Venous malformation mimicking epidermoid cyst at the peri-ocular location in adults

Sir,

We hereby report an observation seen in six patients over the last five years. They had skin-coloured peri-ocular swellings clinically suspected to be epidermoid cysts, which turned out to be cavernous haemangiomas on histopathological examination. Cavernous haemangiomas are venous malformations formed by a dermal or subcutaneous collection of dilated vascular channels, lined by flattened endothelial cells and thick fibrous septae.

All patients had a similar presentation— a single asymptomatic skin-coloured oval swelling at peri-ocular location [Figure 1a]. The swellings were firm and slightly mobile but not adherent to the overlying skin. There was no history of any discharge or presence of a true punctum. A bluish-green hue with surface lobulation, but without any opening, was misinterpreted as a punctum in two cases. We termed this feature of cavernous haemangioma as a 'pseudo-punctum' [Figure 1b]. The swelling was located over the eyelid in three cases, temporal region in two and forehead in one case. All patients reported onset in adulthood, with gradual progression over years to the present size. Epidermoid cyst was the chief clinical diagnosis, although a differential of vascular malformation was considered in one case. During surgical excision, intra-operative bleeding exceeded the expected amount in two cases. Three excised lesions were brown-black, ill-defined and unencapsulated on gross examination [Figure 1c and d]. No post-operative hematoma was reported in any case.

Histopathological examination revealed multiple large endothelial-cell-lined spaces congested with erythrocytes [Figure 2a]. Two cases showed muscle in the wall, implying a venous component, highlighted by Masson's trichrome [Figure 2b].

Epidermoid cysts are frequently seen over the head and neck. They present as single-to-few, asymptomatic slow-growing skin-coloured ovoid firm swellings within the skin. A punctum, when present, is pathognomonic, but is absent in many cases. Many different lesions may mimic an epidermoid cyst, including developmental cysts, benign cell proliferation such as nodular hidradenoma, as well as primary and secondary malignant lesions such as cutaneous B-cell lymphoma, epithelioid sarcoma, cutaneous meningioma, Merkel cell carcinoma, metastatic lung adenocarcinoma, to name a few.[1] Vascular lesions have not yet been reported to mimic epidermoid cysts; however, the reverse has been seen; true epidermoid cysts may mimic lymphatic malformations.[2]

Cavernous haemangioma of cutaneous venous malformation, presents as a single, small skin-coloured to slightly bluish deep-seated swelling which may be elevated. About 50% of the cases are localised to the head and neck, although virtually any site may be affected. Contrary to conventional venous malformations which present as congenital large bluish-green soft compressible plaques, cavernous haemangiomas are late-presenting small skin-coloured to blue firm swellings.[3] In skin of colour, a blue hue is often not appreciated.

The older term, cavernous haemangioma still dominates pathological and non-dermatological literature. 'Haemangioma' is not appropriate for these lesions, as they are not tumours but malformations. They are probably present at birth, but manifest late and do not regress spontaneously. They have been included under the umbrella term 'slow-flow vascular malformation' in the latest classification by the International Society for the Study of Vascular Anomalies.[4]

Radiological investigations are not helpful in their differentiation from epidermoid cysts. Both entities are predominantly hypoechoic on ultrasound, hyperintense on T1-weighted magnetic resonance imaging and poorly visualised by computed tomography. Doppler imaging is also not helpful as the flow is extremely slow.[5] The calibre of these malformations is usually too small to allow phlebolith formation. The thick fibrous septae and endothelial growth within lesions may introduce hyperechoic areas, but such changes can also be seen in epidermoid cysts. Lastly, the small lesion size at presentation does not allow detailed ultrasonographic evaluation.

