Case Report
Olgu Sunumu

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Tufted angioma masquerading as a granulomatous condition: A rare case report

Granülomatöz bir hastalığı taklit eden tufted anjiyom: Nadir bir olgu sunumu

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Abstract

Tufted angioma (TA) is an uncommon benign vascular tumor that manifests as erythematous macules, plaques, and nodules and progresses slowly. It is histopathologically characterized by tufts of endothelial cells in a cannonball pattern in the dermis. It primarily affects children and adolescents. A 12-year-old child presented to us with a single, painful, sizeable annular plaque on the right side of his neck that had been there since he was 5 years old. Lupus vulgaris, borderline tuberculoid Hansen, annular elastolytic giant cell granuloma, acquired hemangioma, and benign vascular tumors were all evaluated as differential diagnoses. Ultrasound of the neck revealed an ill-defined hypoechoic region on the right side of the neck with no extension into the deeper plane. TA was diagnosed based on histopathology. We report this case because of its uncommon clinical form of TA, which presented as a single large erythematous annular plaque resembling a granulomatous lesion, which was later confirmed by histological features.
Keywords: Angioma, histopathology, tufted angioma, vascular tumors

Öz

Tufted anjiyom (TA), eritematöz maküller, plaklar ve nodüller şeklinde ortaya çıkan ve yavaş ilerleyen nadir bir benign vasküler tümördür. Histopatolojik olarak, dermiste cannonball paterni oluşturan endotel hücre demetleri ile karakterizedir. Öncelikle çocukları ve ergenleri etkiler. On iki yaşındaki bir çocuk, 5 yaşından beri var olan, sağ boynunda tek, ağrılı ve büyük boyutlu halka şeklinde bir plak ile bize başvurdu. Lupus vulgaris, borderline tüberküloid Hansen, halka şeklinde elastolitik dev hücreli granülom, edinilmiş hemanjiyom ve benign vasküler tümörler ayırıcı tanılar olarak değerlendirildi. Boyun ultrasonunda, sağ boyunda derin plana uzanımı olmayan, belirsiz sınırlı hipoekoik bir bölge tespit edildi. TA, histopatolojik bulgulara dayanarak teşhis edildi. Bu olguyu, histolojik bulgularla doğrulanan ve granülomatöz bir lezyonu andıran tek, büyük, eritematöz halka şeklinde bir plak olarak ortaya çıkan nadir bir TA klinik formu olması nedeniyle sunuyoruz.
Anahtar Kelimeler: Anjiyom, histopatoloji, tufted anjiyom, vasküler tümörler

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Introduction

Tufted angioma (TA) is a rare benign vascular tumor characterized by tufts of endothelial cells in a cannonball pattern in the dermis, which is most common in children and adolescents, with no gender or ethnic preference[1]. It is a painful and tender tumor. The name TA was coined to describe the characteristic dense clumps and lobules of endothelial cells on capillaries visible on histology[2]. It appears as slowly spreading erythematous macules, plaques, and nodules that are generally found on the neck, upper trunk, and extremities[2]. Kasabach-Merritt syndrome (KMS), which is thought to be caused by platelet entrapment in lesions, is a rare complication that must be recognized early. Sometimes, TA can be readily misinterpreted, and histopathological examination (HPE) and immunohistochemistry are used to confirm the diagnosis. Super potent topical corticosteroids, systemic corticosteroids, pulsed dye laser, and surgical excision are among the therapy options. We report this case because of its uncommon clinical presentation, which presented as a single large erythematous annular plaque resembling a granulomatous lesion, which was later confirmed by histological features.

Case Report

A 12-year-old boy, accompanied by his mother, presented to the dermatology outpatient department with complaints of a solitary, painful, erythematous raised lesion on the right side of his neck (Figure 1) that had been present since he was five years old. There had been no history of trauma prior to the development of the lesion. The lesion grew progressively until it reached its current size. There was no fever, weight loss, or other constitutional symptoms. There was no family or personal history of tuberculosis. On dermatological examination, a single, well-defined, tender erythematous annular plaque measuring 8x2 cm was found on the right side of the neck, with elevated indurated borders and mild atrophy (Figure 1). There was no evidence of concomitant lymphadenopathy.

Lupus vulgaris, borderline tuberculoid Hansen, annular elastolytic giant cell granuloma, acquired hemangioma, and benign vascular tumors were all evaluated as differential diagnoses.

Complete blood count, erythrocyte sedimentation rate, coagulation profile, renal function test, and liver function test were all within normal limits. A chest X-ray showed clear lung fields with no signs of tuberculosis. An ultrasound of the neck was performed, which revealed an ill-defined hypoechoic region on the right side of the neck with no extension into the deeper plane. There was no indication of an underlying arteriovenous abnormality.

