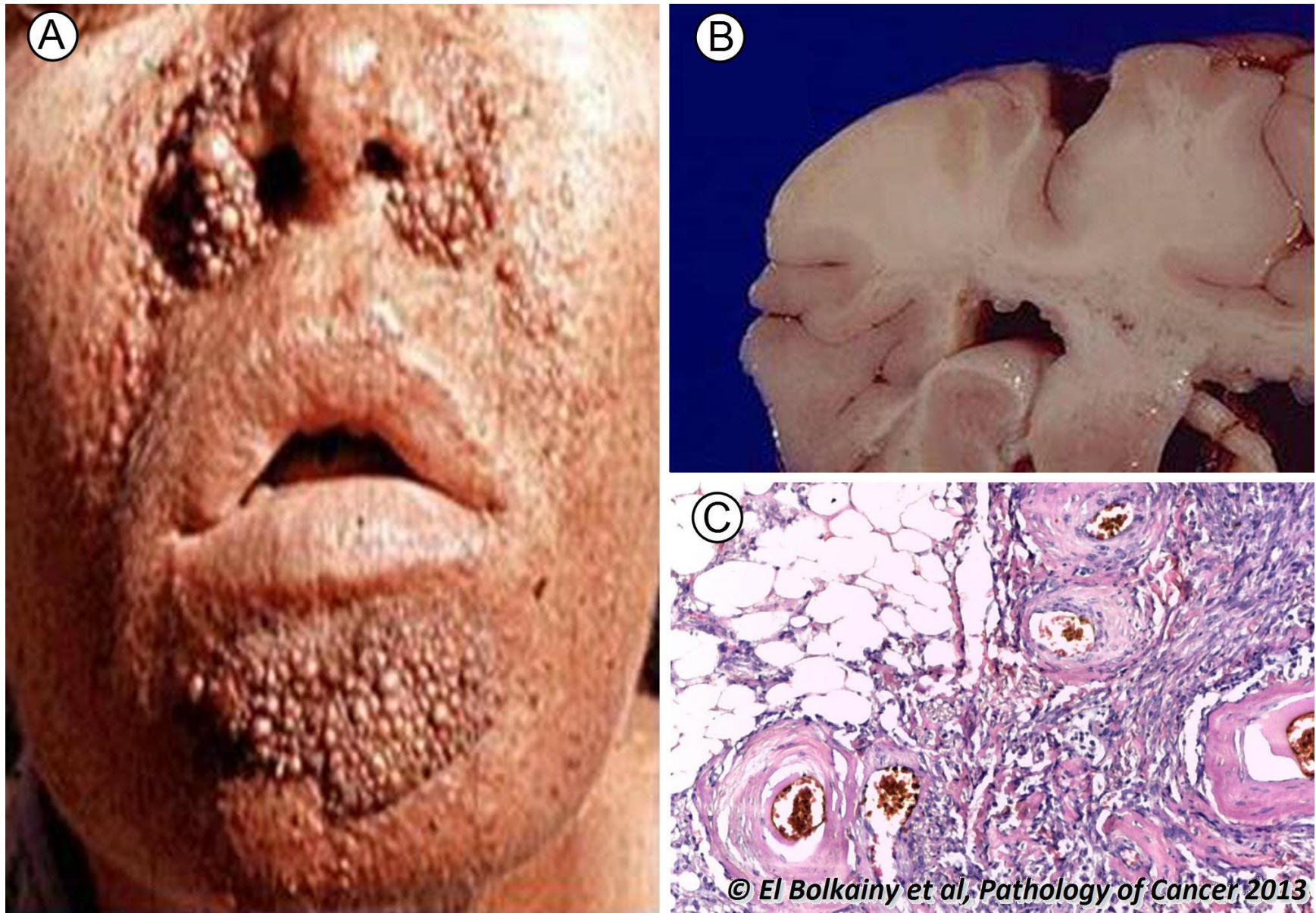
22.78 Cowden syndrome.



