Spindle cell hemangioma reoccurrence in the hand: case report

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Introduction

Spindle cell hemangioma (SCH) was first described by Weiss and Enzinger in 1986 [44]. They presented the initial 26 cases and at the time concluded that the vascular lesion was a vascular tumor of low-grade malignancy, and therefore, they termed the lesion a spindle cell hemangioendothelioma. Since then, various investigations have been conducted, including immunohistochemical studies, revealing they are benign lesions [4, 13, 14].

These lesions affect both sexes equally and have a predilection for the extremities. There are about 186 cases of SCH reported in the English literature, and 40 of the cases involved the hand [1–11, 13–26, 28–30, 32–47]. SCH can present as a solitary lesion (spindle cell hemangioma) or multifocal lesions (spindle cell hemangiomatosis) [33]; both types have been found in the hand. Lesions tend to develop during childhood or early adulthood. They most often have

an indolent course [9, 13], but can become painful and disfiguring [8, 17]. Most lesions occur in the dermis and subcutaneous tissues, but have been found in the viscera, spinal cord, cervix, and oral cavity [26, 36, 40, 41, 43].

Spindle cell hemangiomas have features of both cavernous hemangiomas and Kaposi's sarcoma. Microscopically, this vascular lesion consists of vascular proliferations lined by a thin layer of endothelium within solid areas of mostly spindle cells [6]. There is minimal or no mitosis. The percentage of cavernous areas versus solid areas varies greatly [10, 14]. Thrombi or phleboliths can be found within these cavernous spaces [14, 40] and can be evident in radiographs [8].

The standard of care is wide local excision, but caution must be taken when there is multifocal disease not to cause further disability. Additionally, the reoccurrence rate is greater than 50% [33]. We present a case of a female patient who had a local reoccurrence of SCH in her hand 14 years after the original lesion was excised.

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Case Report

A 30-year-old female was referred to us by Vascular Surgery for a “bulging vein” in her non-dominant hand. The lesion had been slowly growing since the original excision of the lesion in 1993. She noticed an accelerated rate of growth after her child was born in 2010. Over the last year, the lesion became painful and was causing her grip strength to diminish, often dropping objects she was holding. On exam, two bluish masses were appreciated. One lesion measuring 1.5 cm × 1.2 cm was on the radial side of the left third digit, which had a well-healed overlying 1.2-cm scar (Fig. 1). A second lesion measuring 1.3 cm × 1 cm was evident in the third web space encroaching on the dorsum of the hand.

Fig. 1 Preoperative views of the lesions involving the volar left third digit and the third web space on the dorsum of the hand

Both masses were slightly mobile, compressible, and tender to palpation. The only deficit found was decreased two-point discrimination in the distribution of the third radial digital nerve. She did not have any ulceration of the overlying skin. The patient could not recall how long the decreased sensation had been present. No other masses were identified. She had asthma and was otherwise healthy.

Magnetic resonance imaging was performed on the hand to further characterize and define the extent of the mass. T2 fast spin echo-weighted MR image with chemical fat saturation showed a hyperintense, irregular mass in the volar aspect of the third proximal phalanx. The lesion demonstrated a clear cortical break of the phalanx with extension into the bone marrow (Figs. 2a, 3, and 4). It was evident that the same mass also extended into the third web space (Fig. 2b). T1 fast spin echo MRI image shows that the lesion was isointense in signal compared to the muscle (Fig. 3) and additionally showed heterogeneous enhancement of the

mass (Fig. 5). The rest of the bones within the hand were unremarkable.

An incisional biopsy revealed a vascular malformation. We proceeded with full excision of the mass. Blunt dissection was used to separate the mass from the surrounding structures. It was very adherent to the overlying skin. The mass appeared like a sac of grapes and was not well demarcated (Fig. 6). Much care was taken to dissect the mass off of the third radial neurovascular bundle. It appeared to have several small feeding vessels, which were cauterized or clipped. Near the third web space, the mass coursed under the sagittal band of the third digit and continued deeper, penetrating the proximal phalanx. Cautery was used to amputate the mass from the bone.

Post-operatively, the patient healed well from her incision, and it is currently 6 months from the excision of the mass without signs of further reoccurrence. She reports to have full function of her hand and is pain free.

Fig. 2 Axial T2 fast spin echo weighted image with chemical fat saturation shows a hyperintense, irregular mass in the volar aspect of the third proximal phalanx (*asterisk*); the lesion demonstrates a cortical

break on the phalanx (*arrow head*) and extends into the bone marrow (*arrow*) (a). A lower slice (b) shows how the mass extends into the third web space (*arrow*)

Fig. 3 Axial T1 fast spin echo shows that the lesion is isointense in signal compared to the muscle (*arrow*)

Histological examination revealed spindle endothelial cell proliferation with adjacent slit-like spaces with extravasated red blood cells (Figs. 7 and 8). Additionally, medium-size blood vessels showed evidence of intravascular thrombosis and were surrounded by cavernous-like blood spaces (Fig. 9).