A 4 mm punch biopsy was obtained and sent for histopathological analysis. HPE revealed normal epidermis and dermis, showing many lobules of tufts of capillaries with a "cannonball" appearance in the deep dermis and extending focally into the subcutaneous tissue (Figures 2, 3). Endothelial cells lined the lobules of closely packed capillaries, which were surrounded by pericytes. There was no evidence of granuloma, vasculitis, cellular atypia, or atypical mitotic figures. As a result, the final diagnosis of TA was based on HPE findings. The patient was advised to undergo surgical excision and was referred to the surgery department for further treatment.

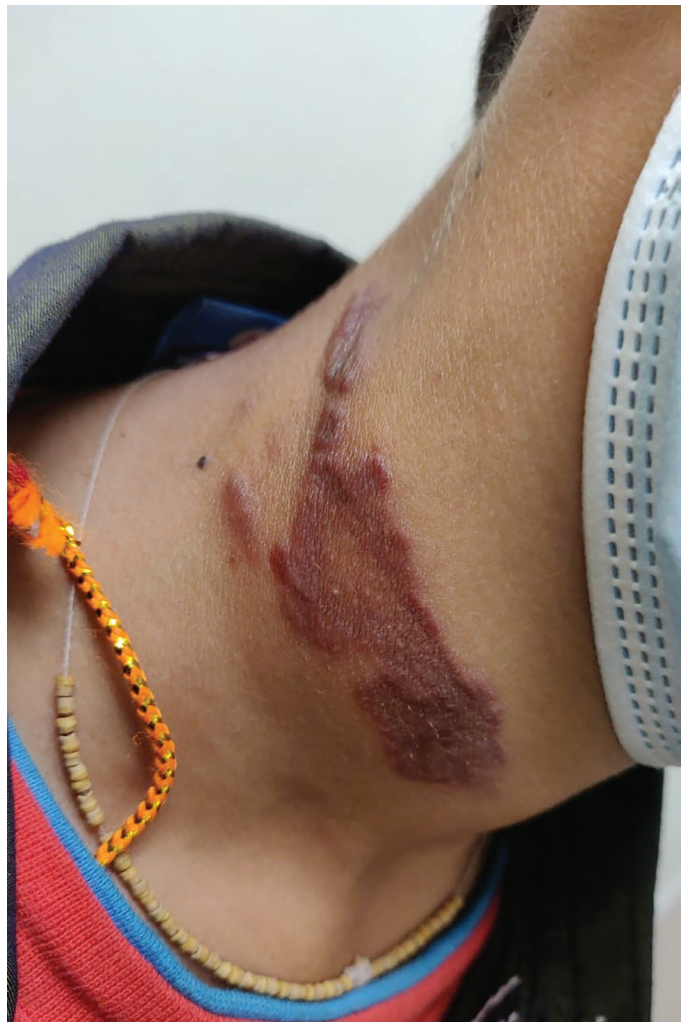

Figure 1. A solitary, well-defined erythematous annular plaque with raised indurated borders with mild atrophy over the right side of the neck

Discussion

Wilson Jones first gave the name TA to describe an unusual acquired vascular proliferation[2]. It is also known as Nagakawa angioblastoma and progressive capillary hemangioma[3]. Angiogenesis is facilitated by an increase in endothelial and vascular growth factors, which allows for the development of capillary lobes. TA is most common in infancy or early childhood[1]. According to many studies, 60-70% of TAs develop before the age of 5 years, and 25% of tumors appear before the age of one year[4]. In this case, the lesion appeared at the age of five. It can start as a small macule that resembles a port wine stain and then coalesce into plaques or nodules of varying sizes. The patch or plaque gradually grows for 5 months to 10 years, then stops growing[5]. The majority of TA cases exhibit little tendency for spontaneous regression. Some lesions resemble connective tissue disorders. It can be accompanied by overlying hypertrichosis and hyperhidrosis[6]. A previous case describing multifocal TA with annular morphology in a child similar to ours has been reported[7]. It can present in three clinical patterns: uncomplicated

