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Glomuvenous Malformation: A Rare Periorbital Lesion of the Thermoregulatory Apparatus

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Abstract

Glomuvenous malformations (GVMs), previously referred to as glomus tumors or glomangiomas, are benign, mesenchymal venous malformations arising from glomus bodies. Glomus bodies are modified smooth muscle neuromyoarterial structures involved in temperature regulation via blood shunting. These classically occur in the digits but can occur in other locations. The authors present a case of a periorbital GVM presented following blunt trauma to the area.

Glomuvenous malformations (GVMs), previously referred to as glomus tumors or glomangiomas, are benign, mesenchymal venous malformations arising from glomus bodies. Glomus bodies are modified smooth muscle neuromyoarterial structures involved in temperature regulation via blood shunting. GVMs are most commonly found in the subungual areas of the hand due to the high concentration of glomus bodies in the digits. Rarely, GVM have been reported in the head and neck. The authors report a case of periorbital GVM that presented following trauma to this location. Collection and evaluation of protected patient health information were Health Insurance Portability and Accountability Act compliant.

CLINICAL CASE

A healthy 41-year-old woman presented with a 2-cm, exquisitely tender mass in the right temple, worsening over the previous 5 years (Fig. 1A,B). Six years prior, she experienced blunt trauma to the right temple in a motor vehicle accident that caused a hematoma. Following resolution of the hematoma, she noted a residual mass associated with sharp pain

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that was sensitive to changes in temperature, particularly cold air. This was initially diagnosed as a traumatic neuroma by her neurologist, and she was unsuccessfully treated with nonsteroidal anti-inflammatory drugs, gabapentin, opioids, and onabotulinum toxin.

Examination demonstrated visual acuity of 20/20 in each eye, normal pupils, and full extraocular movements. Ocular examination was normal. She had mild right brow ptosis and mild weakness of the temporal branch of the facial nerve. There was a 2-cm, spongy, freely mobile mass over her right temple that was tender to palpation. MRI revealed a T1-hyperintense, well-circumscribed mass in the subcutaneous tissue along the temporal branch of the facial nerve (Fig. 1C).

Excisional biopsy was performed through a superolateral eyelid crease incision (Fig. 1D). On gross inspection, the lesion appeared vascular (Fig. 2A,B). Histopathology demonstrated a well-circumscribed mass surrounded by a fibrous capsule (Fig. 3A). The neoplasm was composed of a uniform population of cells with round-to-oval nuclei, salt and pepper chromatin, and pale eosinophilic cytoplasm. No necrosis or mitotic bodies were identified (Fig. 3B). Immunohistochemical staining was positive for type IV collagen (Fig. 3C), smooth muscle actin, myosin smooth muscle heavy chain antibody smooth muscle myosin heavy chain antibody, and vimentin. It was negative for desmin, factor XIIIa, S-100, chromogranin, and synaptophysin. Rare cells were positive for Ki67/MIB-1 (Fig. 3D). These findings were consistent with the diagnosis of GVM.

Postoperatively, the patient was pleased with both relief of symptoms and aesthetic improvement. She experienced mild weakening of the ipsilateral frontalis muscle that resolved after 6 weeks. At 2-year follow up, she remains well.

DISCUSSION

This case is unusual due to the location of the GVM and its presentation following local trauma. In a case series of 56 extradigital GVM, only 2 were reported in the face: 1 on the cheek and 1 on the nose.¹ Other reports of facial GVM include lesions in the cheek, mandibular area, oral mucosa, eyelid, and retrobulbar orbit but not in the brow region.²⁻⁶ Masson⁷ provided the first clinical description of this condition in 1924 with the classic triad of excruciating pain out of proportion to size, localized tenderness, and temperature sensitivity, particularly to cold.

The relationship between trauma and the development of GVM is not known, although proliferation of neural and vascular structures at locations of vascular anastomosis has been hypothesized.⁸ Malignant potential has not been demonstrated for GVM, and treatment is indicated for symptom relief after histologic diagnosis is confirmed. Treatment is primarily via surgical excision, although successful treatment of GVM with laser (including ND:YAG), sclerotherapy, and electron beam radiation has been reported.⁹⁻¹¹

Of note, the previously used term glomus tumor has also been used historically, and inaccurately, as a synonym for paraganglioma in the head and neck, although these are distinct clinical entities. Paragangliomas arise from paraganglia, neural crest-derived endocrine cells that form part of the neuroendocrine system, while GVM are derived from

neuromyoarterial glomus bodies. Paragangliomas can also occur in the head and neck and are typically benign. In the English literature, there are 12 cases of paragangliomas arising along the facial nerve, so called glomus faciale. These primarily presented clinically with progressive facial nerve palsy.¹²

CONCLUSION

GVM occur rarely in the periorbital region, yet are essential to include in the differential diagnosis of patients presenting with soft tissue masses, pain out of proportion to examination, and sensitivity to temperature change. Otherwise, incorrect treatment modalities may be attempted unsuccessfully. CT and MRI are useful to narrow the differential diagnosis and identify the extent of involvement, but tissue biopsy for histology and immunohistochemistry is necessary to make the diagnosis. Treatment is primarily surgical resection, with the goal of preserving cranial nerve function.

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FIG. 1.

A, Clinical photograph of the patient at presentation (*arrow*). **B**, Profile photograph of lesion (*arrow*). **C**, Axial, T1-weighted MRI showing hyperintense lesion (*arrow*). **D**, Preoperative markings of lesion (*dotted line*) and planned surgical incision (*solid line*).

FIG. 2.
A, Vascular lesion after resection and surgical site. **B,** Excised lesion measuring 14 mm in diameter.

FIG. 3.

A, Hematoxylin and eosin (H&E) staining at low power shows well-circumscribed, encapsulated mass. **B**, H&E staining at high power shows prominent basophilic nuclei but no mitotic bodies or necrosis. **C**, Collagen type IV positivity. **D**, Rare cells are positive for Ki67/MIB-1.