We describe a case of recurrent intravascular papillary endothelial hyperplasia involving the middle finger which was successfully-treated with photon and proton radiotherapy following two previous surgical excisions.

Intravascular papillary endothelial hyperplasia is a rare benign neoplasm of endothelial origin which was first described by Masson in 1923\(^1\) as “vegetant intravascular hemangioendothelioma”. It comprises approximately 2% of all vascular tumours of the skin and soft tissue. The prognosis is usually excellent following resection.\(^2\) Few cases of recurrence, which is most often associated with an underlying vascular tumour,\(^3\)-\(^6\) have been reported. The role of radiotherapy in its treatment is unclear. We present a case involving a finger managed by such treatment following two previous excisions.

**Case report**

A 15-year-old girl presented with a painful mass in the middle finger of her dominant hand. Her medical history was unremarkable, and she had no history of trauma to the finger. Physical examination revealed a reddish-blue soft-tissue mass 2 cm in diameter on the volar aspect of the proximal phalanx of the right middle finger (Fig. 1). The finger was normal in terms of the range of movement, vascular examination, and neurological assessment.

Magnetic resonance imaging revealed a heterogeneous mass with no flow voids or calcification (Fig. 2). An excision biopsy was performed, and the histological examination showed a haemangioma with organising thrombus and papillary endothelial hyperplasia.

During the following four weeks, the patient developed a painful red mass, increasing in size, which ulcerated the skin and bled. There was a black eschar, 1 cm in diameter, at the site of operation. The finger was red, warm and tender with a bluish discoloration surrounding the eschar. Magnetic resonance imaging revealed a heterogeneous mass on T1- and T2-weighted images, consistent with recurrent tumour.

Repeat excision was performed through the pre-existing scar, which was extended...
proximally and distally to obtain adequate exposure. The eschar was excised in an elliptical fashion, and the residual tumour was seen just under the skin over the flexor tendon sheath. It was extensive and adherent to the surrounding structures with the exception of the radial neurovascular bundle. Frozen section revealed a highly cellular neoplasm, suggestive of a high-grade sarcoma. The ulnar neurovascular bundle was encased in the mass, and the ulnar digital nerve was sacrificed with excision of the tumour. An intraoperative digital Allen test\(^7\) to assess collateral circulation revealed the ulnar vessel to be predominant, and therefore it was preserved. The mass was isolated and excised completely. The wound was left open to heal by secondary intention. The final pathological diagnosis was of a recurring papillary endothelial hyperplasia and the surgical margins contained residual tumour. This was expected because the tumour had enveloped the ulnar vessel, which had been preserved.

Three weeks later, she presented with an ulcerating mass consistent with further recurrence (Fig. 3). Flexion of the finger was limited. Following consultation with a radiation oncologist (TFD), she started treatment with 19.8 Gy of external beam radiation therapy using photons, delivered in daily fractions of 1.8 Gy for a period of three weeks, following which the pain and swelling were reduced. In order to minimise the cumulative dose of radiation to the extensor aspect of the finger, she was then given two weeks off therapy before continuing treatment with protons for 21.6 Cobalt Gray Equivalent (CGE), in daily fractions of 1.8 CGE. Therefore, her entire treatment totalled 41.4 Gy over 58 elapsed treatment days. She tolerated the treatment well. Magnetic resonance imaging performed three weeks after the end of treatment showed marked reduction in the size of the mass.

She recovered normal use of her hand four months after completing treatment. She had full flexion of the finger, although 5° to 10° loss of terminal extension at the proximal interphalangeal joint remained. There was no swelling. Magnetic resonance imaging showed thin subcutaneous tissue consistent with post-operative changes, and no evidence of recurrence of the tumour. The appearances on MR scans at nine months were similar and at one year there was no clinical evidence of recurrence (Fig. 4).

**Discussion**

Intravascular papillary endothelial hyperplasia is a rare benign lesion showing reactive proliferation of the endothelium associated with thrombosis. It typically presents in one of three forms: 1) in an isolated, thrombosed blood vessel; 2) in association with a pre-existing pyogenic granuloma, haemangioma, or other vascular malformation; 3) from a haematoma. Its aetiology remains uncertain. Masson\(^1\) regarded it as a true neoplasm with endothelial proliferation followed by necrosis and thrombosis. However, recent evidence suggests that it is an unusual organising thrombus with reactive proliferation of endothelial cells in response to inflammation and stasis.\(^2,3\)

It typically presents as a firm, occasionally tender mass of ≤ 2 cm in diameter, often with red or blue discoloration of the skin. It is usually seen in the third or fourth decade, with a slightly greater incidence in women.\(^3\) The most commonly affected sites are the head, neck and fingers.\(^3\) The differential diagnosis includes pyogenic granuloma, haemangioma, and angiosarcoma. It is important to distinguish this benign...
lesion from an angiosarcoma. Intravascular papillary endothelial hyperplasia typically has an excellent prognosis following simple excision. Recurrence is extremely rare, and the few reported cases have usually been associated with incomplete resection or an underlying vascular lesion, such as an intramuscular haemangioma or pyogenic granuloma.

To our knowledge, this is the first reported case of the use of radiation therapy to treat recurrent intravascular papillary endothelial hyperplasia of the hand. The role of radiation therapy in the treatment of this condition is unclear. Treatment by radiotherapy of intracranial intravascular papillary endothelial hyperplasia has been described, but there is little evidence of its efficacy. We used a photon and proton radiation treatment in the range of 36 Gy to 45 Gy, typically used to treat haemangiomas. This dose was in the same range as that employed for the intracranial cases referenced above.

Protons are biologically similar to photons (both are low linear energy transfer radiation), but their distal range is related to their energy, and the depth of treatment can be controlled with a very high degree of precision. In our case, photon or electron irradiation for the course of the treatment would have entailed a full dose of circumferential irradiation to the finger, with a high risk of late radiation-related oedema. In order to minimise the damage to normal tissue on the extensor surface of the finger, the patient was changed to a proton-beam radiotherapy treatment plan. We were able to use a single volar port to adequately encompass the lesion for the last 21.6 CGE of treatment, and constructed a custom-built immobilisation and shielding mould to further spare the extensor portion of the finger from additional radiation. While protons could have been used theoretically for the entire course of the treatment, the extent of involvement at presentation allowed photon irradiation of the entire circumference of the finger at the level of the tumour to adequately encompass the lesion. After 19.8 Gy, there was sufficient regression to allow sparing of the extensor surface if the volar beam could be stopped beyond the deepest point of tumour involvement. Protons have no exit dose beyond their maximum range. In this case, the dose could be set to spare the extensor space of the patient’s finger. While most sarcomas that have been treated with protons have involved the spine and the base of the skull, they have also been used for soft-tissue sarcomas where photon techniques would have caused circumferential irradiation of the extremity, or for truncal lesions to spare underlying normal abdominal and thoracic tissues.

In our patient, a third excision was not considered a satisfactory option as the lesion encased the neurovascular bundle on the ulnar side. An intra-operative digital Allen test revealed the ulnar vessel to be predominant; resection would have compromised the viability of the finger. Therefore, in patients where complete re-excision of intravascular papillary endothelial hyperplasia is not possible or if there is a high risk of peri-operative complication, radiation therapy may be a viable alternative. Discussion with the patient regarding the possibility of future amputation in the event of a further recurrence is clearly mandatory.

Although the prognosis for intravascular papillary endothelial hyperplasia in the hand is excellent, rare cases of recurrence following excision have been reported. Based upon our experience in this patient, although the period of follow-up is short, radiation therapy may be successful in preventing recurrence and restoring function. No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References