keratinising foci and had weak or absent cytokeratin and/or p63 expression. TTF1 expression was not reported in these tumours.

Together with this case, these studies suggest that it may not be uncommon for NC to show either TTF1 expression, or lack squamous differentiation. This may be an important potential source of misdiagnosis. NC with TTF1 expression could be erroneously classified as either lung adenocarcinoma or thyroid carcinoma. NC lacking expression of squamous markers and weak/negative low molecular weight cytokeratin expression may be classified as a sarcoma, a carcinoma of unknown primary or an undifferentiated malignancy. The likelihood of such errors would be compounded for NC with an atypical clinical presentation or variant tumour morphology, as in such cases NUTM1 IHC is less likely to be performed. For these reasons it would seem judicious to have a low threshold for screening high grade or poorly differentiated malignancies by NUTM1 IHC or FISH, and in positive cases to perform RNA sequencing to identify the specific NUTM1 fusion partner in order to confirm a diagnosis of NC.

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Keratosis pilaris with adjacent haemosiderin deposition: a clue to scurvy

Sir,

We present the case of a 50-year-old man with a misleading 'classical' presentation and the histological assessment which was the key to his diagnosis. This patient developed a purpuric rash several weeks prior to his hospital presentation. The rash arose in his lower limbs which rapidly progressed proximally with increasing generalised pain. It was non-blanching, confluent, palpable and purpuric in nature. Given that the clinical picture resembled typical features of a vasculitic rash, extensive laboratory tests including haematological, biochemical and immunological tests were performed. A biopsy of the rash was undertaken which marked a critical turning point in his clinical course.

Two skin punch biopsies were received for histological assessment. Both biopsies showed a superficial perivascular lymphocytic infiltrate. In one biopsy, keratosis pilaris with perifollicular haemosiderin deposition was present (Fig. 1A–D). No vasculitis was seen and there were no extravasated red blood cells. Perls stain (used to highlight iron) was performed which demonstrated perifollicular haemosiderin deposition (Fig. 1E,F). The combination of keratosis pilaris and perifollicular haemosiderin deposition raised the strong possibility of scurvy. The histology result was conveyed back to the treating clinicians which prompted the investigation of scurvy.

A review of systems revealed that the patient had not consumed fresh vegetables or fruits for the past 10 years. He did not eat regularly and had been consuming large amounts of alcohol since his employment had been terminated. Further examination revealed gingival bleeding and ecchymosis. His body mass index was borderline underweight at 18.9 kg/m² and he had lost 16 kg (21% weight loss) in the preceding 2 years. This patient's serum ascorbate (vitamin C) level was indeed low at 9 µmol/L (normal range 20–120 µmol/L), further supporting the diagnosis of scurvy. The diagnosis of vasculitis was dismissed, and a nutritional screen and supplementation were undertaken. Unfortunately, dermatological specialty opinion was not sought nor was a comprehensive skin examination performed, which may have revealed some of the key cutaneous findings of scurvy as discussed below.

Fig. 1 Histological features of scurvy in our case. (A–D) H&E stain. (A,B) Keratosis pilaris is present with corkscrew-like hairs buried in keratotic follicular material. (C,D) Perifollicular haemosiderin deposition. (E,F) Perl's stain was used to highlight perifollicular haemosiderin deposition (haemosiderin stained blue).

The clinical manifestations caused by vitamin C deficiency are called scurvy. As the human body is unable to synthesise vitamin C, adequate body supply depends wholly on oral intake and absorption.¹ Vitamin C is abundant in a variety of foods including fruits, vegetables, animal products and milk. The recommended daily intake for vitamin C for an average adult is 75–90 mg, which all but the most restrictive diets provide.²

One of the main functions of vitamin C is collagen synthesis. This is important for the structural integrity of blood vessels, skin development and normal hair growth. As a result of impaired collagen formation, one of the most common presentations is small vessel haemorrhage resulting from minor bruising due to abnormal vessel wall fragility. Characteristically, this is perifollicular in distribution but spontaneous petechiae and ecchymoses can also develop.³ The haemorrhagic skin lesions are initially flat but may coalesce and become palpable, especially on the lower extremities.⁴ Other typical features such as follicular hyperkeratosis and corkscrew hairs may also be visible.³ Unless one has clinical suspicion for scurvy, these signs can be easily mistaken as signs of vasculitis as they have strongly

overlapping features. However, a thorough skin examination reveals the distribution of the rash seen in scurvy to be perifollicular in nature, differentiating it from other entities such as vasculitis.⁵

