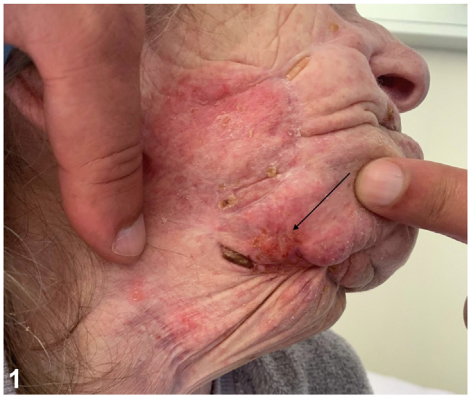
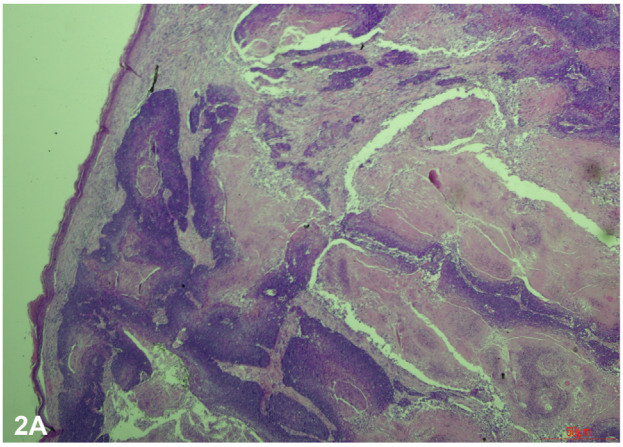
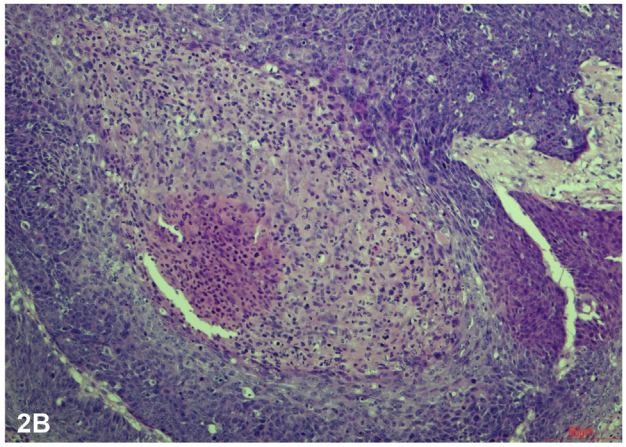
A rapidly-growing erythematous nodule on the right upper neck

Siobhan Rafferty, MB, BCh, BAO,[a] Paula Finnegan, MB, BCh, BAO,[a] Brian Ryle, MDS,[b] Vourneen Healy, MB, BCh, BAO,[c] Mark Wilson, MB, BCh, BAO,[b] and Sinead Field, MB, BCh, BAO[a]

Key words: adnexal tumor; ghost cells; hair follicle matrix; nodule; pilomatrical carcinoma.







From the Charles Centre for Dermatology, University Hospital Limerick, Limerick, Ireland[a]; Department of Maxillofacial Surgery, University Hospital Limerick, Limerick, Ireland[b]; and Department of Pathology, University Hospital Limerick, Ireland.[c]

Funding sources: None.

IRB approval status: Not applicable.

Patient consent: Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

Correspondence to: Siobhan Rafferty, MB, BCh, BAO, Charles Centre for Dermatology, University Hospital Limerick, Limerick, Ireland, V94 F858. E-mail: siobhanrafferty@gmail.com.

JAAD Case Reports 2023;37:134-6.

2352-5126

https://doi.org/10.1016/j.jdcr.2023.05.024

HISTORY

A rapidly-growing, eroded, mobile, erythematous nodule on right upper neck adjacent to an old hemangioma scar was incidentally noted in a 67-year old female who presented for follow up after excision of a squamous cell carcinoma on her left cheek 10 weeks earlier (Fig 1). There was a surrounding transient nonspecific dermatitis extending over right cheek (Fig 1).

Comorbidities included hypothyroidism post thyroidectomy, parathyroid adenoma, osteoporosis, congenital mitral valve prolapse, atrial fibrillation, esophageal stricture and intellectual disability.

Delineating margins and assessing for lymphadenopathy was challenging as she had surgeries in childhood for oropharyngeal haemangiomas. Computed tomography of the neck due to challenging anatomy showed normal calibre lymph nodes.

Hematoxylin-eosin staining (Fig 2, *A* and *B*) and immunohistochemistry were performed following complete excision.

Question 1: Given the history and images, what is the most likely diagnosis?

A. Epidermoid cyst

B. Squamous cell carcinoma

C. Pilomatrical carcinoma

D. Basal cell carcinoma

E. Pilomatrixoma

Answers:

A. Epidermoid cyst — Incorrect. Epidermoid cysts typically present as firm, skin colored or yellowish papules or nodules in middle aged adults. They generally have a central punctum and are mobile. They are characterized histologically by unilocular or multilocular cystic structures in the dermis. Cyst rupture can result in foreign body granulomatous reaction in adjacent dermis.

