

# Sclerotic Glomus Tumor

Felix A. Vigovich, MD, Mark A. Hurt, MD, and Daniel J. Santa Cruz, MD

**Abstract:** We report an unusual histopathological variant of a glomus tumor that arose in a peculiar topographic site, a *sclerotic* glomus tumor. Unlike conventional glomus tumors or glomangiomas that have a loose fibrous stroma with variable hyaline and myxoid changes, the case reported herein had a diffuse, hyalinized, sclerotic stroma. A further difference was that the majority of glomus tumors and glomangiomas occur in the subungual area, trunk, or extremities, whereas the present tumor occurred on the ear. Due to the peculiar histological features and location, other tumors were considered in the differential diagnosis to include Merkel cell carcinoma, primitive neuroectodermal tumor, and small cell melanoma. This article illustrates a unique variant of a glomus tumor, which to our knowledge has not been previously described.

**Key Words:** glomangioma, glomus tumor, stromal sclerosis, differential diagnosis with basal cell carcinoma, differential diagnosis with melanoma, acrospiroma, hidradenoma

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## INTRODUCTION

Glomus tumors are benign mesenchymal neoplasms composed of cells similar to the epithelioid smooth muscle cell located within the walls of a specialized arteriovenous anastomosis (the Sucquet–Hoyer canal), a part of the glomus body. The glomus vessels are dermal neurovascular organs involved in temperature regulation.<sup>1–5</sup>

Most glomus tumors develop in the dermis or subcutis of extremities (especially the subungual region but also the hand, wrist, or foot); however, they can affect gastrointestinal tract, genitourinary tract, mediastinum, lung, liver, pancreas, ovary, bone, and soft tissues.<sup>2,6,7</sup> There is no gender predilection except for the digital and subungual neoplasms, which tend to occur in women predominantly.<sup>2,5</sup>

Cutaneous forms of glomus tumors present as blue-red dermal nodules are characteristically associated with paroxysmal pain and are usually less than 1 cm in greatest dimension.<sup>5,6</sup>

Histologically, glomus tumors are classified as<sup>1,5,7–12</sup>

1. *Glomangioma*
2. *Glomangiomyoma*
3. *Solid glomus tumor*
4. *Glomangiomatosis* (histologically benign glomus tumor with diffuse growth)
5. *Glomangiosarcoma* (malignant analogue of glomus tumor)

Reprints: Felix A. Vigovich, MD, British Hospital of Buenos Aires, Perdriel 74, Buenos Aires, Argentina 1426 (e-mail: felixvigo@yahoo.com.ar).  
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6. *Glomus tumors of uncertain malignant potential* (in essence, these are lesions that firm criteria for benignancy or malignancy, which have not been established objectively.)

7. *Symplastic glomus tumors* (tumors with high nuclear grade in the absence of any other malignant feature)

Rare variants show oncocytic and/or epithelioid cytology.<sup>13,14</sup>

## MATERIALS AND METHODS

A 28-year-old white woman presented with a discrete mass on the right ear. The lesion was excised. Gross examination revealed a 0.5 × 0.25-cm ellipse of skin with an underlying 0.3 × 0.3 × 0.25-cm ovoid, firm, semitranslucent, light tan nodule. The specimen was fixed in 10% neutral buffered formalin, processed, sectioned, and stained with hematoxylin and eosin and Masson trichrome. Immunohistochemical staining was performed using an avidin–biotin–peroxidase technique employing antibodies to a cytokeratin cocktail (AE1/AE3), CD31, CD34, Melan-A (A-103), and smooth muscle actin.

## RESULTS

### Light Microscopic Findings

Sections from the specimen showed a well-circumscribed monomorphic proliferation of small uniform basophilic cells with central, round to oval, punched-out nuclei in a fairly dense hyalinized stroma (Fig. 1). They were composed of interwoven coarse fascicles of thickened and homogenized eosinophilic collagen bundles arranged in laminated fashion (Fig. 2). The glomus cells were either present in solid lobules or as a ribbon pattern of thin columns, cords, and strands (Fig. 3). A relatively scant vascularity was identified within the solid lobules. Masson trichrome highlighted the prominent collagenous stroma (Fig. 4).

