Multiple tender bluish nodules

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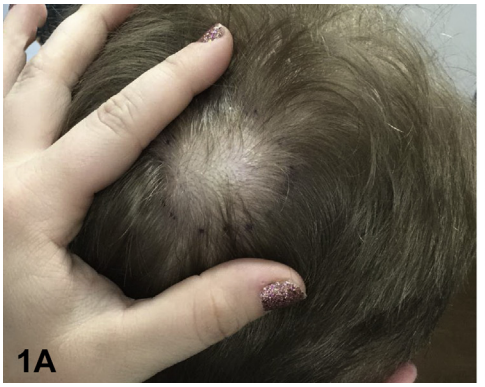

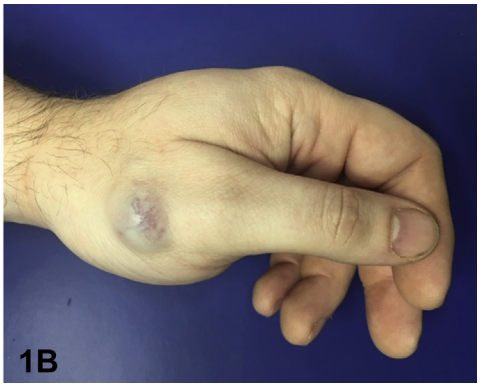

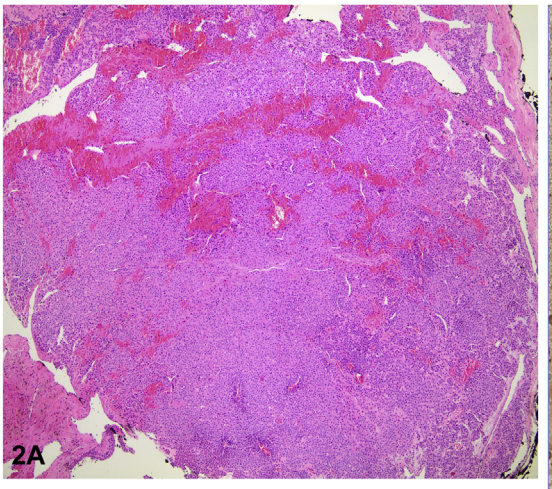

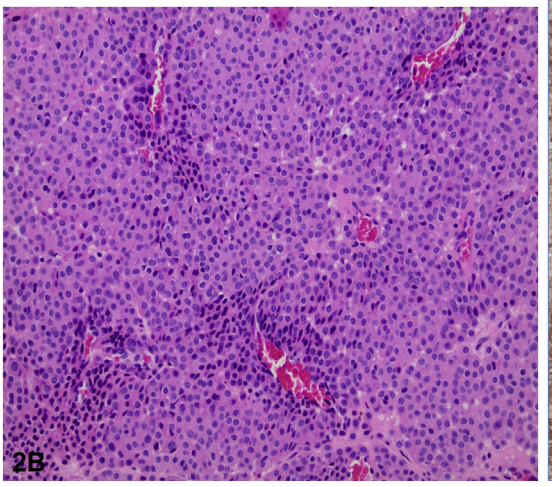

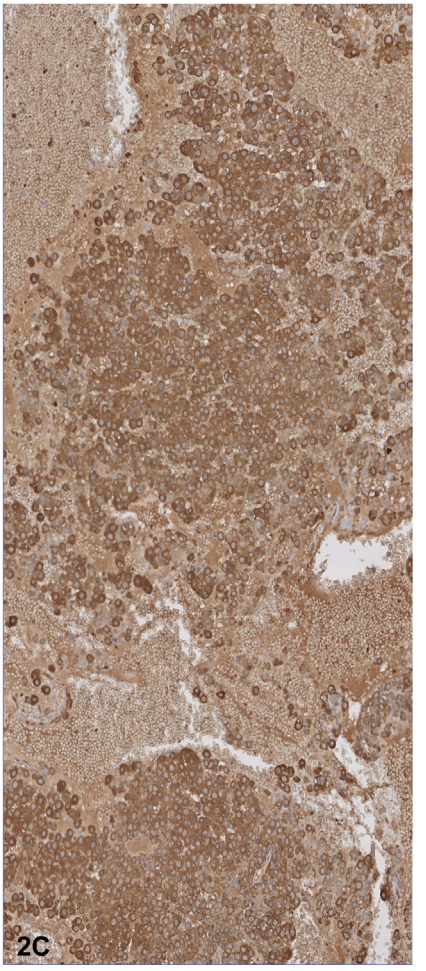

A 35-year-old man presented for evaluation of nodules that were present since childhood. He reported that his father had similar nodules. Physical examination found multiple tender, compressible subcutaneous nodules with bluish hue on the forearms, hands, and fingers, and a similar nodule without bluish coloration on the scalp (Fig 1). He underwent surgical excision of the scalp nodule because of tenderness. Histopathologic findings are shown in Fig 2 with panel C showing smooth muscle actin immunohistochemistry.

Question 1: What is the correct diagnosis?

A. Blue rubber bleb syndrome

B. Multiple glomus tumors

C. Multiple spiradenomas

D. Arteriovenous malformations

E. Nodular Kaposi sarcoma

Answers:

A. Blue rubber bleb syndrome — Incorrect. This genetic syndrome also presents with multiple bluish papules and nodules.[1] However, the presence of monomorphous smooth muscle actin positive cells is consistent with glomus cells,[2] which is not seen in this entity.

B. Multiple glomus tumors — Correct. The histopathologic findings of vascular spaces with monomorphous round cells that stain positively with smooth muscle actin show that these tumors are made up of glomus cells, hence, the diagnosis of multiple glomus tumors. Multiple glomus tumors have a genetic component with an autosomal dominant inheritance, variable expressivity, and incomplete penetrance.[1] They are most commonly located in the extremities[1] but are also described in other locations including trunk, scalp, face, and visceral organs.[1,3] Complete excision can be both diagnostic and therapeutic for these tumors.

C. Multiple spiradenomas — Incorrect. Although spiradenomas clinically present as subcutaneous nodules sometimes with a bluish to bluish-red color, the pathology in this case is not consistent with this diagnosis. The histologic hallmark of spiradenoma is a dermal nodule with basophilic cells arranged in a trabecular network and presence of lymphocytes.[4]