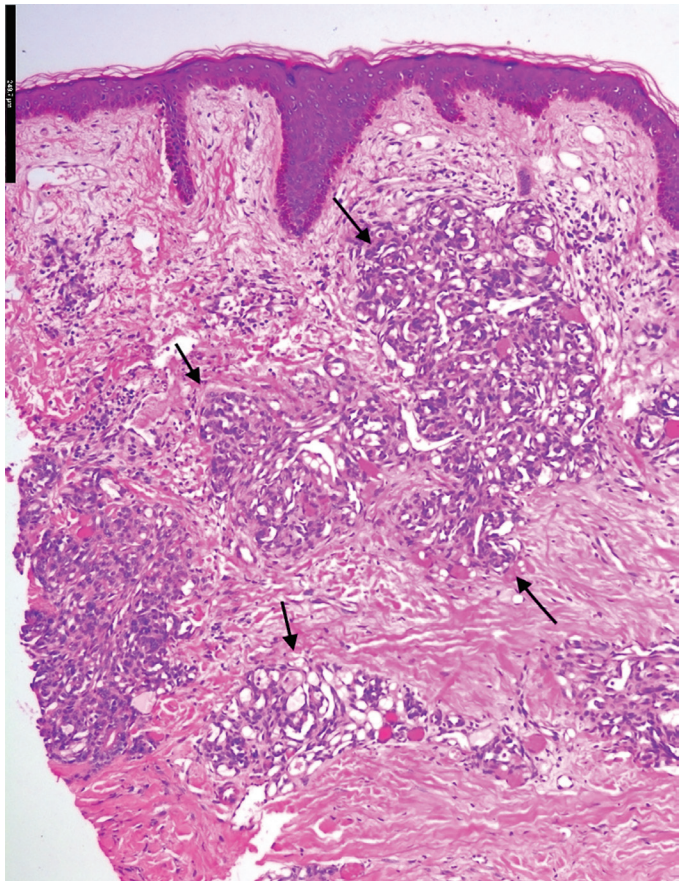

Figure 2. Photomicrograph showing multiple lobules of tufts of capillaries resembling a "cannonball" (black arrow) (H&E, x10). No granuloma or cellular atypia was observed
H&E: Hematoxylin and eosin

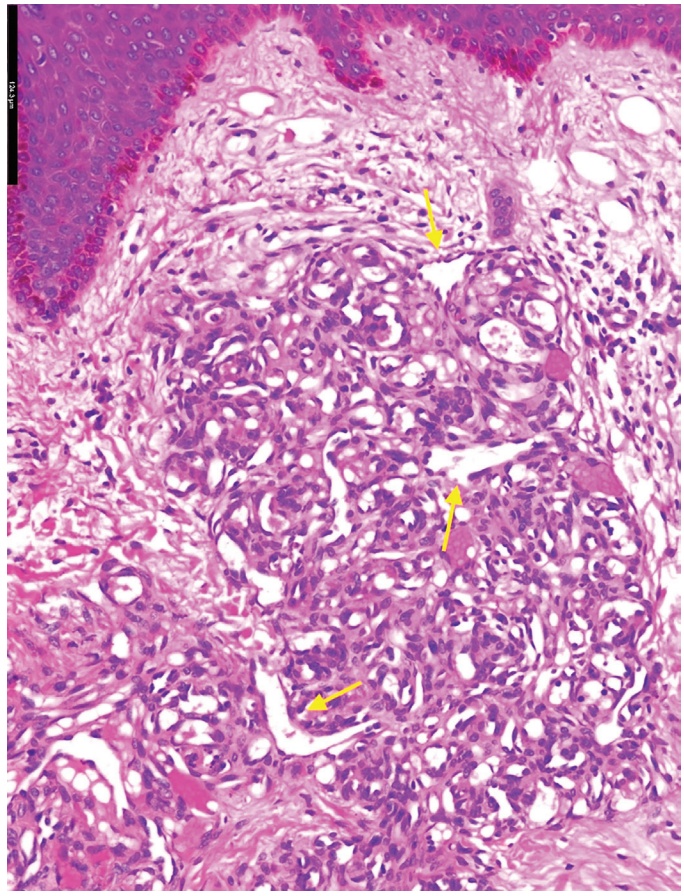

Figure 3. Photomicrograph showing dermis with a lobule composed of tufts of capillaries (yellow arrow) with endothelial cells surrounded by pericytes (H&E, x40)
H&E: Hematoxylin and eosin

thrombocytopenia, thrombocytopenia with chronic coagulopathy, and thrombocytopenia complicated by KMS[8]. KMS develops as a result of platelet sequestration and the activation of the fibrinolytic system. The presence of petechial and ecchymotic patches in TA should raise the possibility of KMS development[9].

Infantile hemangioma, pyogenic granuloma, vascular malformations, Kaposiform hemangioendothelioma (KHE), and Kaposi sarcoma are considered differential diagnoses for TA. Table 1 shows the characteristics that distinguish these conditions. KHE is an infiltrative rare vascular tumor that presents as a brown-red stain at birth, which thickens and becomes purpuric or as plaques, nodules, and bulky tumors. KHE may be present at birth or develop in early childhood. Adult cases have also been reported. It can occur on the extremities, trunk, head, neck, and deeper viscera can be affected. The histology of KHE is characterized by infiltrative nodules and sheets of spindled endothelial cells with minimal atypia and crescentic vessels containing hemosiderin. KMS is one of the complications associated with KHE. Spontaneous involution is rare[10].