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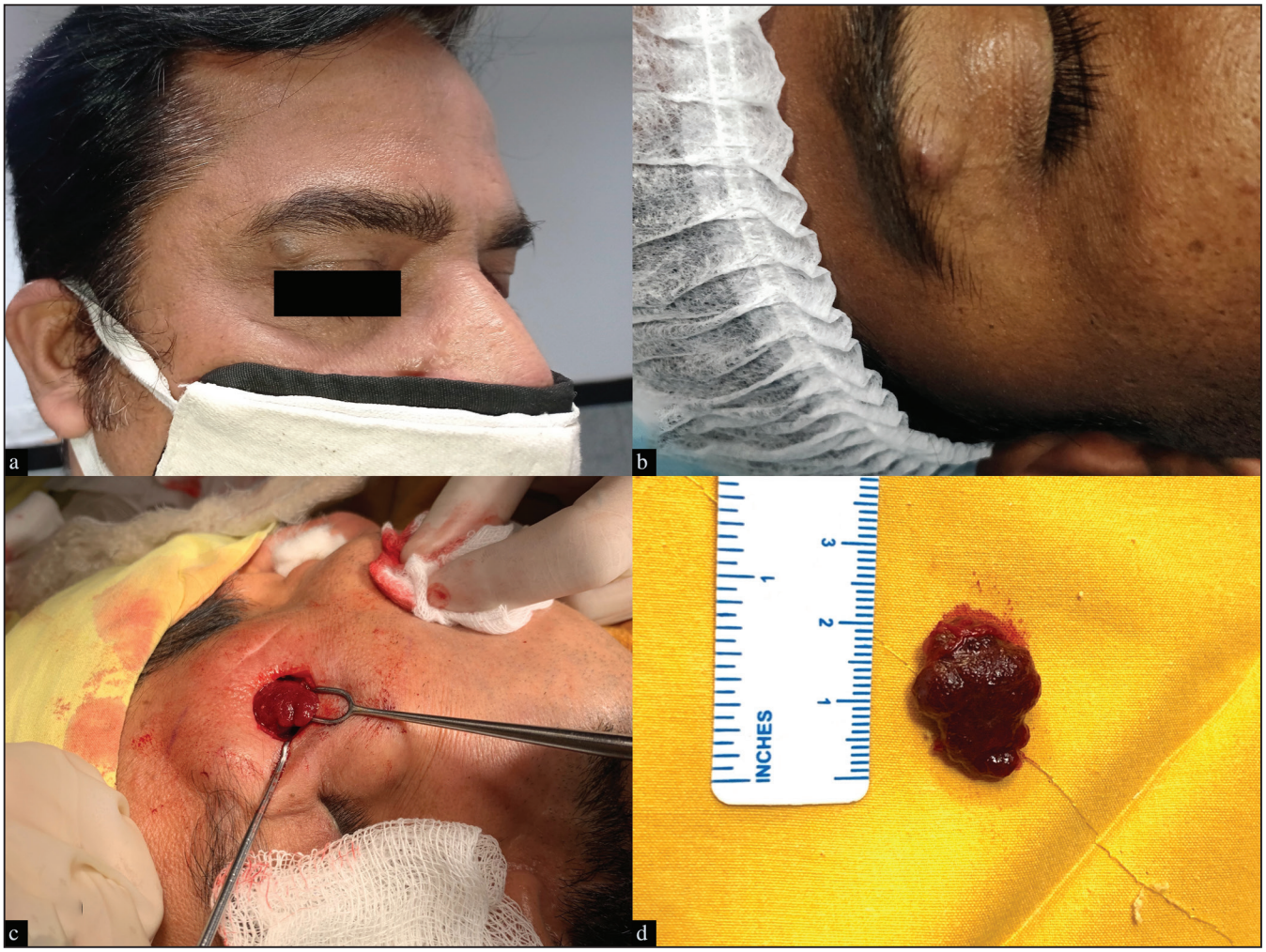

Figure 1: Venous malformations at peri-ocular area in adults mimicking epidermoid cysts. (a) Swelling in the upper eyelid just beneath the eyebrow in Patient 1, with a bluish-hue. (b) A nearly identical presentation in Patient 2, along with a 'pseudo-punctum'. Grossly dark haemorrhagic lesion seen intra-operatively (c) and after excision (d) in Patient 3.