B. Squamous cell carcinoma — Incorrect. Squamous cell carcinomas are keratinocyte carcinomas that typically present as tender, painful lesions on sun-exposed sites that may ulcerate. Typical histological features include conspicuous keratinisation with nests of squamous epithelial cells extending into the dermis.

C. Pilomatrical carcinoma — Correct. Pilomatrical carcinomas are rare, locally aggressive adnexal tumors arising in the hair follicle matrix. They usually present in males in the sixth or seventh decades of life on the head and neck. The most common clinical presentation is a solitary non-tender firm dermal swelling. Diagnosis is based on histological examination; basaloid cells with atypia and a raised mitotic index with or without ghost cells are the commonest pathological finding.[1]

D. Basal cell carcinoma — Incorrect. Although basal cell carcinomas can also present as skin colored nodules on the head and neck, they are typically slow growing with clinically distinct features on both gross examination and dermoscopy.

E. Pilomatrixoma — Incorrect. Differentiating between benign and malignant pilomatrixomas remains challenging in clinical practice. Pilomatrixomas also present as skin colored, red or white lesions on the head and neck, however they typically remain stable for years or grow slowly in size. Immunohistochemistry is also not able to reliably distinguish between benign and malignant pilomatrical (hair follicle) tumors.[2]

Question 2: What is the most appropriate treatment for this patient?

A. No treatment

B. Complete surgical excision with clear margins

C. Primary radiotherapy

D. Systemic chemotherapy

E. Wide local excision with adjuvant radiotherapy

Answers:

A. No treatment — Incorrect. Pilomatrical carcinoma is a malignant adnexal tumor. Metastasis occur in 10%-16% of cases; lung and regional lymph nodes are the most common sites for distant metastasis.[1]

B. Complete surgical excision with clear margins — Correct. Standards for surgical management are not well defined given the rarity of this malignancy. Complete margin control is essential given the aggressive nature of this tumor.[3] Surgical excision with wide margins is the current mainstay of treatment. Local recurrence rate is high (between 50% to 60% after simple excision).[1] Most studies recommend complete excision with wide margins; recommendations range between 5 and 30 mm margins.[1,4] Mohs micrographic surgery has been

reported as a successful treatment for pilomatrical carcinoma, achieving complete margin control and no local recurrence at 5-6 months.[3]

C. Primary radiotherapy — Incorrect. Complete surgical excision is the current mainstay of treatment; however, radiotherapy may be a good option in patients in whom wide excision is not possible.[4]

D. Systemic chemotherapy — Incorrect. Systemic chemotherapy has been used in systemic disease but there is a poor response.[1]

E. Wide local excision with adjuvant radiotherapy — Incorrect. Adjuvant radiotherapy is recommended in recurrent or residual macroscopic disease.[1]

Question 3: Which of the following histopathologic features is present in pilomatrical carcinoma?

A. Islands of large oval squamous cells with eosinophilic cytoplasm, vesicular nuclei and evidence of clear cell differentiation

B. Nodules, nests and trabeculae of neoplastic cells within the dermis with round nuclei, scant cytoplasm, conspicuous mitoses and apoptotic bodies

C. Irregularly shaped nests of large anaplastic basaloid cells with an infiltrative growth pattern, follicular matrical differentiation and intra-tumoral melanocytic hyperplasia

D. Basaloid lobules with peripheral palisading and the presence of shadow cells

E. Solid nests of basaloid cells undergoing trichilemmal-type keratinization with foreign body reaction and calcification

Answers:

A. Islands of large oval squamous cells with eosinophilic cytoplasm, vesicular nuclei and evidence of clear cell differentiation — Incorrect. These are features of clear cell squamous carcinoma.

B. Nodules, nests and trabeculae of neoplastic cells within the dermis with round nuclei, scant cytoplasm, conspicuous mitoses and apoptotic bodies — Incorrect. These are histopathological features typically seen in Merkel cell carcinoma, a cutaneous neuroendocrine carcinoma typically affecting older and immunosuppressed individuals.

C. Irregularly shaped nests of large anaplastic basaloid cells with an infiltrative growth pattern, follicular matrical differentiation and intra-tumoral melanocytic hyperplasia — Correct. Pilomatrical carcinomas also demonstrate central areas with necrotic debris, shadow cells, clear cytoplasmic cells, transition to squamous cells, invasion of blood and lymphatic vessels and surface ulceration.

D. Basaloid lobules with peripheral palisading and the presence of shadow cells — Incorrect. The combination of typical histological features of BCC and pilomatrixoma indicates a diagnosis of basal cell carcinoma with matrical differentiation. This is rare variant of BCC but also typically presents in men at an average age of 69 on the head and neck.[5]

E. Solid nests of basaloid cells undergoing trichilemmal-type keratinisation with foreign body reaction and calcification — Incorrect. These are the typical features of pilomatrixoma, a benign hair follicle tumour.

Conflict of interest

None disclosed.

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