The histological findings of scurvy in the skin include follicular hyperkeratosis with coiled, fragmented, corkscrew-like hairs buried in keratotic follicular material, known as keratosis pilaris (Fig. 1A,B). One of the characteristic features is the presence of extravasated erythrocytes around blood vessels in the superficial dermis in a perifollicular distribution. Haemosiderin deposition as a result of previous haemorrhage is also found and this can be highlighted using Perl's stain (Fig. 1E,F). Other features that can also be seen include perifollicular fibrosis, perivascular inflammation and ulceration of the skin. There should be no evidence of vasculitis. Electron microscopy, which is often not required for the diagnosis, shows alterations in fibroblasts with defective collagen formation. There may also be alterations in the endothelial cells of vessels and their junctions.⁵

At times, the true diagnosis is only apparent on histopathology such as in our case. As the incidence of scurvy is rare in the Australian population, the clinical suspicion is low and

may not be immediately considered. However, the histological features of keratosis pilaris with adjacent haemosiderin deposition formed a clue to scurvy in our case. Being able to make a histological diagnosis in situations with misleading or overlapping clinical presentations is critical in driving appropriate clinical care.

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A unique composite tumour of the adrenal gland in a paediatric patient, with adrenal cortical and neural crest cell-like components

Sir,

The adrenal gland has a dual embryological origin, with adrenal cortical cells being derived from the intermediate mesoderm in the area of the urogenital ridge and medullary chromaffin cells being derived from the neural crest.¹ Due to this developmental background, a diverse range of tumours may occur in the adrenal, including occasional composite tumours. We present a case of a unique composite tumour presenting in the adrenal gland of a paediatric patient.

A 7-year-old girl was referred to the paediatric surgical department after an ultrasound for recurrent abdominal pain identified an adrenal mass. She had no significant medical history, no family history of note, and no dysmorphic features or cutaneous stigmata to suggest an underlying syndrome. The ultrasound showed a heavily calcified adrenal mass, with an imaging differential of teratoma or myelolipoma.

A primary excision was performed, and the pathology department received a soft tissue mass weighing 39 g, with a fibrotic to calcified cut surface. Histological examination showed a circumscribed tumour mass with extensive degenerate changes and a rim of compressed normal adrenal gland at one end. Approximately 95% of the tumour mass comprised areas of dense fibrosis admixed with mature adipose tissue, foci of calcification and ossification. In this fibrotic background, small viable nests of tumour cells were present (Fig. 1) which displayed a cytomorphology distinct to the included normal adrenal. They comprised large epithelioid cells with abundant eosinophilic cytoplasm, vesicular nuclei and prominent nucleoli. Immunohistochemistry revealed an adrenal cortical-type phenotype, with positivity for CK8/18, inhibin, synaptophysin and Melan-A. Focal cells also showed positivity for PHOX2B. Also present were distinct tumour cell nests showing an admixture of epithelioid cells and spindle cells (Fig. 2). The epithelioid cells in these areas showed an intermediate phenotype on immunohistochemistry, being positive for CK8/18 and inhibin, but negative for synaptophysin and Melan-A. The spindle cells showed wavy nuclei and pale eosinophilic cytoplasm and were immunoreactive for NFP and S100. No neuroblastic or ganglion-type cells were identified and immunohistochemistry for chromogranin was negative throughout the tumour. No areas of significant mitotic activity, atypical mitoses, pleomorphism or necrosis were seen. The tumour was felt to be best classified as an adrenal cortical adenoma with extensive degeneration and focal unique neural crest cell-like differentiation.

Adrenal cortical neoplasms are rare in the paediatric population, with a reported incidence of 1 in 500,000 boys or 1 in 87,000 girls. The majority of such tumours in children will be classified as adrenal cortical carcinomas.² The median age of diagnosis is 3–4 years, and the most frequent presentation is with paraneoplastic endocrine phenomena, particularly virilisation in girls.^{2,3} Syndromes and other phenotypic abnormalities are reported in association with adrenal cortical neoplasms in children in some cases, including hemihypertrophy, neurofibromatosis type 1, Beckwith–Wiedemann syndrome, Li–Fraumeni syndrome and other low penetrance p53 mutation disorders, and congenital anomalies of the urinary tract.^{2–4} In this case, clinical work-up did not reveal any features concerning for a familial syndrome or congenital abnormality.

The majority of composite adrenal tumours, as described in the World Health Organization Classification of Tumours of Endocrine Organs,⁵ derive from the medulla. They typically present as a pheochromocytoma composite with a neuroblastic tumour of varying levels of differentiation. Composite tumours with components from both the cortex and medulla are exceedingly rare, existing in the literature only as isolated case reports. They have been described only in adults, and occur nearly exclusively in women. Benign examples of such tumours have been reported as showing admixed adrenal cortical cells and pheochromocytes,^{6,7} or a mix of adrenal cortical cells, pheochromocytes, and ganglioneuroma.⁸ Additionally, malignant examples have been reported, including a malignant mixed corticomedullary tumour,⁷ and a tumour with admixed adrenal cortical morphology and areas of undifferentiated spindle cell/primitive neuroectodermal-