A 2-year-old male child presented to us with 1-year history of single, asymptomatic, reddish to bluish swelling with hypertrichosis over upper back. The lesion began as a pinpoint reddish mark. There was no history of trauma before onset, increased sweating over the lesion, bleeding or discharge from the lesion, or bleeding from any other site. There was no family history or any significant past medical or surgical history.

On examination, a single, ill-defined, 4 cm × 6 cm sized, erythematous to violaceous plaque was present over upper back of the patient. Multiple lightly pigmented vellus hair was present over the surface of the lesion [Figure 1]. On palpation, lesion was nontender, indurated in the center, adherent to overlying skin and free from the underlying soft tissue structure. Lesion was blanchable on diascopy. No bruit was heard on auscultation.

Histopathological examination showed blood vessels with tightly packed rounded or spindle-shaped endothelial cells organized as clusters in the dermis [Figure 2]. Immunohistochemistry was not performed due to financial constraints of the patient.

**Question**

What is your diagnosis?

**Figure 1:** An ill-defined, erythematous to violaceous plaque present over upper back with multiple lightly pigmented vellus hair over the surface

**Figure 2:** Blood vessels with tightly packed rounded or spindle-shaped endothelial cells organized as clusters in the dermis (H and E, ×10)
Answer
Tufted angioma.

Discussion
Tufted angioma (TA) is a rare benign vascular tumor with endothelial origin characterized by slow and indolent growth. This lesion was first described by Nakagawa in 1949, who named it as angioblastoma and later, Macmillan and Champion renamed it as a progressive capillary hemangioma. In 1989, Jones and Orkin defined the term TA based on the microscopic features of this pathology as the endothelial cell proliferation presents a typically tufted organization.

This is a rare tumor which occurs most commonly in prepubertal children. It was, however, originally described in adults. Less commonly, it has been described in pregnancy and in an immunosuppressed individual. No causes of TA have been established. Trauma does not appear to be a predisposing factor, although a report describes the appearance of a lesion of TA at the site of a previous arthropod bite. Some authors have postulated that high hormonal levels during pregnancy and puberty may induce the development of TAs.

TA appears as a purplish red to red-brown patch or plaque that predominantly appears on the upper thorax, neck, shoulders, and less commonly, on the face, scalp, and proximal extremities. The diameter of the patch or plaque generally ranges from <1 cm to several centimeters. It may be solitary or multifocal and it can be round to polycyclic in more extensive cases. Lesions may be tender and show hyperhidrosis and hypertrichosis. The presence of hair aids in diagnosis.

Histologically, there is a lobular proliferation of plump, oval cells surrounding tiny slit-like lumina. These blood vessels are tightly packed and organized in rounded tufts scattered in the dermis, often described as resembling cannonballs. The tufts may occur deeply in the dermis, and into the subcutis. Immunohistochemical stains show strong positivity for *Ulex europaeus* I lectin and EN4 and unlike infantile hemangioma, negative staining for glucose transporter 1. Their natural history shows that after a period of expansion they stabilize and may either resolve spontaneously or more rarely progressively worsen. Malignant transformation has not been reported.

As with kaposiform hemangioendothelioma, it may be associated with the Kasabach–Merritt phenomenon. In our case, there was no rapid increase in size of the lesion and no sign of overlying inflammation. Platelet count, fibrinogen level, prothrombin time, and partial thromboplastin time were found to be within normal limits in the patient.

Based on the histologic features, it is important to distinguish TA from other vascular lesions such as pyogenic granuloma, hemangioma, and kaposiform hemangioendothelioma.

Pyogenic granuloma shows a high capillary vascular proliferation and presence of intense inflammatory infiltrate containing young fibroblasts that resemble granulation tissue, while TA is characterized by proliferation of spindled and polygonal cells surrounding vascular channels, organized as tufts. Capillary hemangioma presents as the proliferation of endothelial cells forming lobular arrangement of well-formed capillaries; however, the clusters of endothelial cells in TA are larger and more irregular in shape. Moreover, the “cannon ball” distribution of nodules is only observed in TA. The kaposiform hemangioendothelioma exhibits spindled endothelial cells with slit-like vessels, and these cells grow in sheets or coalescing nodules rather than dispersed tufts as observed in TA.

Treatment of choice is surgical excision, although systemic steroids, interferons, propranolol, pulse dye laser, superficial X-ray therapy, cryotherapy, and chemotherapy have been tried. Propranolol was given to our patient in a dose of 2 mg/kg after prior investigations. However, the patient was lost to follow-up.

Learning points
1. TA is a rare benign vascular tumor with endothelial origin
2. It is not present at birth, as also observed in our patient. Hence, helps in distinguishing it from hemangioma
3. In the absence of immunohistochemical staining, diagnosis can be made on the basis of histopathological examination which shows a characteristic cannon-ball appearance
4. Hypertrichosis aids in diagnosis
5. It has a slow and indolent growth. Any rapid increase in size or inflammation is usually a sign of Kasabach–Merritt phenomenon and should be appropriately investigated and managed.

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Conflicts of interest
There are no conflicts of interest.

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