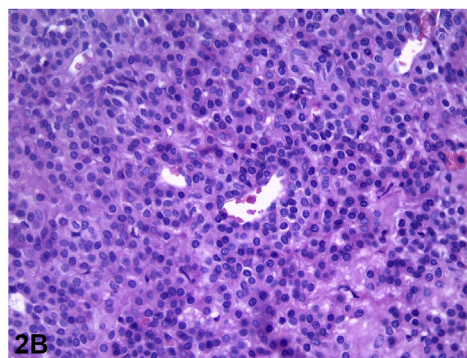
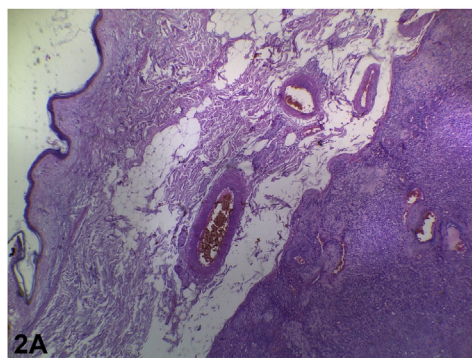


Long-term telangiectatic nodule in left shoulder



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Key words: extradigital; glomus cells; glomus tumor; shoulder.



A 70-year-old healthy male presented with a 12-year history of a painful cutaneous vascular lesion on his left shoulder. It had begun as a slowly growing, small, erythematous macula covered by telangiectasias. The pain was triggered by pressure or palpation on the shoulder. Neither cold intolerance nor paroxysmal pain was reported. The patient noticed that symptoms had been increasing in severity over the past two years. Physical examination found a single, well-demarcated, purplish-red dermal nodule, which was 12 mm in diameter.

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Funding sources: None.

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JAAD Case Reports 2021;10:38-40.

2352-5126

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<https://doi.org/10.1016/j.jidcr.2020.12.039>

Additionally, it showed multiple, radiating, non-blanching telangiectasias (Fig 1). The lesion was extremely painful on palpation, and restricted shoulder abduction was observed. Complete blood count, coagulation tests, and comprehensive metabolic panel results were within normal ranges. Preoperative imaging studies were not requested. The nodule was completely removed using a 7-mm surgical margin. Only hematoxylin-eosin-staining was carried out (Fig 2). There was no recurrence of the skin lesion in the first year.

Question 1: What is the most likely diagnosis?

- A. Giant spider nevus
- B. Cutaneous angiosarcoma
- C. Cutaneous epithelioid angiomatous nodule (CEAN)
- D. Glomus tumor
- E. Telangiectatic Kaposi sarcoma (KS)

Answers:

A. Giant spider nevus—Incorrect. This lesion is characterized by multiple blood vessels in a radial pattern that surrounds a central reddish spot; giant variants could resemble the patient's lesion.¹ Histologically, it's composed by a central arteriole in the upper dermis which feeds a net of ectatic capillaries.

B. Cutaneous angiosarcoma—Incorrect. This vascular neoplasm is rare and aggressive. It generally affects the head and neck of elderly patients. It presents as a single or multiple, ulcerated, reddish-purple lesion. It has several histological variants and shows an infiltrative growth pattern, cytological atypia, and numerous mitoses.

C. CEAN—Incorrect. CEAN is a reactive vascular proliferation. It appears as a rapidly growing nodule, mainly on the trunk and extremities. Histopathology shows a well-circumscribed nodule of epithelioid cells with eosinophilic cytoplasm, vesicular nuclei, and conspicuous nucleoli; focally, there may be scattered vascular channels.

D. Glomus tumor—Correct. This vascular hamartoma arises from the glomus body and usually appears in the subungual region of the fingers; at present, there are only 15 reported cases that have been located in the shoulder. They can occur as a painful or tender mass involving subcutis, muscle, or periarticular structures. The average time from the onset of symptoms to an accurate diagnosis is 10 years (range, 0.5-20 years).^{2,3} Pathology shows a well-circumscribed dermal nodule composed of solid aggregates of uniform round small cells with eosinophilic cytoplasm and surrounding normal, dilated, or congestive capillary vessels.

