

Intravascular Papillary Endothelial Hyperplasia Simulating Malignant Melanoma

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Sir,

Intravascular papillary endothelial hyperplasia (IPEH) was first described by Masson (1) as a neoplastic lesion “vegetant intravascular haemangioendothelioma”. In 1976, Clearkin & Enzinger (2) proposed the term IPEH, which is currently known as a non-neoplastic reactive endothelial proliferation. The lesions are frequently situated in the deep dermis and subcutis of the fingers, head and neck. In subcutaneous localizations the lesion may appear clinically as a small, firm, superficial nodule imparting a red-to-blue discoloration of the overlying skin. We describe here a rare case of IPEH on the lower leg, which resembled malignant melanoma in appearance.

CASE REPORT

A 62-year-old woman presented with a 1-month history of abruptly grown skin lesion on her lower leg. The patient did not recall any preceding trauma. Physical examination revealed a black-coloured, dome-shaped nodule, 8 mm in diameter (Fig. 1). She had been diagnosed with end-stage renal disease 20 years previously, and since then she had received continuous ambulatory peritoneal dialysis (CAPD). Histopathological examination revealed a mass of vascular channels with prominent intraluminal papillary projections in the dermis (Fig. 2a). The papillary projections were composed of a single layer of plump endothelial cells around a core of fibrous connective tissue (Fig. 2b). Characteristically, the

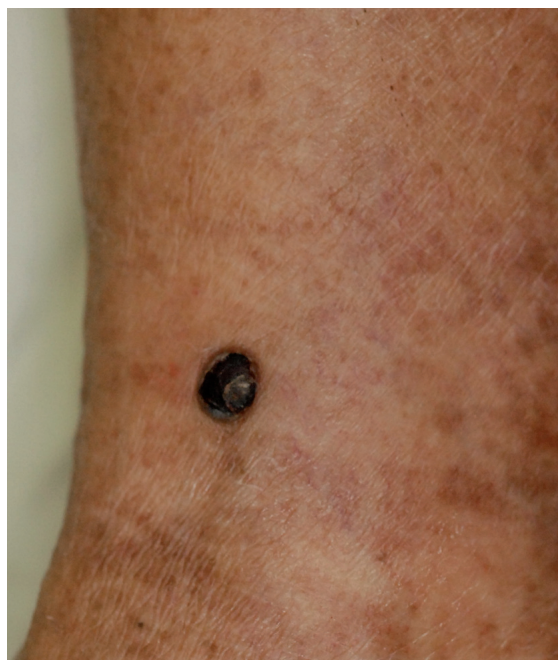


Fig. 1. A 62-year-old woman presented with a black, dome-shaped, firm nodule on her lower leg.

papillary structures were either attached to the internal surface of the vessel wall or apparently lying in the lumen. The endothelial cells showed no significant nuclear atypia or mitotic figures, no necrosis or invasion into surrounding tissues. Thrombus formation and excessive fibrin deposition were not observed. The pathological diagnosis was established as IPEH.

DISCUSSION

IPEH is characterized by the development of endothelial papillary projection into the vascular lumen. IPEH arises most often in middle-aged adults, with a slight predilection for females. Although it may occur in any location in the body, only a few cases of IPEH on lower leg have been reported (2–4). Three different types of IPEH have been recognized: (i) a primary (pure) form, in which changes are observed in a dilated vessel or organizing thrombi; (ii) a secondary (mixed) form with incidental microscopic findings or pre-existing vascular tumour; (iii) an uncommon type that is extravascular in origin (3). The pure form of IPEH is reported to comprise approximately 33% of all IPEHs (3). Our case was also a pure form of IPEH that occurred in a dilated vessel, but organizing thrombi were not observed.

The exact pathogenesis of IPEH remains unknown, but an unusual form of thrombus organization following a trauma is considered to play a role (3). Levere et al. (5) demonstrated elevated levels of basic fibroblast growth factor (bFGF) in cases of IPEH compared with

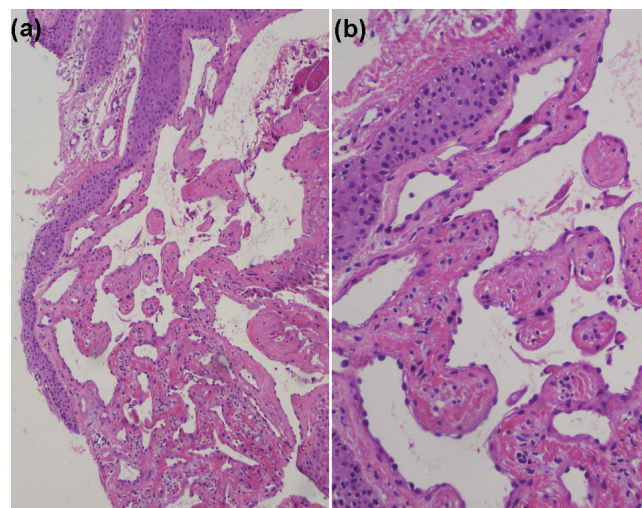


Fig. 2. (a) The mass of vascular channels with prominent intraluminal papillary projections in the dermis. (b) The papillary projections were composed of a single layer of plump endothelial cells around a core of fibrous connective tissue (haematoxylin and eosin staining: (a) $\times 100$; (b) $\times 200$).

non-IPEH organizing thrombi. They proposed that the release of bFGF from macrophages recruited to the lesion is proposed to trigger the proliferation of endothelial cells, which in turn release more bFGF, leading to a vicious cycle (5). Hormonal influence also has been suggested due to its slight female preponderance (6).

The differential diagnosis of IPEH includes other benign and malignant vascular proliferative disorders, such as pyogenic granuloma, Kaposi's sarcoma, haemangioma, and angiosarcoma (7). However, the main concern in forming clinicopathological diagnosis is differentiation of IPEH from angiosarcoma. A helpful differential point is its intravascular location, since angiosarcomas are almost never confined to a vascular lumen. It also differs from angiosarcoma in that the endothelial cells lack necrosis, marked pleomorphism, significant mitotic activity, and solid sheet formation (8). Clinically, it is almost impossible to make an exact diagnosis of IPEH just by inspecting the lesion. The usual clinical appearance of IPEH is an elevated, firm and sometimes tender nodule that imparts a slight purplish red colour to overlying skin or mucous membrane. We report here an IPEH on lower leg of female with unusual clinical appearance. As far as we know, there have been no reports of IPEH presenting as a strongly-black coloured firm nodule, which is very similar to malignant melanoma. However, malignant melanoma can easily be differentiated by pathological examination.

It is interesting that IPEH, which is a completely benign condition, may resemble two of the most notoriously malignant skin cancers, malignant melanoma and angiosarcoma, in clinical appearance and histological features, respectively.

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