**Picture  
22-78**

**Cowden syndrome.** **A** Gross, multiple trichilemmomas affecting head. **B** Trichilemmoma, histology. The internal malignancy include: breast carcinoma (50 %), thyroid (10%) and endometrium (10%). (Reproduced with permission, Thiers et al, 2009).

22.79 Tuberosus sclerosis.

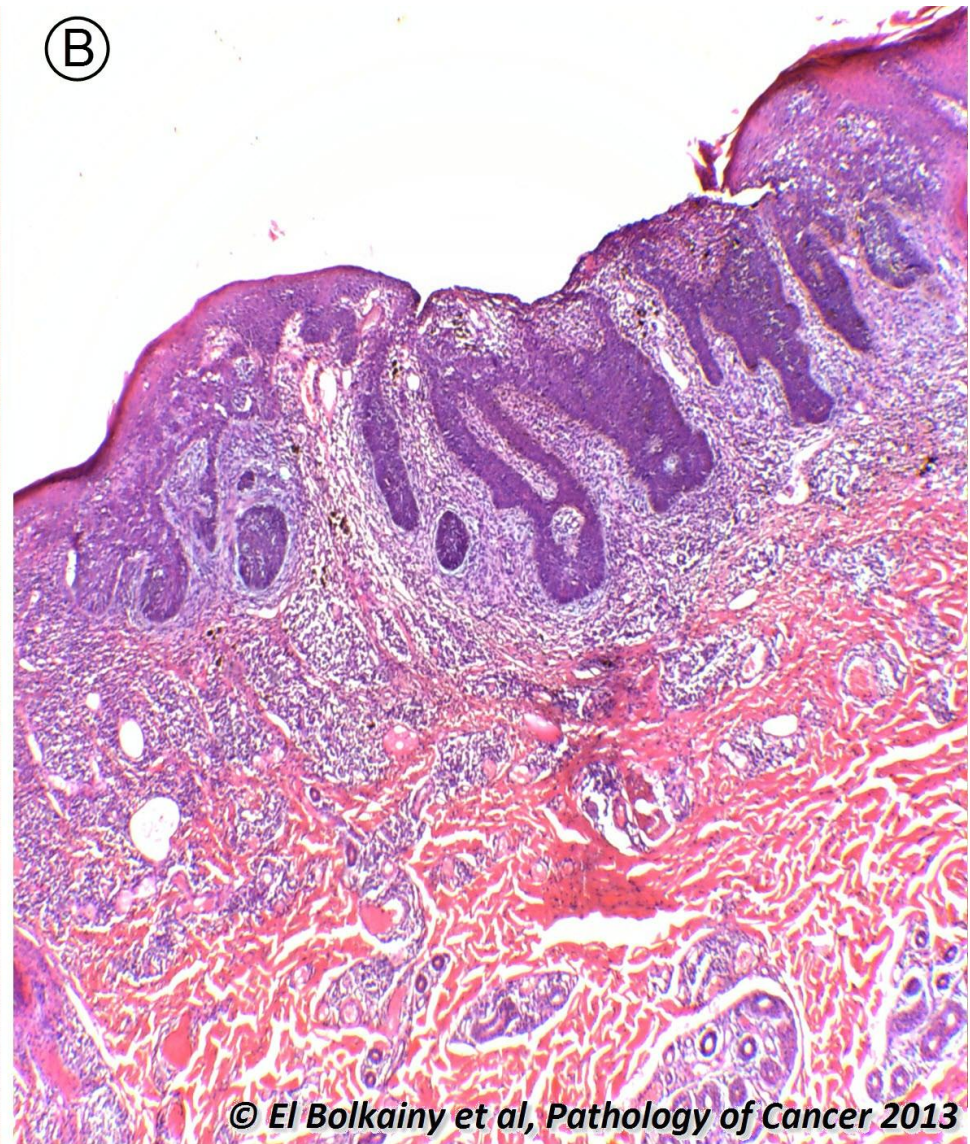


**Picture 22-79** Tuberosus sclerosis. **A** Clinical picture, multiple angiofibromas of skin. **B** Associated so-called subependymal giant cell astrocytoma of lateral ventricle (multiple benign hamartomatous lesions) and **C** angiomyolipoma of kidney.