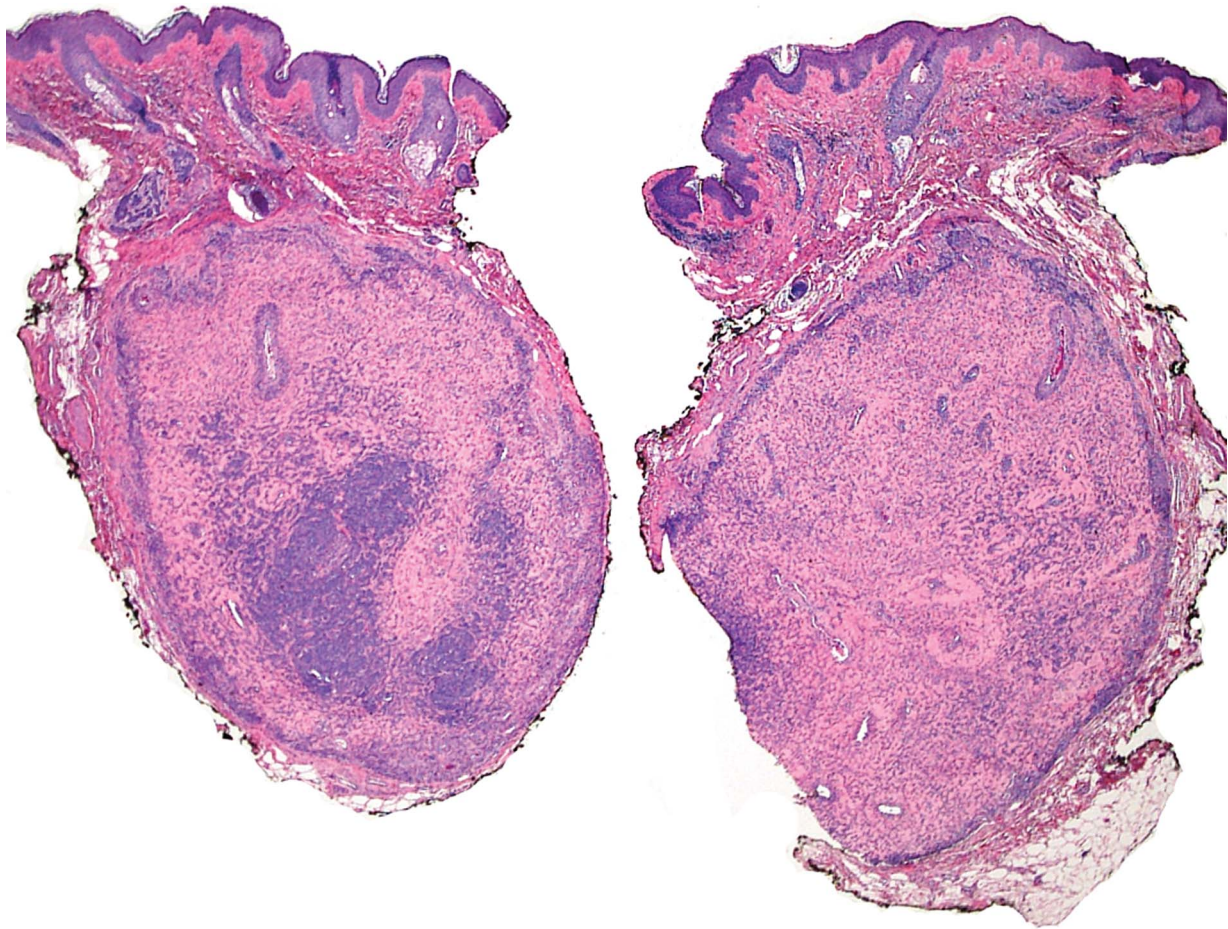
### Immunohistochemistry

Staining with CD31 highlighted only the endothelial component of the neoplasm (Fig. 5). Actin was positive in almost all these neoplastic cells (Fig. 6). Pancytokeratin, CD34, and Melan-A were negative in the lesion.