E. Telangiectatic KS—Incorrect. KS is a low-grade vascular neoplasm, which arises from lymphatic endothelium. Telangiectatic KS is a very unusual histological variant.⁴ It presents as a nodular proliferation of spindle cells with multiple, dilated, and congested vascular spaces.

Question 2: What associations have been involved in the physiopathology of this entity?

- A. Chronic liver disease
- B. Neurofibromatosis type I (NF-1)
- C. None
- D. Chemical carcinogens
- E. Human herpesvirus 8 (HHV-8)

Answers:

A. Chronic liver disease—Incorrect. Chronic liver disease, mainly due to alcoholic cirrhosis, has been described in patients with spider nevi. Higher plasma levels of vascular endothelial growth factor and substance P have been found in these patients, but their exact role still remains unknown.¹

B. NF-1—Correct. NF-1 is an autosomal dominant disorder. The gene involved is located on chromosome 17q11.2 and codes for neurofibromin. Glomus tumors have been described in individuals with NF-1. They appear as solitary or multiple lesions and show a predilection for acral sites, such as fingers and toes. NF1-associated glomus tumors present hyperactivation of the Ras/Mitogen-activated protein kinase (MAPK) pathway produced by loss of inhibition of neurofibromin.⁴

C. None—Incorrect. An unremarkable medical history is described in patients with CEAN. This entity is rare, and usually occurs in healthy individuals.

D. Chemical carcinogens—Incorrect. Chemical carcinogens, such as vinyl chloride, have been associated with the development of angiosarcoma.⁴

E. HHV-8—Incorrect. HHV-8 plays an important role in the physiopathology of KS, and the HHV-8 genome has been isolated in all clinical subtypes.⁴

Question 3: Which of the following would be an appropriate paraclinical test for this patient?

- A. Magnetic resonance imaging (MRI)
- B. Liver function panel
- C. None
- D. HIV test
- E. Color duplex Doppler sonography

Answers:

A. MRI—Incorrect. MRI would be more appropriate in a patient with cutaneous angiosarcoma. It helps to delineate the local extent of the primary tumor before surgery. In addition, a computed tomography scanning could exclude metastatic disease. Lymph nodes and lungs are the primary sites of metastasis.⁴

B. Liver function panel—Incorrect. This would be the initial lab test used to exclude chronic liver disease in a patient with spider nevi, especially if these are multiple or there are risk factors for liver disease.

C. None—Incorrect. This would be the most appropriate conduct in a patient with CEAN, since CEAN usually occurs in healthy individuals and with no relevant clinical associations.

D. HIV test—Incorrect. It would help to exclude AIDS-related KS. The diagnostic approach is based on the type of extracutaneous manifestation. The gastrointestinal tract, lymph nodes, and lungs are commonly involved sites.⁴

E. Color duplex Doppler sonography—Correct. Doppler sonography shows a well-defined hypo-echoic lesion; it can also determine the location and

size, even for small nodules of only 2 mm in diameter. Additionally, this is the preferred preoperative imaging technique due to its accessibility and low cost. Some studies have reported a 100% detection rate and no false-negative results with color duplex Doppler sonography. MRI has also proven to be a very useful diagnostic technique, but its main indication is for multiple or recurrent glomus tumors. MRI reveals a hypointense tumor on T1-weighted sequence, a hyperintense tumor on T2-weighted sequence, and enhancement on T1-weighted sequence after receiving an injection gadolinium. It has a sensitivity of 90% but a specificity of only 50%.⁵

Abbreviations used:

CEAN: cutaneous epithelioid angiomatous nodule

HHV-8: human herpesvirus 8

KS: Kaposi sarcoma

MRI: magnetic resonance imaging

NF-1: neurofibromatosis type I

Conflicts of interest

None declared.

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