Discussion

Spindle cell hemangiomas have a predilection for the extremities. We found a 21.5% incidence in the hand upon review of the literature. These vascular lesions most often present as solitary or multiple dermal or subcutaneous masses. SCH has been reported to occur in viscera such as the pancreas and spleen [11, 36], as well as the spinal cord [1, 26]. Osseous involvement has only been reported in three other patients [5, 34, 37], and two of these involved

Fig. 4 Coronal T2 fast spin echo weighted image with chemical fat saturation shows the intermedullary extension of the lesion seen as hyperintense signal in the bone marrow (*arrow*) and soft tissue mass (*asterisk*)

Fig. 5 Coronal T1 fast spin echo fat saturated image with contrast shows heterogeneous enhancement of the mass (*arrow*)

the hand. In these two cases, the SCH appeared to cause only osseous erosions. The SCH in our patient not only extended deep into the subcutaneous tissue and traversed the extensor compartment, but also penetrated the proximal phalanx and entered the medullary canal. No other reports in the literature show this type of intramedullary bony involvement. Steinbach et al. [37] report on a case that has MRI imaging of an SCH in the thumb causing bony erosion. Chung et al. [5] present an SCH in the ankle that eroded into the medial malleolus. This case is histologically different from other cases of SCH because there was pleomorphism and increased mitotic activity. In Scott and Rosai's [34] case series of seven patients with SCH, they report on a patient with spindle cell hemangiomatosis of the entire upper extremity. Bony erosions were found in the areas where the lesions were contiguous.

The etiology of SCH has yet to be fully elucidated. Fletcher et al. [13] advocate that the lesions arise in the area

Fig. 6 Intraoperative view of mass protruding through the third web space

Fig. 7 High power photomicrograph of the lesion showing the spindle endothelial cell proliferation with adjacent slit-like spaces

Fig. 9 Low power photomicrograph showing medium-size blood vessels with evidence of intravascular thrombosis and surrounded by cavernous-like blood spaces

of abnormal vasculature. Imayama et al. [20] propose that injury incites these lesions, causing a reactive vascular proliferation. It can occur at any age, but has a propensity for patients before and during the second decade of life [13, 33].

Despite the initial appearance that our patient had two separate lesions, on MRI and surgical exploration, it was clear that it was one continuous lesion that traversed the extensor compartment. Of the 34 cases in the literature involving the hand, the vast majority present as multiple lesions [9, 13, 20, 34]. Multifocal SCH's have been associated with Maffucci's syndrome [13, 17, 32, 46], Ollier disease [27], lymphadema [42, 47], Klippel Trenaunay syndrome, and early onset varicose veins [13]. Of these, Maffucci's syndrome is the most common, and many advocate that a patient with multifocal SCH should be evaluated for enchondromas. Perkins and Weiss [33] advocate that surveillance in patients with Maffucci's syndrome is particularly important because they can develop malignant degeneration of the enchondromas and angiosarcomas.

The differential diagnosis of SCH includes other types of hemangiomas (including cavernous, histiocytoid, and disseminated lobular capillary hemangiomas), dermatofibroma,

Fig. 8 Spindle endothelial cell proliferation forming irregular vascular spaces and kaposi slit-like spaces with extravasated red blood cells

Kaposi's sarcoma, epithelioid angiomatosis, kaposiform hemangioendothelioma, and intravascular papillary endothelial hyperplasia [34, 37, 40, 46]. The combination of SCH's cavernous spaces with organized thrombi, plump endothelial cells, multitude of spindle cells, and minimal mitotic activity can histologically differentiate it from the other diagnoses [46].

Preoperative planning should include an MRI because, as evident in our case, deeper structures can be involved [21, 37]. This can also aid in diagnosing enchondromas in patients with multifocal disease. Fletcher et al. [13] suggest angiographic examination owing to the associated histological evidence of locally malformed vasculature.

Although SCH is now believed to be a benign lesion [12, 13, 20], it can range from a solitary lesion that is excised and does not reoccur to multiple lesions that can progress to disfigure an entire extremity [8, 34]. Additionally, the lesions can be present for decades before growing large enough to be noticed or to become painful, but they can also reoccur after surgery or increase in number within months [9, 38]. The standard treatment is wide surgical excision [31, 44]. Limited excisions almost certainly reoccur, but there is no consensus on the incidence of recurrence [38, 44]. When deciding how to treat each individual case, it must be taken into consideration the slow growth of the tumor and the morbidity that the surgery may cause. In our rare case, the tumor enters the medullary canal, so complete wide local excision would have included a third digit amputation. We discussed with the patient the possibility of amputation due to the high likelihood of recurrence, but decided to do a more conservative excision with close follow-up. In other cases where there are multiple lesions, excision may be even more disfiguring and morbid. Radiotherapy, low-dose interferon α -2b, and intralesional and intraarterial administration of recombinant interleukin-2 have been successful in treating aggressive disease or lesions that are difficult to surgically excise [7, 26, 36, 40,

44]. There has been no reported patient mortality or metastasis from SCH in the literature. Spontaneous regression has been reported [13]. In light of the data, it is important to counsel the patient with SCH on the high risk of reoccurrence and provide appropriate follow-up.

Conflicts of interest Authors declare that they have no conflicts of interest.

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