

Annular tufted angioma

Sir,

Tufted angioma (TA) also called angioblastoma of Nagakawa, is a very rare pattern of angioma. Congenital or adult-onset cases are rare.^[1] In 50% of the cases, it develops in infancy and it is congenital in only 15% of cases.^[2] It starts as a poorly delineated macule with mottled appearance. It is pinkish, erythematous or purplish in color resembling a port wine stain. Gradually it progresses to deep red to purple plaques or nodules of 2-10 cm size.^[3] Clinically they can have variable presentations mimicking pyogenic granuloma, hemangioma, vascular malformation or Kaposiform hemangioendothelioma (KHE).^[4] However, annular configuration of lesions has not been reported.

An eleven year-old male child presented with three progressively enlarging, annular, erythematous, tender, well-defined plaques over the upper trunk and shoulder without any deeper swelling for the last three years. There were also a few small papules at the periphery. There was no telangiectasia and the lesions were not compressible [Figure 1].

The patient also complained of localized hyperhidrosis. No other family members were affected. There was no lymphadenopathy. Systemic examination was normal.

Investigations including complete hemogram, liver and renal function assessment, chest X-ray and routine urine examinations were done and were within normal limits. Punch biopsy showed mild acanthosis and multiple lobules composed of solid staining cells and narrow vascular channels compressed by closely packed spindle shaped cells. There were multiple eccrine glands noted around the angiomatous

lobules [Figure 2].

Tufted angioma was described by Jones and Orkin in 1989.^[2] Clinically it presents as pinkish macules to purplish plaques with an ill-defined border.^[4]

Our case was atypical in presentation as it had a very well-defined lesion without any signs of vascular pathogenesis like a port wine stain, telangiectasia or any deeper tissue swelling. Moreover two of the three lesions were almost annular with marked central depression. They resembled granuloma annulare. However, there was no beading of the margins in any of the lesions. Age of onset and distribution of lesions in the present case is classical. Classical sites of involvement are the neck, the upper trunk and the shoulder.^[2,5,6] Males are more frequently affected than females.^[2] However, not all studies support this pattern.^[5]



Figure 1: Annular tufted angioma over the back

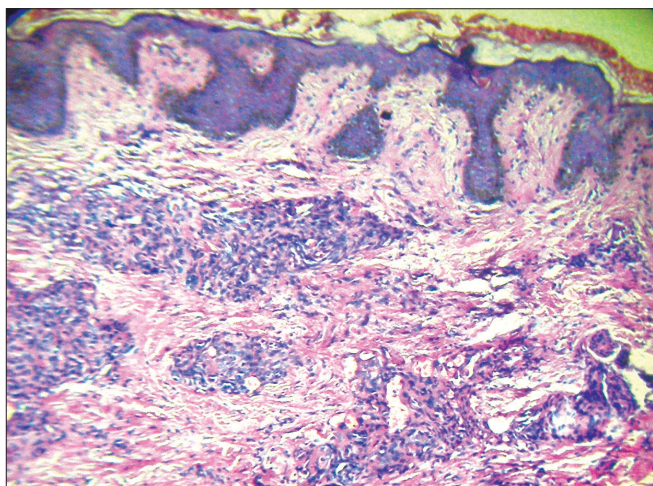


Figure 2: Histopathology showing classical 'cannon ball' appearance (H and E stain, X100)

Histologically, there are numerous vascular lobules in the dermis composed of solid stained perithelial cells. Multiple capillaries lined by small immature endothelial cells are scattered within these lobules. Frequently, the enlarged endothelial cells compress the lumen of the capillaries making it slit-like. The gross microscopic morphology of the lobules is termed as cannon ball appearance. It is said that the presence of eccrine glands nearby is a specific finding for this angioma, which is present in our case.

Classically TA is characterized by indolent growth^[7] although a faster rate of progression has also been described.^[2] However, malignant changes are never seen. The most dreaded complication of TA is the rare occurrence of Kasabach-Merritt syndrome. Small tumors can be excised. Other treatment options are often required like systemic corticosteroid, interferon gamma (IFN- γ), radiotherapy, cryotherapy, pulse dye laser etc.

Tufted angioma is a rare tumor. Most of the previous reports are from Japan. No case has been reported in Indian patients. This case of tufted angioma in an Indian male child is reported here due to its unusual clinical presentation.

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