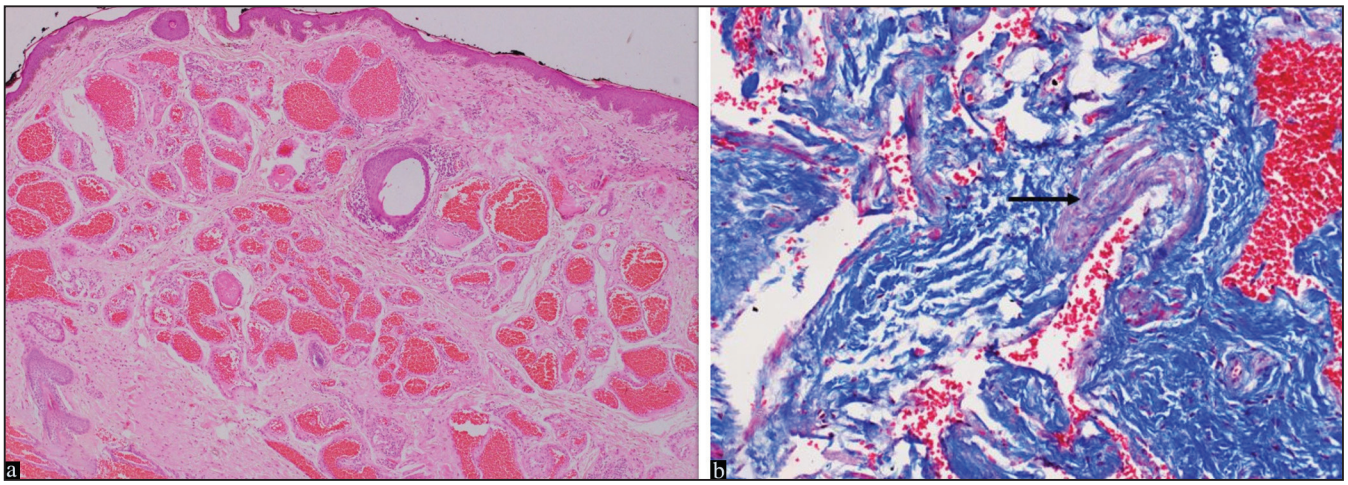

Figure 2: Histopathology of cavernous haemangioma type of venous malformation. (a) Multiple large endothelial-cell lined spaces are congested with erythrocytes throughout the dermis. Haematoxylin and eosin, magnification × 400. (b) Myocytes in the wall, implying a venous component, in one of the specimens. Masson's trichrome, magnification × 1000.

In conclusion, clinical clues to a cavernous malformation include peri-ocular location, lack of true punctum and the presence of a pseudo-punctum. This differential diagnosis of an epidermoid cyst enables the surgeon to suspect a vascular malformation during on-table gross evaluation and change their approach to pre-empt haemorrhagic losses and post-operative hematoma formation.

Declaration of Patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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Pseudoxanthoma elasticum-like changes in longstanding gadolinium-naïve nephrogenic systemic fibrosis in a patient with chronic kidney disease

Sir,

Nephrogenic systemic fibrosis is a rare systemic fibrosing disorder which has been recently described in settings of renal insufficiency, especially with gadolinium exposure.[1] The exact etiopathogenesis is still to be elucidated. Almost no treatment described for the disease has been completely successful. Histopathological features are similar to the sclerodermoid spectrum of disorders. A pseudoxanthoma elasticum like pattern has been rarely reported as an incidental finding in the setting of calciphylaxis. We present such a case in an adolescent with chronic kidney disease.

A 16-year-old boy presented with a history of pruritic skin thickening of one year duration, initially presenting over thighs and later involving the abdomen and legs. It was also associated with multiple, soft swellings which developed from the thickened skin. He was diagnosed with nephrotic syndrome at the age of five years and was on long-term systemic steroids, at a starting dose of 60 mg prednisolone. This dose was tapered gradually to 5 mg till the age of 12 years. He was not on medication at present and was recently detected with stage 5 chronic kidney disease, for which he was on multiple sessions of haemodialysis and erythropoietin injections. Patient also had a history of hypothyroidism and hypogonadotropic hypogonadism, but no exposure to gadolinium. Patient was on 50 μg thyroxine and currently on no drug for hypogonadotropic hypogonadism.

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