A Case Report of Late-Onset and Pregnancy-Related Tufted Angioma in Thailand.

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ABSTRACT:

PONGCHOMPORN M, PATTAMADILOK B. A CASE REPORT OF LATE-ONSET AND PREGNANCY-RELATED TUFTED ANGIOMA IN THAILAND. THAI J DERMATOL 2016; 32: 199-204.

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Tufted angioma is a rare benign vascular tumor of the skin, usually occuring during infancy or childhood by either congenital or acquired causes. Adult onset has rarely been reported and pregnancy-related incidences are also infrequent. However, we reported a Thai woman with adult-onset, pregnancy-related tufted angioma. Such a case has seldom been seen in Thailand or even in world-wide medical literature. In our case, the patient presented with a slowly progressing lesion with tender, dull erythematous macules and papules which coalesced into patches and plaques with a mottled appearance on the right inframammary area. The lesion began when she was in the seventh month of pregnancy. She was later treated with vascular laser with excellent results.

Keyword : tufted angioma, Kasabach-Merritt syndrome, pregnancy

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บทคัดย่อ:

มณสิญา พงษ์ชมพร, เบ็ญจ์สชีว์ ปัทมดิลก รายงานผู้ป่วยที่มีความผิดปกติของหลอดเลือด ชนิด TUFTED ANGIOMA ที่เกิดในผู้ป่วยหญิงตั้งครรภ์ในประเทศไทย วารสารโรคผิวหนัง 2559; 32: 199-204.

สถาบันโรคผิวหนัง

ความผิดปกติของหลอดเลือดชนิด tufted angioma เป็นโรคที่พบได้น้อยมาก ส่วนมากมักพบในผู้ป่วยเด็ก อาจจะเกิดตั้งแต่แรกคลอดจนตลอดช่วงวัยเด็ก รายงานฉบับนี้นำเสนอผู้ป่วยที่มีความผิดปกติของหลอดเลือดชิด tufted angioma ในผู้หญิงตั้งครรภ์ ซึ่งพบได้น้อยมากในประเทศไทย

คำสำคัญ : ความผิดปกติของหลอดเลือด คนท้อง เนื้องอกเส้นเลือด หลอดเลือดผิดปกติ

Introduction

Tufted angioma is a rare slow-growing benign vascular neoplasm. It is also known as “Angioblastoma of Nakagawa” in Japanese literature.[1,2,3] It usually presents in infancy and early childhood but late-onset has been reported on occasion.[4,5] Tufted angioma has rarely been reported in pregnancy.[6] The etiology and pathogenesis remains unknown and its morphology is variable. Tufted angioma presents as erythematous to dull-red stain-like macules progressing into a thick, large plaque-like lesion with mottled appearance, varying from 2-5 cm in diameter, mostly on the neck and upper trunk area.[7] Histologically, it is characterized by lobules or tufts of capillaries, which is also termed “cannonball distribution”.[8] We reported a case of late-onset and pregnancy-related tufted angioma in Thailand.

Case report

A 34-year-old Thai female presented with a two-year history of slowly enlarging red-blotch-lesions under the right breast. She reported that the lesions first appeared during the seventh month of her pregnancy. At that time, she experienced occasional pain on the affected area. She also noticed that the lesions gradually progressed, but stabilized after the delivery of her baby. She had no other diseases or family history of any systemic disease. The dermatological examination demonstrated mildly tender, non-blanchable erythematous papules which coalesced into plaques, 6 cm by 6 cm in diameter, located on the right inframammary area. [Figure1] The remaining physical examination was unremarkable.

Histopathology confirmed several islands of closely-set capillaries in the dermis. Thrombosis in some blood vessel was noticed. Neither pleomorphism nor increased mitosis was noted. [Figure2,3]

The CBC, including platelet count and the coagulogram, were also normal. Platelet was 290,000 cell/mm3. PT was 10.5 sec and PTT was 24.8 sec. She was diagnosed with late-onset, pregnancy related tufted angioma without complication.

She was treated with topical 0.25% timolol eye drops for several months without any visible improvement. She was then persuaded to be treated with sessions of Pulse dye laser (PDL, 595nm). The parameters were 7 mm spot size, fluence of 9 J/cm2, and pulse duration of 1.5 msec. Obvious improvement was observed after 3 sessions of laser treatment, and excellent results persisted up to the last follow up six months after stopping laser treatment. [Figure4] No complication was noted.