To exclude other skin lesions from TA, histopathologic examination and immunohistochemistry are required. Because of the appearance of red, swelling, warm, and occasionally painful masses, TA can be misdiagnosed as a local infection and inflammation. The presence of dense agglomerates of endothelial cell lobules and capillaries in cannonball appearance, surrounded by the spindle and polygonal cells, on histopathology, is characteristic of TA. An ultrasound of the affected area can be performed to differentiate it from other tumors and to assess the plane and consistency of the lesion. Immunohistochemistry reveals positivity for CD31, CD34, and, in rare cases, smooth muscle actin[11].

Therapy for TA is limited and individualized due to the lack of specific treatment guidelines. Lesions can be monitored in a few cases due to the possibility of spontaneous regression[12]. Topical or systemic corticosteroids, pulse dye laser, cryotherapy, surgical excision, chemotherapy, and radiotherapy are among the treatment options[13]. Corticosteroids, vincristine, or interferon should be used aggressively to treat the Kasabach-Merritt phenomenon[14].

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Table 1. Differentiating features between tufted angioma, infantile haemangioma, vascular malformation, pyogenic granuloma and Kaposi sarcoma

	Tufted angioma	Infantile haemangioma	Vascular malformation	Pyogenic granuloma	Kaposi sarcoma
Age of onset	Present at birth/ childhood	Appears few weeks to months after birth	Present at birth	Childhood or pregnancy	Classic type - 5th to 7th decade; endemic type- children, adults iatrogenic type- younger adults
Gender	No predilection	Female preponderance	No predilection	Male preponderance	Male preponderance
Location	Neck, trunk and extremities	Face, neck	Neck, forehead, glabellar areas, lower legs	Face, fingers, gingival, labial or nasal mucosa	Feet, hands, ears, nose, lymphnodes, mucosae and viscera
Clinical presentation	Dusky red to violaceous macules, plaques or nodules	Asymptomatic, sharply demarcated, bright red, protuberant plaque/nodule with smooth /bosselated surface	Superficial, reddish or salmon-coloured irregular flat areas crossed by linear telangiectasia	Well circumscribed, red coloured dome-shaped, sessile or pedunculated nodules with collar of scales	Multifocal purplish macules, plaques and tumours
Dermoscopic findings	Skin coloured to light pink structureless area	Mixed vascular pattern: red dots, globules, lacunae, and structureless area	Red to reddish -purple globules, and lacunae	Red structureless area and shiny white structures	White lines, white clods, surface scale, polychromatic colour change, four dot clods, collarette sign, serpentine vessels
Histological findings	Vascular tufts of capillaries throughout dermis in cannonball distribution.	Multinodular pattern with nodules composed of hyperplastic endothelial cells, pericytes with or without lumens and prominent basement membranes	Ectasia of mature dermal blood vessels in cobblestone appearance	Typical lobules of capillaries in a myxoid background	Dense whorled arrangement of spindled cells in dermis with unlined slit like spaces with extravasated RBCs
Course	Spontaneous regression if onset is before six months	Rapid growth, quiescent phase and phase of regression	Increase in size as patients grows	No spontaneous regression Prone for recurrences	Systemic involvement- poor prognosis
Treatment	Topical or systemic corticosteriods, pulse dye laser, cryotherapy, surgical excision	Topical/systemic/ intralesional steroids, beta blockers, surgical excision, laser therapy	Flashlamp pulsed dye laser	Shave removal, curettage, electrocautery, cryotherapy and pulse dye laser	Localised cutaneous disease: excision & Cryotherapy for systemic involvement: radiotherapy, chemotherapy- liposomal doxorubicin and paclitaxel
Complications	Kasabach-Merritt syndrome	Ulceration, thrombosis, bleeding, infection	Facial and limb asymmetry, ulceration	Recurrent bleeding and ulceration	Edema, disfigurement of skin; pulmonary involvement leads to death

RBCs: Red blood cells

We present this case to emphasize the importance of including TA in the differential diagnosis of any lesion with annular morphology, especially if accompanied by pain.

Ethics

Informed Consent: The patient's informed consent have been obtained for the patient's clinical informations and images to be published in the journal.

Footnotes

Authorship Contributions

Surgical and Medical Practices: A.A.A., N.M., S.G.V., Y.M., Concept: A.A.A., N.M., S.G.V., Y.M., Design: A.A.A., N.M., S.G.V., Y.M., Data Collection or Processing: A.A.A., N.M., S.G.V., Y.M., Analysis or Interpretation: A.A.A., N.M., S.G.V., Y.M., Literature Search: A.A.A., N.M., S.G.V., Y.M., Writing: A.A.A., N.M., S.G.V., Y.M.

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