Persistent facial angioedema
Mei-Chin Ho Ming-Tuo Chuan Shu-Ling Hu

CASE REPORT
The 61-year-old male had peripheral vascular occlusive disease and hypertension under medical control. He had no allergic history of drug and food. He presented with asymptomatic facial edema for more than 4 months. The edema began at forehead then extended to eyelids and bilateral cheeks as well as scalp (Fig. 1). No response to antihistamine and diuretics were also noted. Laboratory data showed BUN: 21 mg/dl, Cr: 0.8 mg/dl, urinalysis: no proteinuria, C3: 119 mg/dl, C4: 22.1 mg/dl and ANA: negative. The cardiac sonography showed no abnormality. The sonography of renal vein and jugular vein revealed no stenosis. The carotid color duplex showed normal extracranial vascular ultrasonography study and no significant stenosis of the intracranial arteries. The skin biopsy over his left cheek was performed and the histopathology of the tissue specimen was shown as follows (Fig. 2, 3). The tumor cells revealed positive for CD34 and Ulex europaeus lectin 1 (Fig. 4), but negative for factor VIII and cytokeratin.

Fig. 1
Persistent facial swelling for 4 months.

Fig. 2
The neoplastic cells are arranged in short fascicles, small nests and irregular anastomosing vascular channels in dermis and subcutis. (H & E, x40)

Fig. 3
The neoplastic cells are characterized by moderate to marked nuclear pleomorphism and hyperchromasia. (H & E, x200)

Fig. 4
The tumor cells are positive for Ulex europaeus lectin 1. (400X)

From the Department of Dermatology, Cathay General Hospital
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Reprint requests: Ming-Tuo Chuan, M.D., Department of Dermatology, Cathay General Hospital, No 280, Section 4, Jen-Ai Road, Taipei, Taiwan
TEL: 886-2-27082121 ext. 5078 FAX: 886-2-2707-4949
**DIAGNOSIS: Cutaneous Angiosarcoma**

**DISCUSSION**

Most cutaneous angiosarcoma arise in the following clinical settings: angiosarcoma of the face and scalp in the elderly, angiosarcoma secondary to chronic lymphoedema and angiosarcoma as a complication of chronic radiodermatitis or arising from the effects of severe skin trauma or ulceration. Three clinical types of the angiosarcoma of the scalp and face in the elderly exist: an ulcerative type, a diffuse superficial type, and a nodular type. All three types may be intermingled. Severe uncommon manifestations have been reported such as purpuric blister, deep seated cyst, scaring alopecia, rosacea-like lesion and recurrent angioedema of the face.\(^1,2\)

The clinical differential diagnosis of angiosarcoma includes dermatitis, infections, bruise, benign tumor, malignant tumor, collagen vascular diseases, rosacea, and angioedema.

Angiosarcoma presenting with angioedema of the face is uncommon. The edema may be due to the lymphatic vessels infiltrated by malignant cells or due to lymphedema. However, the actual mechanism has not been confirmed.

The etiology is unknown and the prognosis is poor. The current treatment strategy is to excise focal tumor followed by radiation therapy and/or chemotherapy. Doxorubicin and paclitaxel\(^3\) are the common chemotherapeutic agents recently. Many new therapies have been tried including intraliesional rIL-2 with OK-432,\(^4\) liposomal doxorubicin following radiation therapy,\(^5\) and IFN-α 2a with 13-cis-retinoid acid. The search for effective systemic treatment for angiosarcoma is necessary.

Cutaneous angiosarcoma is a highly aggressive neoplasm and the first challenge is early diagnosis. However, diagnosis is often delayed for many reasons: asymptomatic early lesions, patient in advanced age, low suspicion of malignancy and often misdiagnosed. Our case indicated that angiosarcoma may initially present as persistent facial angioedema. Thus we should be alert to such unusual manifestation.

**REFERENCE**