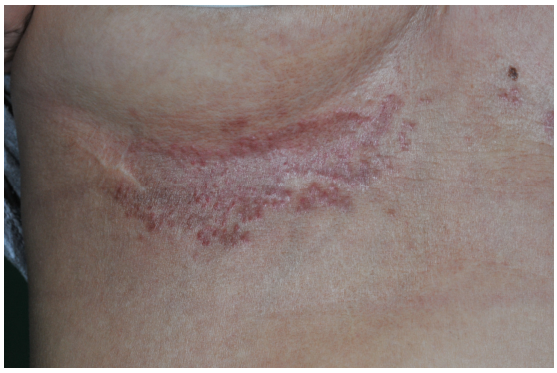

Figure 1 Large area of painful, erythematous papules which coaelesced into plaques on the right inframammary area

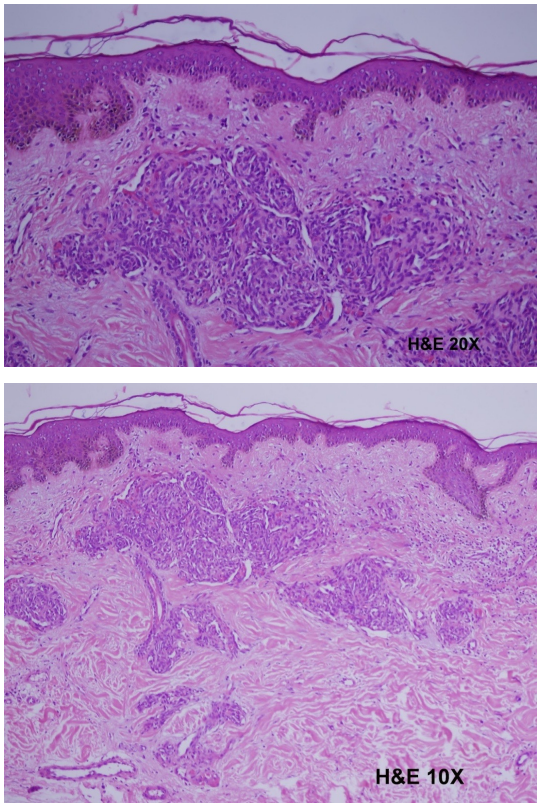

Figure 2 Well-circumscribed lobules of densely packed bloodless capillaries scattered throughout the dermis and subcutaneous tissues giving a special pathognomonic “cannonball” appearance without any mitotic figures. These capillary lobules have normal separating collagen and each lobule is surrounded by crescent-shaped vascular channels [10x].

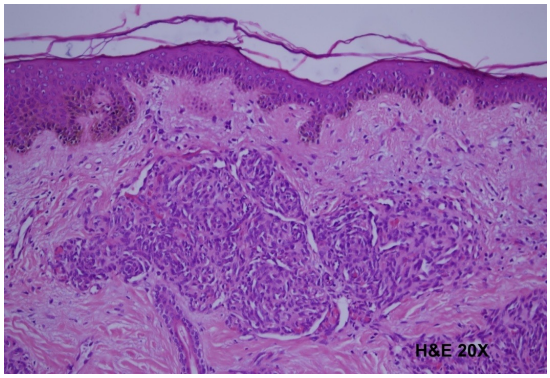

Figure 3 Well-circumscribed lobules of densely packed bloodless capillaries scattered through the dermis and subcutaneous tissues giving a special pathognomonic "cannonball" appearance without any mitotic figures. These capillary lobules have normal separating collagen and each lobule is surrounded by crescent-shaped vascular channels[20x]

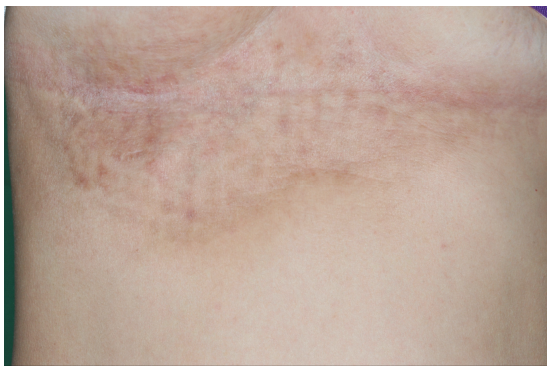

Figure 4 Marked improvement after three sessions of pulse dye laser, 595 nm

Discussion

Tufted angioma, also known as "Angioblastoma of Nakagawa" in Japanese literature, is a rare slow-growing benign vascular neoplasm. It usually presents in infancy and early childhood, especially in children under 5 years old.[1] In addition, congenital and adult-onset tufted angioma have also been rarely reported in medical literature.[1,2,3] It was first described as an angioblastoma in 1949 by Nakagawa.[4,5]

