Keratoacanthoma was first described by Hutchison in 1889 as “crateriform ulcer of the face.” Since then several variants, both solitary and multiple, have been described. Generally these variants have a benign clinical course with spontaneous resolution. Uncommonly, keratoacanthomas may be associated with aggressive histologic features, such as perineural invasion and lymphovascular invasion; however, these features do not appear to have adverse impact on prognosis. [1-5] Multinodular keratoacanthoma is characterized by multiple keratoacanthoma nodules at the periphery of a progressively expanding tumor with central resolution.[6-8] Generalized eruptive keratoacanthoma of Grzybowski is characterized by a generalized eruption of hundreds to thousands of follicular papules with marked facial involvement, which can lead to masked facies and ectropion. Individual tumors heal over weeks to months, but new tumors continue to occur.[9, 10]

Our patient developed a unique plaque-like lesion studded with multiple cystic tumors immediately adjacent to where she had developed a previous conventional solitary keratoacanthoma. A similar case has been previously reported in the literature by Washington et al.[11] Their patient developed multiple keratoacanthomas in the setting of immunosuppression, including a 4.5 x 3.5 cm scaly, erythematous plaque studded with numerous milia-like papules, nodules, and small cysts on the right cheek. The authors referred to this lesion as eruptive keratoacanthoma en plaque.

Aggressive histologic features such as perineural and lymphovascular invasion in keratoacanthoma are uncommon with a limited number of reported cases.[12,13] The English literature includes four case series and several case reports, which cite perineural invasion to occur in 1-4% cases.[3, 5] Perineural invasion, found within the adjacent dermis as well as subcutaneous adipose fat and skeletal muscle, has been noted at distances of up to 2 cm from the center of the lesion by Janeka et al[4] and seen to affect multiple nerves, up to 59, within a single resection. [2] Keratoacanthomas with perineural invasion are noted for their predilection for the head and neck and generally favorable clinical course. In a review of 73 cases reported in literature, 57 occurred on the head and neck. The increased incidence in the head and neck region is thought to be due to the more superficial location of large neurovascular bundles within this region. All cases with perineural invasion were treated with primary excision and a few selected cases with additional radiation, re-excision, and lymph node dissection due to concern over the more aggressive histologic features. With a follow-up ranging from 6 months to 12 years, one recurrence is cited by Gobalt et al, which they contribute to an incomplete excision. No metastatic disease has been reported. Also of note, several cases of keratoacanthoma with perineural invasion present at the resection margins did not develop recurrence, despite no further treatment.

Eruptive Keratoacanthoma en plaque
Although less frequent, lymphovascular invasion in association with keratoacanthoma has also been previously described in cases with a predilection for the head and neck area.[1, 2, 4] Cooper et al reported cases affecting large vessels (0.2-0.5 mm diameter) as well as multiple veins within a single resection.[2] Intravascular invasion up to 3 mm from the tumor was reported in Caljone et al.[1] With a follow-up of up to 8 years, no metastatic disease has been reported.

The literature and our case suggest that perineural and lymphovascular invasion may not necessarily predict an aggressive clinical course when seen in keratoacanthomas of the head and neck. With the predilection of keratoacanthomas with perineural invasion for the face, careful consideration should be made before aggressive treatments, including re-excision, are pursued in this anatomic area.

REFERENCES:

Superficial granulomatous pyoderma

Superficial granulomatous pyoderma (SGP) is a rare chronic inflammatory disorder that severely disfigures patients.

Microscopic examination reveals superficial ulcers and abscesses. Superficial dermal abscesses of neutrophils are noted in close relationship to proliferative epidermis at the edges of ulcers or forming sinus tracts. Acanthosis and pseudoepitheliomatous hyperplasia are seen. The abscesses/granulomas are frequently formed in three layers: the innermost layer with neutrophils, the surrounding layer with histiocytes and giant cells, and the outermost layer of plasma cells.

Although considered a superficial, vegetative variant of pyoderma gangrenosum (PG), SGP exhibits certain clinical, histologic, and prognostic features that distinguish it from PG: SGP is indolent and slowly progressive, exhibits chronic granulomatous inflammation on histology, and typically lacks association with underlying systemic disease.

REFERENCES:

Metastatic intravascular endometrial adenocarcinoma

A 65-year-old woman presented with erythematous and purpuric plaques on both thighs and massive edema of both lower extremities. Four years prior to that the patient was diagnosed with stage IV endometrial adenocarcinoma, endometrioid type. Hysterectomy and bilateral salpingo-oophorectomy was performed. Both ovaries and adjacent soft tissue showed extensive involvement by cancer. One common iliac lymph node was positive. The patient underwent chemotherapy and subsequently developed deep venous thrombosis and stroke. A biopsy from rash on the right leg was signed out as spongiotic/eczematous dermatitis. The large atypical neoplastic cells within vessels were not noted. A subsequent biopsy 5 months later from the left leg rendered a correct diagnosis of metastatic intravascular endometrial adenocarcinoma.

CAM 5.2 low molecular weight cytokeratin has been reported to be positive in 100% of endometrial adenocarcinomas.

The following entities should be considered in the differential diagnosis:

1. **Intravascular lymphoma**
   Intravascular lymphoma (angiotropic lymphoma) is defined as the intravascular proliferation of clonal lymphocytes with little to no involvement of the organ parenchyma. The diagnosis is established by histologic examination of a skin biopsy and immunohistochemical stains. Intravascular lymphomas have large cell morphology, i.e. the malignant cells are two or more times the size of a normal lymphocyte, and typically have a prominent nucleolus. Most cases of cutaneous intravascular lymphoma show a B–cell phenotype but a T–cell variant of the disease has been reported.

2. **Reactive angioendotheliomatosis**
   Clinically presents as red-brown or violaceous nodules or plaques over the face, arms, and legs. It may be associated with underlying disease such as infection, subacute bacterial endocarditis, cryoproteinemia, leukemia etc. Histologically there are increased numbers of variably dilated vessels, mainly capillaries, with intraluminal proliferation of plump endothelial cells. The lumina are often obliterated by endothelial cells or fibrin thrombi. Focally, “glomeruloid” blood vessels, especially in cases associated with cryoglobulinemia, are seen.

3. **Intravascular histiocytosis**
   Intralymphatic histiocytosis is a rare condition characterized by the presence of dilated lymphatic vessels containing aggregates of mononuclear histiocytes within their lumina. The phenomenon seems to occur almost exclusively in the reticular dermis. Clinically, lesions are located predominantly on the upper and lower limbs and consist of asymptomatic and poorly demarcated erythematous plaques and livedo reticularis-like lesions. Double immunohistochemistry with podoplanin and CD68 shows endothelial cells positive for podoplanin and intralymphatic histiocytes positive for CD68.

References: