

### Case 19

A 59-year-old Thai female from Phetchabun

**Chief complaint:** Multiple erythematous papules and plaques on right chest wall for 3 years



#### Present illness:

The patient had developed tender erythematous papules and plaques on right chest wall for 3 years. She was diagnosed with irritant contact dermatitis at first time but the lesion was not relieved by topical corticosteroids. Then she sought second medical attention and had been diagnosed with herpes zoster with postherpetic neuralgia, treated with oral acyclovir and gabapentin. However, the lesion was not improved.

**Past history:** She has no underlying disease.

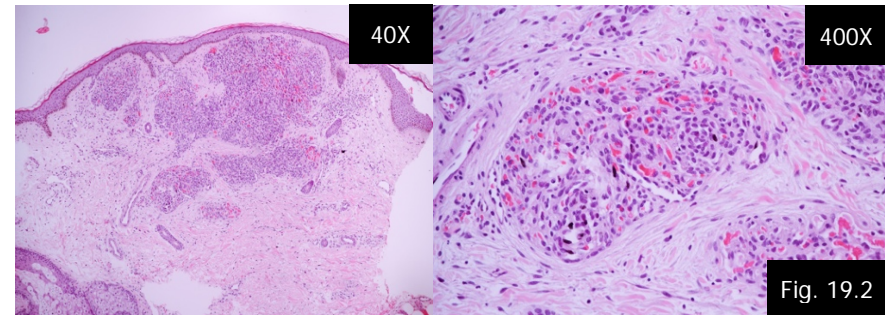
**Family history:** There was no family history of similar lesions.

**Physical examination:** Other systemic examination revealed no abnormality.

**Dermatological examination:** Multiple non-blanchable erythematous papules coalescing to form plaques on right chest wall (Fig. 19.1)

**Histopathology:** (SC19-00327, chest) (Fig 19.2)

- Scattered, round lobules composed of bloodless capillaries surrounded by dilated crescentic vascular channels, in the upper dermis



#### Laboratory investigations:

- CBC: Hb 12 g/dL, Hct 36%, Plt 211,000 /mm<sup>3</sup>, WBC 6,350 /mm<sup>3</sup> (N 46%, L 40%, M 10%, E 3%, B 1%)

**Diagnosis:** Tufted angioma

**Treatment:** Pulse dye laser

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#### Discussion:

Tufted angioma (TA) is a rare benign vascular tumor characterized histologically by small tufts of capillaries associated with dilated lymphatic vessels in the dermis.<sup>1</sup> It is usually affected children and young adults, but both congenital cases and late-onset cases have been also reported. Approximately 60-70% of TA develop

before the age of 5 years.<sup>3</sup> Less than 10% of TA occur after the age of 50 years like in our case. It occurs in both sexes and predilection for the neck, upper chest, back, and shoulder.<sup>2</sup> Lesions are initially seen as solitary tumors or infiltrating plaques that are dusky red or violaceous, sometimes associated with hyperhidrosis or hypertrichosis. Evolution of the disease seems to vary considerably, including complete or partial spontaneous regression<sup>4-6</sup>, persistence over the years<sup>4</sup>, or Kasabach-Merritt syndrome (KMS).<sup>7-8</sup>

The characteristic histology consisting of a proliferation of endothelial cells forming lobules in the middle and lower part of the dermis, and in subcutaneous fat, with a specific "cannon-ball" pattern. The aggregates of endothelial cells are concentrically whorled along a preexisting vascular plexus.<sup>3</sup> Some lobules bulge the walls of dilated thin-walled vascular structure, giving a semi-lunar appearance to the vessels: this aspect, in addition to the angiocentricity, prompted the name "Tufted angioma".<sup>3</sup> Immunohistochemistry (IHC) demonstrates that the cells in the capillary tufts are positive for CD31, CD34 and, rarely, for smooth muscle actin and negative for GLUT1.<sup>3</sup> Histopathologic examination with IHC is necessary to exclude other lesions such as hemangioma of infancy, vascular malformations, infantile myofibromatosis, and congenital dermatofibrosarcoma protuberans.<sup>9</sup>

No specific laboratory study is useful in the diagnosis or treatment of TA. If the coexistence of KMS is suspected, a complete blood cell count with a determination of the platelet count, prothrombin time and/or activated partial thromboplastin time and full disseminated intravascular coagulation profile is indicated.<sup>10</sup>

Medical treatment is not necessary for TA, due to the benign nature, no malignant transformation, and slow progression of these vascular tumors: only clinical observation is recommended. Treatment may be categorized as that administered for aesthetic reasons or as that instituted because of complications or anticipated complications such as KMS or functional compromise. Several treatment regimens for TA have been reported, including compression therapy, surgery, pulse

dye laser, topical or systemic corticosteroids, interferon- $\alpha$ , and chemotherapy.<sup>11-12</sup>

In our case, the clinical presentation, histopathology and IHC most suggest TA. She has been treated with pulse dye laser. After 3 sessions of treatments, the lesion was minimally improved in size.

#### References:

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