D. Arteriovenous malformations — Incorrect. The pathology of this entity consists of central thin-walled vessels surrounded by thick-walled vessels.

E. Nodular Kaposi sarcoma — Incorrect. Kaposi sarcoma presents as violaceous nodules and plaques with pathology showing proliferation of spindled endothelial cells with slit-like vascular spaces.[5] The presence of smooth muscle actin positive monomorphous cells is consistent with glomus tumors.[2]

Question 2: What is the most common location of these tumors?

A. Scalp

B. Face

C. Acral extremities

D. Mucosal

E. Trunk

Answers:

A. Scalp — Incorrect. Although glomus tumors can be present on the scalp, as seen in our patient, this is not a common location.

B. Face — Incorrect — Glomus tumors are not frequently observed on the face.[1,3]

C. Acral extremities — Correct. Acral extremities are the most common location of both solitary and multiple glomus tumors. Approximately 90% of patients with multiple glomus tumors had tumors in their extremities in a retrospective study.[1,3]

D. Mucosal — Incorrect. Although mucosal glomus tumors have been reported, these are rare entities.[3]

E. Trunk — Incorrect. Although glomus tumors can often be found on the trunk, this is not the most common location.[1,3]

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Question 3: Besides smooth muscle actin, which of the following immunohistochemistry stains can be used to identify glomus cells?

A. S-100

B. Desmin

C. Vimentin

D. Cytokeratin

E. CD31

Answers:

A. S-100 — Incorrect. Glomus cells do not stain with S-100.[2]

B. Desmin — Incorrect. Although, smooth muscle typically stains with desmin, vascular smooth muscle cells including glomus cells do not stain with desmin.[2]

C. Vimentin — Correct. Glomus cells stain with both smooth muscle actin (Fig 2, *C*) and vimentin.[2]

D. Cytokeratin — Incorrect. Glomus cells do not stain for cytokeratin.[2]

E. CD31 — Incorrect. Although CD31 is normally positive in endothelial cells, glomus cells, which are specialized vascular smooth muscle cells, do not stain for CD31.[2]

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