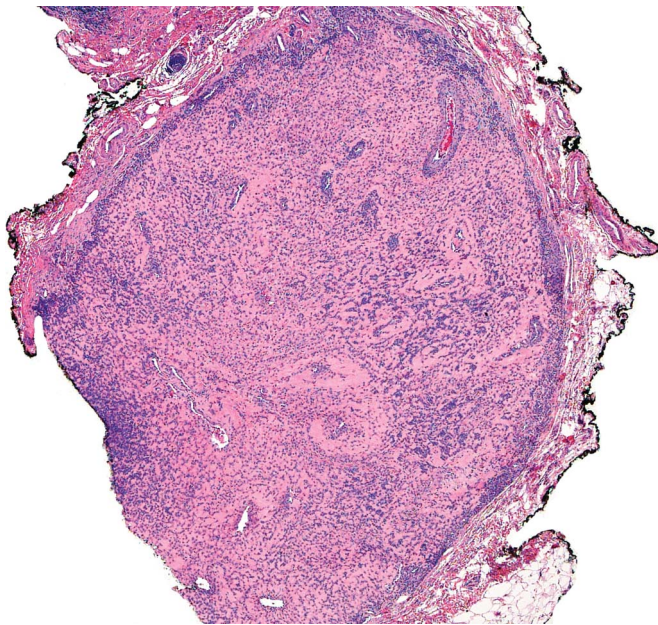
## DISCUSSION

In 1924, Masson described the first example of glomus tumor, a distinctive mesenchymal neoplasm composed of cells similar to modified smooth muscle cells, present in a specialized neurovascular organ serving as a thermoregulatory function, namely, the glomus body.<sup>15</sup>

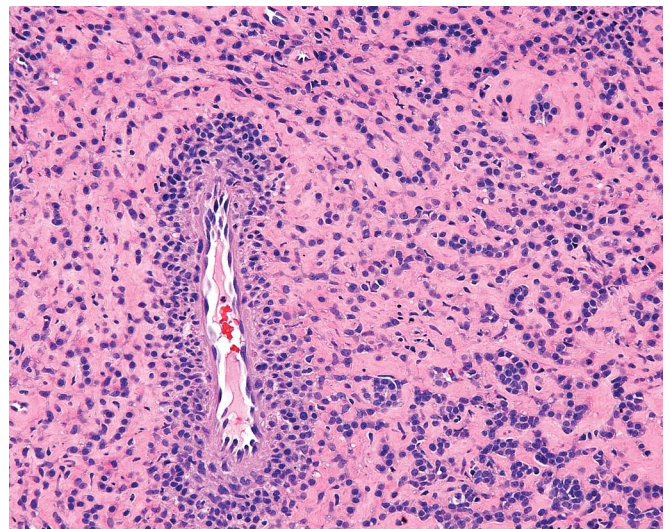
It was also Masson who published a case (in his textbook) that was somewhat similar to the one presented here except that our case is more sclerotic than the one he illustrated.<sup>4</sup>



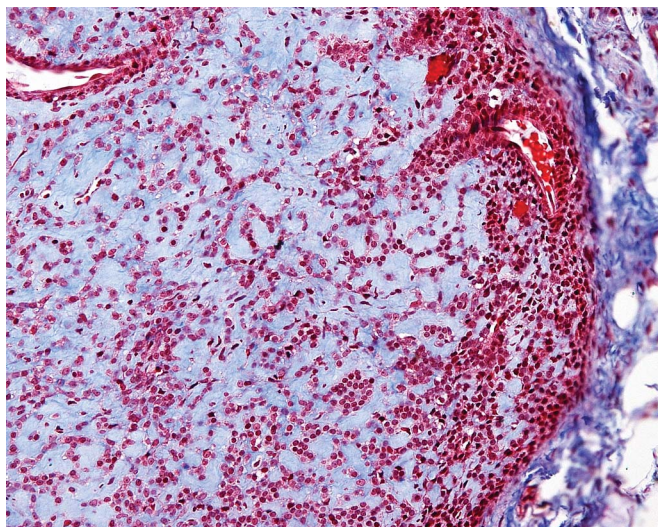
**FIGURE 1.** Low magnification showing a deep, dermal, well-circumscribed tumor.



