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## Dermatology Online Journal

### Title

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### Permalink

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### Journal

Dermatology Online Journal, 25(9)

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### Publication Date

2019

### DOI

10.5070/D3259045513

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Peer reviewed

# Tufted angioma presenting with subclinical coagulopathy

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## Abstract

Tufted angioma is a rare, benign vascular tumor of uncertain pathogenesis, characterized histopathologically by “tufts” of capillaries within the dermis. A life-threatening coagulopathy, Kasabach-Merritt phenomenon, occurs in approximately 10% of cases of tufted angioma and is characterized by profound thrombocytopenia and fibrinogen consumption. We present an asymptomatic 10-month-old boy who presented with an erythematous patch of the right upper extremity and subsequently was diagnosed by biopsy with tufted angioma. Baseline laboratory workup of our patient revealed mildly decreased fibrinogen and elevated D-dimer levels without thrombocytopenia or elevated partial thromboplastin time. This suggests that asymptomatic patients with tufted angioma may present with coagulopathy in the absence of Kasabach-Merritt phenomenon. This also highlights the importance of obtaining baseline laboratory workup in patients presenting with tufted angioma.

*Keywords: Kasabach Merritt, tufted, angioma, vascular*

## Introduction

Tufted angioma is a rare, benign vascular tumor of uncertain pathogenesis, characterized histopathologically by “tufts” of capillaries within the dermis [1]. These tumors typically occur during infancy or early childhood, with approximately half present at birth and the majority of the rest presenting within the first year of life.

## Case Synopsis

A 10-month-old boy presented for evaluation of an erythematous patch of his medial right upper extremity. The patch appeared rapidly at approximately 5 months of age and had not changed significantly since appearance. The patient’s parents reported no symptoms. Prior to referral to the dermatology clinic, the patient’s pediatrician had treated him with topical mupirocin and topical hydrocortisone 2.5% cream without improvement.

On presentation, the patient had an annular and reticulated erythematous patch of the right medial upper extremity without surface change (**Figure 1**).



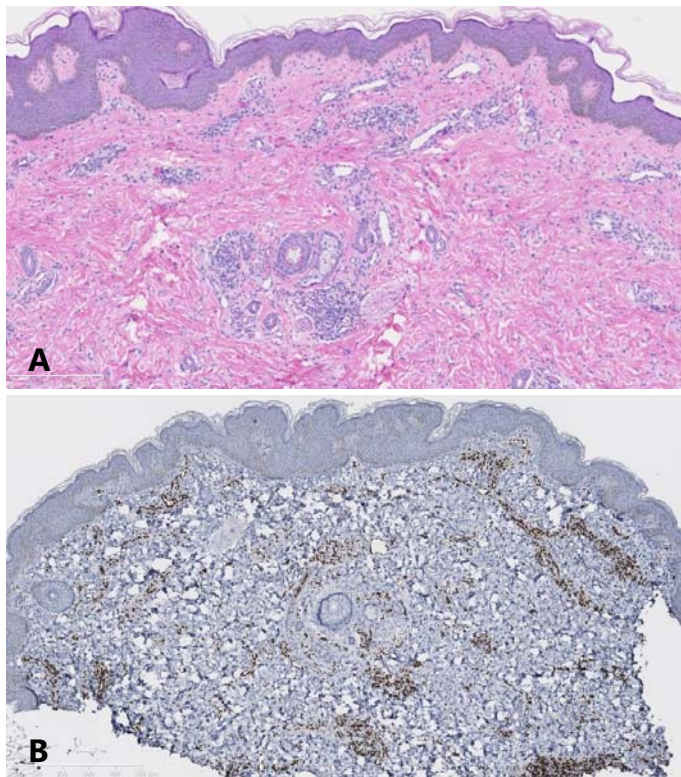
**Figure 1.** Reticulated erythematous patch of the right upper extremity.

A 3mm punch biopsy was performed. Pathology showed a vascular proliferation in the superficial dermis with spindled and polygonal cells (**Figure 2**). The vascular nature of the lesion was confirmed with positive CD31 and ERG immunohistochemical stains. A diagnosis of tufted angioma was made.

Laboratory workup for our patient included a mildly elevated partial thromboplastin time at 32 seconds (reference range (RR): <30 seconds), mildly decreased fibrinogen 141mg/dL (RR: 180-363mg/dL), and elevated D-dimer at 1960ng/mL (RR: 190-500ng/mL). Complete blood count, including platelets, comprehensive metabolic panel, and prothrombin time were within normal limits.

## Case Discussion

Tufted angiomas are rare and the precise incidence is unknown. It is closely associated with kaposiform hemangioendothelioma, a more aggressive tumor,



**Figure 2. A)** Punch biopsy of right medial arm demonstrating vascular proliferation in the superficial dermis mimicking "tufts." H&E, 100 $\times$ . **B)** ERG immunohistochemical staining demonstrating vascular nature of the lesion, 60 $\times$ .

with the processes believed to be part of a continuum [2].

A life-threatening coagulopathy, Kasabach-Merritt phenomenon occurs in approximately 10% of cases of tufted angioma and 70% of cases of kaposiform hemangioendothelioma [3]. In these cases prominent thrombocytopenia and consumption of fibrinogen and coagulation factors can be seen, with tumors becoming tense, painful, and purpuric. This phenomenon is believed to be result of platelet trapping within the tumor vasculature. Infantile hemangiomas, the most common vascular tumors of infancy, are not associated with Kasabach-Merritt phenomenon [4].

Surgical excision is the definitive treatment for small or localized tufted angiomas [5]. Observation is an option in asymptomatic cases and there have been cases of spontaneous regression [6]. For symptomatic, nonresectable cases low-dose aspirin may have a benefit [4, 7]. Although our patient has mild laboratory abnormalities, given the lesion is asymptomatic and stable, we elected for close monitoring.

Previous cases of patients with tufted angioma presenting with chronic coagulopathy in the absence of thrombocytopenia have been reported [6]. This suggests that coagulopathy may be more prevalent among patients with tufted angioma than the rates of Kasabach-Merritt phenomenon would suggest. These findings also suggest that coagulopathies may present in these patients to varying degrees and supports the need to obtain baseline laboratory workup in patients presenting with tufted angioma or kaposiform hemangioendothelioma.

## Conclusion

This is a case of tufted angioma in an asymptomatic patient who was found to have a mild coagulopathy on laboratory workup.

## Potential conflicts of interest

The authors declare no conflicts of interest.

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