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22.80 Gorlin syndrome (Nevoid basal cell carcinoma syndrome).



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**Picture 22-80**

**Gorlin syndrome (Nevoid basal cell carcinoma syndrome).** **A** Clinical picture, multiple basal cell carcinoma of face in young adult. Internal tumors include medulloblastoma, meningioma, craniopharyngoma and ovarian fibroma. (Reproduced with permission, Thiers et al, 2009). **B** Basal cell carcinoma, histology.

22.81 Ataxia-telangiectasia (Louis-Bar syndrome)



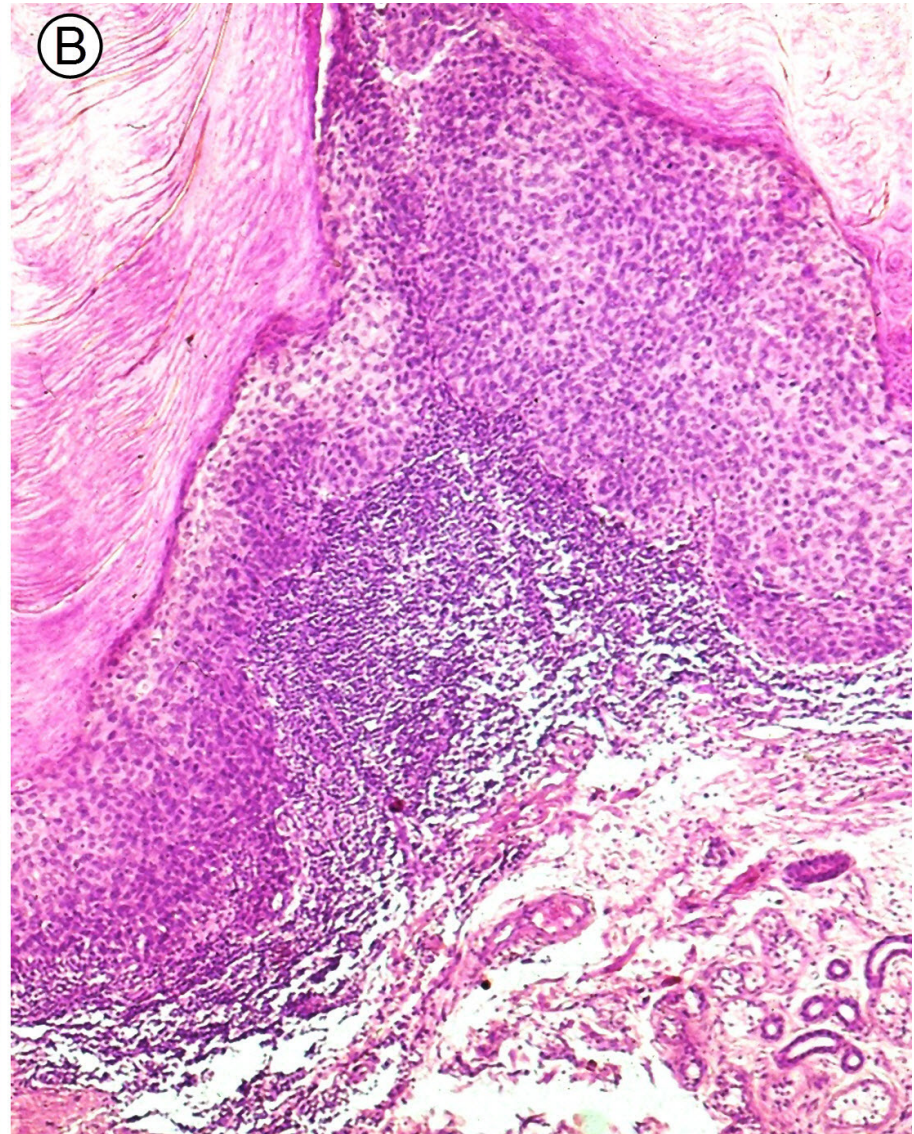
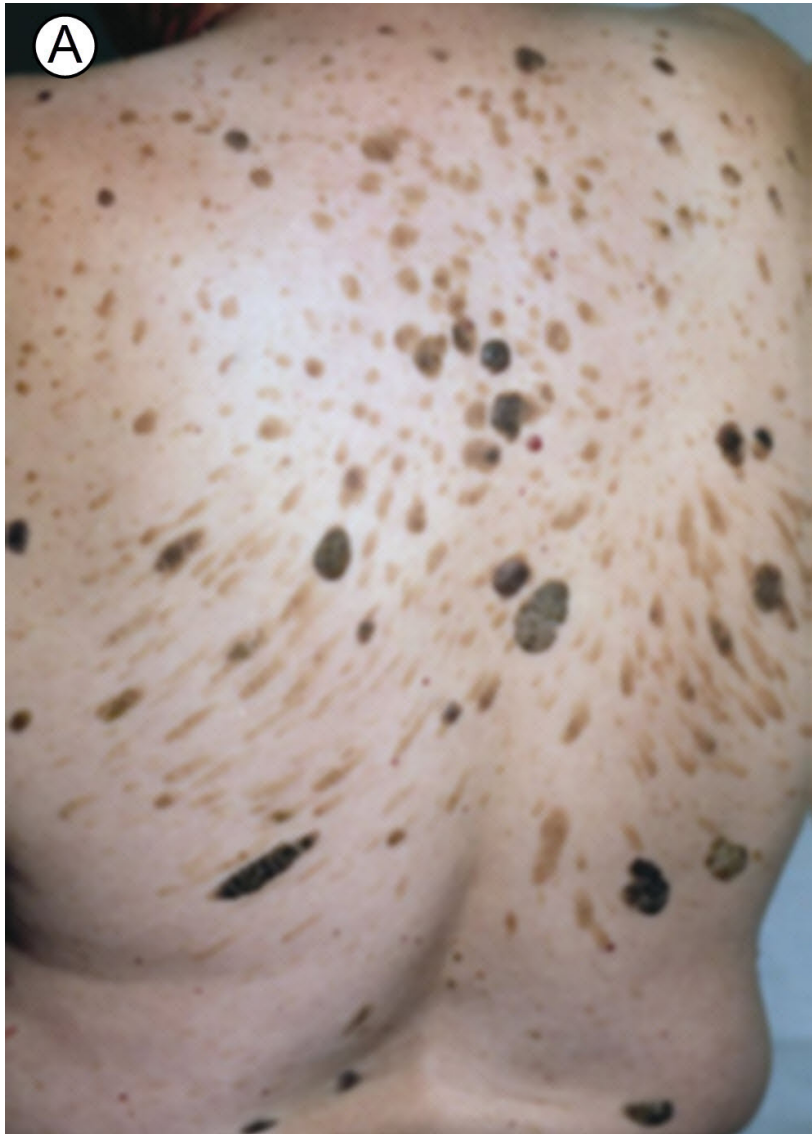
**Picture 22-81** Ataxia-telangiectasia (Louis-Bar syndrome) presents clinically as conjunctival telangiectasia. This inherited immunodeficiency syndrome is complicated by lymphoma and leukemia. (Reproduced with permission, Thiers et al, 2009).