**FIGURE 2.** Higher magnification of Figure 1 displaying a proliferation composed of fairly equal amounts of basophilic cells and eosinophilic stroma.



**FIGURE 3.** Vascular structure cuffed by glomus cells and surrounded by dense hyalinized stroma.

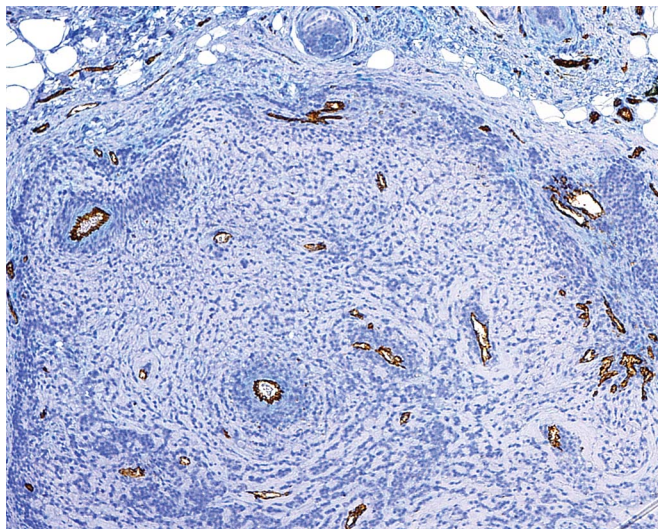


**FIGURE 4.** Masson trichrome stain enhances the sclerotic stroma.

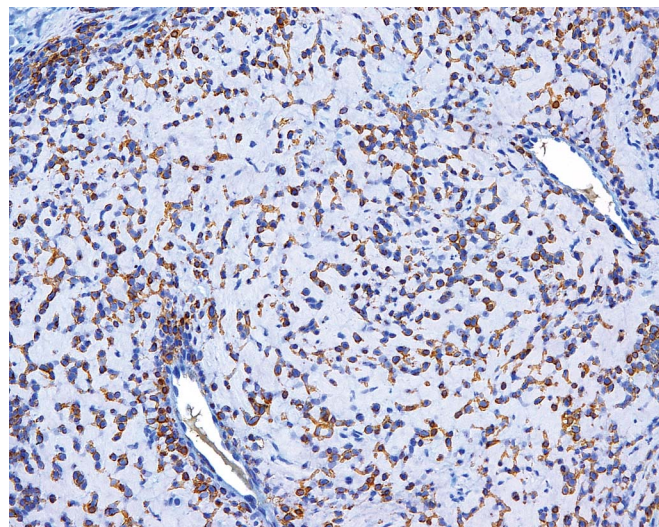
As a rule, glomus tumors contain monomorphous epithelioid cells and vascular structures. The stroma is usually rather inconspicuous in most cases. The malignant analogues are exceedingly rare and are composed of pleomorphic glomocytes that exhibit mitotic figures and necrosis, features simply not found in the benign lesions.

The lesion analyzed in this case report showed some features of classical glomus tumors, namely, a circumscribed mass of convoluted vascular structures cuffed by aggregates of glomus cells. In this particular case, however, the sclerotic pattern of the stromal matrix was very different from the classical cases of glomus tumor. Furthermore, although glomus tumors occur usually in distal extremities, our case developed on the right ear, an unusual site.

Lesions considered in the differential diagnosis were morpheiform basal cell carcinoma, hidradenoma/acrospiroma, and melanoma, which were excluded by morphology coupled with immunohistochemistry.



**FIGURE 5.** CD31 emphasizing vascular component.



**FIGURE 6.** Glomus cells are highlighted with immunoperoxidase technique for actin.

This sclerotic variant seems to be rare (or, at least, identified uncommonly), and it is not detailed in the major textbooks of dermatopathology or soft tissue tumors.<sup>1-3,6</sup>

In conclusion, we emphasize that glomus tumors can display several morphologic patterns including the epithelioid, solid, vascular, oncocytic, and myxoid variants.<sup>2,13,14</sup> This case shows that sclerotic stromal change can also occur in glomus tumors.

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