Tufted angioma occurs as a sporadic genetic mutation. Familial occurrence has also been reported, possibly monogenic autosomal dominant trait with reduced penetrance tendency.[2,3] The etiopathogenesis remains unclear. It maybe aggravated by hormonal effects, cytokine or chemokine.[7] There were cases of tufted angioma associated with pregnancy reported in 2002 but few other cases can be found in literature.[6] It has been hypothesized that cytokine may play a role in angiomatous proliferation, particularly IL-8.[7]

The morphology of tufted angioma is variable. It may present as a dull red, brown-red or purple lesion with large firm patches or thickened indurated plaques with mottled appearance.[8] The size of the lesion ranges from 5cm in diameter to more than 10 cm.[9] It often presents as a solitary lesion but sometimes presents as multifocal. Tufted angioma can be found anywhere on the body but is more commonly seen on the neck, trunk, and shoulder, and more seldom on the extremities. Hypertrichosis, localized hyperhidrosis and

warmth to the touch have been reported in some cases.[10] Extensive involvement is common even though it is entirely benign. There have been no reported aggressive or metastasis cases.[12]

Tufted angioma usually progresses slowly then stabilizes.[8,9] It may persist for years or spontaneously resolve[11]; however, partial self-involution has been reported and complete self-involution is rare.[12,13] In our case, the lesion spread slowly from the onset until delivery of her child and then stabilized. It remained stable for two years until the patient sought medical attention.

Congenital cases of tufted angioma that present with petechiae and ecchymotic patches may be associated with Kasabach-Merritt phenomenon (KMP), a life-threatening complication. However, in our case, the coagulogram was normal therefore KMP was excluded.[12,14,15]

Histopathology of tufted angioma has a specific pattern.[12] It is characterized by circumscribed lobules of densely packed bloodless capillaries scattered throughout the dermis and subcutaneous tissues, giving a special pathognomonic "cannonball" appearance without any mitotic figures. These capillary lobules have normal separating collagen and each lobule is surrounded by crescent-shaped vascular channels.[8,9] [figure2,3] Immunochemical profiles reveal positivity for lymphothelial, endothelial and Wilms Tumor 1(WT1) markers but negative for glucose transporter 1(GLUT1).[16] The differential diagnoses are infantile hemangioma, pyogenic granuloma, kaposi sarcoma and kaposiform hemangioendothelioma. These can be distinguished by clinical, histological, and immunochemical methods.[10]

Treatment focuses on cosmetic issues and complications that are associated with tufted angioma such as obstruction to vital organs, Kasabach-Merritt syndrome and functional compromise. Medical treatments available are topical or systemic corticosteroids, interferon alpha-2a and topical propranolol. Surgical excision and laser therapy are other effective treatments.[11] However, most cases of tufted angioma spontaneously regress within two years.[11] Most of the authors recommend that serious complications be treated with systemic corticosteroid or interferon-α.[12] Patients without complications can be treated with laser, as in our case.

There have been many reported cases of good responses to pulse dye laser treatment, with the concept of selective photothermolysis[17,18,19] It has been used in several vascular lesions such as port wine stain and hemangioma.[20] Robati et al(2002) showed successful treatment of tufted angioma by three

sessions of 595 nm pulse dye laser. The fluence was 10 J/cm 2, with pulse duration of 3 msec and spot size of 7 mm.[17,18] In line with this report, our patient was treated with sessions of pulse dye laser[PDL, 595nm] every month. The laser parameter was 7 mm spot size, fluence of 9 J/cm2, and pulse duration of 1.5 msec. The patient saw a dramatic improvement after 3 sessions of pulse dye laser.[fiqure 4] No further laser treatment was administered.

Conclusion

The incidence of reported tufted angioma in adults is rare and pregnancy-related cases are even more scarce, but such cases have been previously documented in literature. Even though the cause of tufted angioma is unknown, it is postulated that the hormonal effect might be the main etiopathology in pregnancy-related cases. In patients with no complications, topical or laser treatment is the recommended methods for cosmetic purposes, as tufted angioma is a benign vascular tumor. We reported a Thai woman with late-onset and pregnancy-related tufted angioma which responded extraordinarily to Pulse Dye Laser, 595 nm.

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