22.82 Acanthosis nigricans.



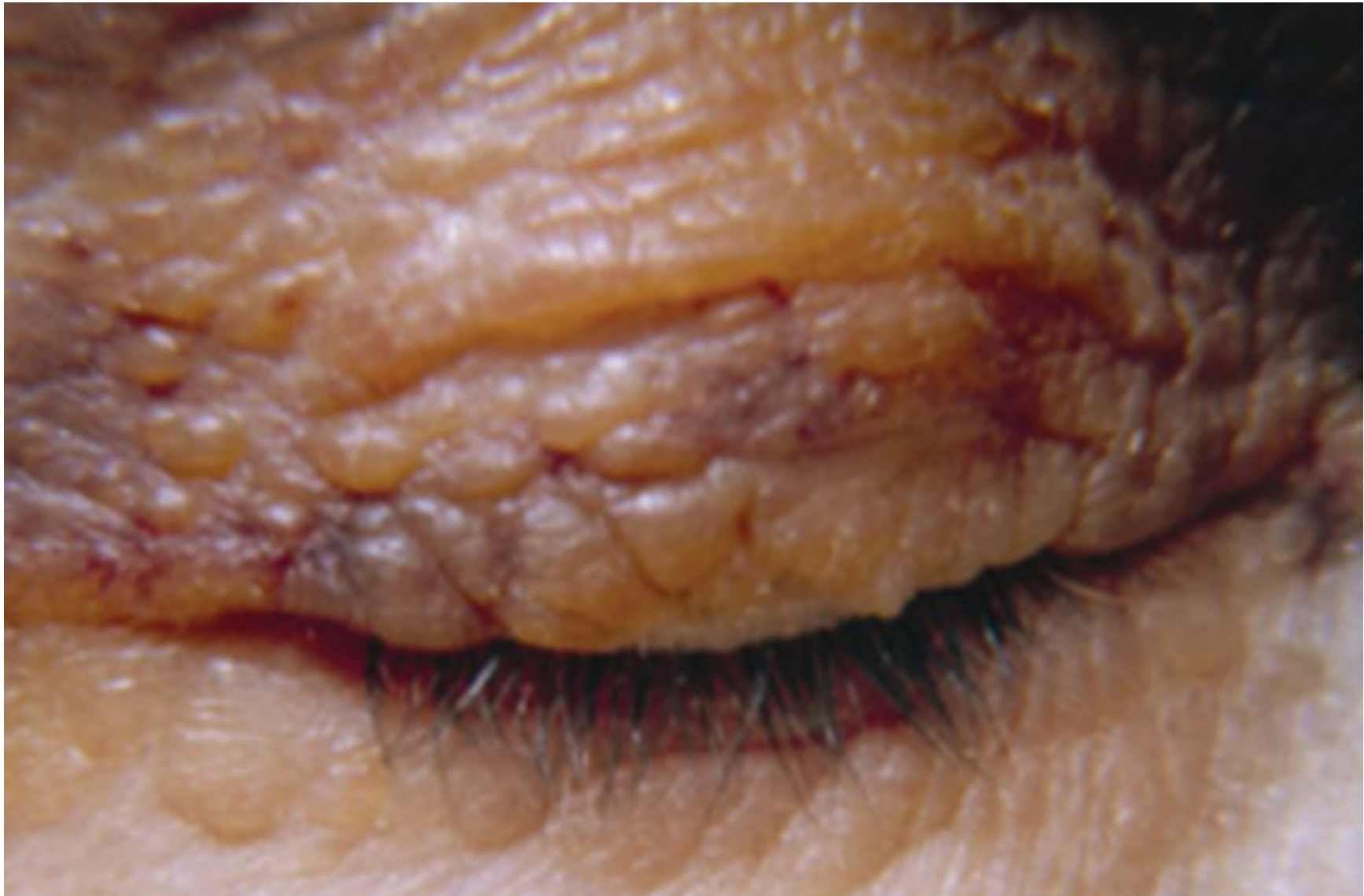
**Picture 22-82** Acanthosis nigricans. **A** Clinically, hyperpigmented areas of skin with velvety surface. **B** Histologically, note papillary hyperplasia and hyperkeratosis with mild melanosis of basal layer. The internal malignancy is adenocarcinoma commonly gastric. (Reproduced with permission, Thiers et al, 2009).

22.83 Seborrheic keratosis.



**Picture 22-83** Seborrheic keratosis. **A** Clinical picture, eruptive seborrheic keratosis, note multiple warty pigmented lesions. **B** Histology shows basaloid proliferation with hyperkeratosis. The internal malignancy is usually gastrointestinal. (Reproduced with permission, Thiers et al, 2009).

22.84 Primary systemic amyloidosis



**Picture 22-84** Primary systemic amyloidosis presents as waxy skin folds and papules in upper eye lid which bleed easily. This may be associated with underlying plasma cell dyscrasia (monoclonal gammopathy or myeloma). (Reproduced with permission, Thiers et al, 2009).

