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Glomangiosarcoma Arising from a Prior Biopsy Site

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Summary: Glomangiosarcoma represents a rare malignant variant of the benign glomus tumor that typically presents as a tender, slowly growing nodule with a predilection for the lower extremities. Unlike their benign counterparts, glomangiosarcomas may display aggressive characteristics such as large size, local invasion, and a tendency to recur after excision. Although wide local excision remains the treatment of choice, rare cases of systemic metastasis have been previously reported. We present a case of glomangiosarcoma arising at a prior biopsy site after excision of an unknown soft tissue lesion. *(Plast Reconstr Surg Glob Open 2017;5:e1219; doi: 10.1097/GOX.0000000000001219; Published online 25 January 2017.)*

Glomus tumors are benign lesions arising from the glomus body, a specialized form of arteriovenous anastomosis in the reticular dermis that aids thermoregulation by regulating capillary blood flow. The majority presents as painful, solitary, slowly growing nodules with a predilection for the deep dermis or subcutis of the extremities. In rare circumstances, glomus tumors may show unusual clinical features such as large size, multicentricity, deep soft tissue infiltration, or extradermal origin.[1]

Glomangiosarcoma represents a rare malignant form of glomus tumor characterized histologically by nuclear atypia, necrosis, and high mitotic index (Park). Clinically, glomangiosarcomas display locally invasive behavior and a tendency to recur after excision. Rare cases of metastatic spread to the brain, bone, lung, liver, small intestine, mediastinum, and small bowel mesentery, which have been documented.[2,3] We present a case of glomangiosarcoma arising at a prior biopsy site.

CASE PRESENTATION

A 68-year-old man presented with a painful, slowly enlarging, palpable subcutaneous mass of the medial left buttock. A similar lesion, a "sebaceous cyst" by history, was excised from the same area 10 years earlier. Pathological evaluation had not been performed.

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Marginal excision of a 1.5-cm diameter, firm, noninflamed, tender solid soft tissue subcutaneous, suprafascial mass, subjacent and fixed to a mature scar, was performed under local anesthesia. Photographs of the gross specimen were not taken. Histopathologic examination revealed a well-circumscribed lesion with solid aggregates of glomus cells surrounding capillary-sized vessels in a myxoid or hyalinized stroma (Fig. 1). Closer magnification showed areas of short spindle cells with hyperchromatic nuclei and prominent mitotic figures (Fig. 2). The lesion was identified as a glomangiosarcoma and was noted to extend to the margins of the resected specimen. Reexcision of the biopsy site with 1.5-cm peripheral margins was performed. Pathologic examination demonstrated no residual tumor and negative margins. X-rays or other imaging studies were not performed at the time of excision or at subsequent visits as the patient was clinically disease free. There was no evidence of recurrence at 10-year follow-up.

DISCUSSION

Review of the literature reveals few reported cases of glomangiosarcoma of the skin and soft tissue. Although clinically and histologically atypical glomus tumors have been previously reported, the designation of glomangiosarcoma or malignant glomus tumor is generally reserved for lesions with features suggesting an increased risk of metastasis. The largest existing case series of 52 patients found a significantly increased 5-year risk of metastasis in tumors with a deep location, size larger than 2 cm, and atypical mitotic figures.[4] Based on these findings, the following diagnostic criteria for malignancy have been established: size greater than 2 cm, subfascial location, atypical mitotic figures, moderate to high nuclear grade, and 5 or more mitotic figures per 50 high powered fields.[1,4]

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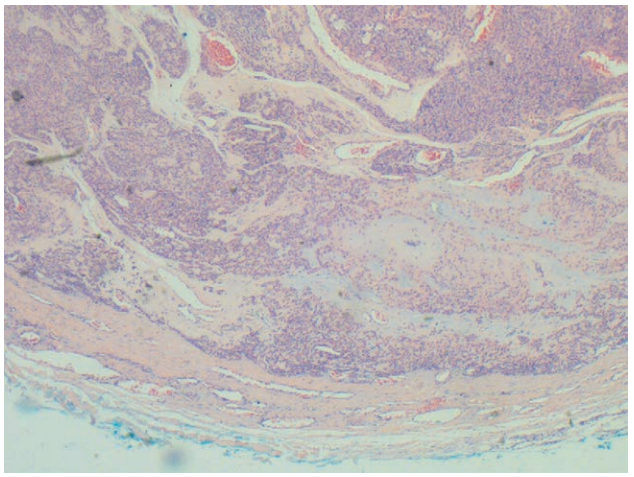

Fig. 1. Well-circumscribed lesion with solid aggregates of glomus cells around capillary-sized vessels in a myxoid or hyalinized stroma. Glomus cells are round and regularly shaped with sharply punched out nuclei (hematoxylin and eosin stain; 40×).

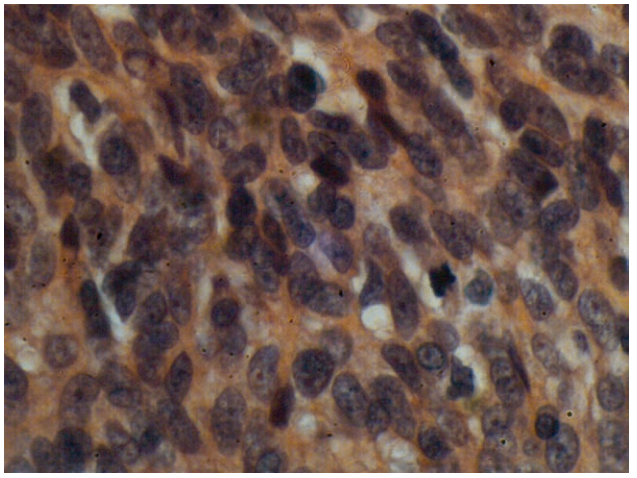

Fig. 2. Sarcomatous area consisting of short spindle cells with hyperchromatic nuclei and prominent mitotic figures (hematoxylin and eosin stain; 400×).

Malignant glomus tumors were initially divided into 3 distinct categories based on their histologic characteristics.[5] Locally infiltrative glomus tumors tend to be larger and more deeply located than their benign counterparts. They may also show a more extensive solid growth pattern or a cavernous glomangioma pattern. Glomangiosarcomas arising from a benign glomus tumor consist of typical glomus tumor merging histologically into cytologically malignant areas containing mitotic figures and nuclear pleomorphism. These lesions are thought to arise from malignant degeneration of an existing glomus tumor. Glomangiosarcomas arising de novo represent tumors with malignant cytologic features and increased mitotic rate that lack any histologic findings consistent with origin from a benign glomus tumor.[5]

In the largest case series of atypical glomus tumors, both malignant and benign tumors were further classified by their gross and histologic features. The term malignant glomus tumor was suggested for lesions with features suggesting an increased risk of metastasis: deep location and size more than 2 cm, presence of atypical mitotic figures, or a combination of moderate to high nuclear grade and mitotic activity. Tumors with atypical features such as large size, deep location, or superficial location with high mitotic activity that did not meet minimum criteria for malignancy were deemed glomus tumor of uncertain malignant potential. The term symplastic glomus tumor was proposed for lesions displaying nuclear atypia as their only unusual feature. A final group of atypical glomus tumors with angiomatosis and a strong glomus component were designated glomangiomatosis.[4]

Histologic and immunohistochemical examination are essential in establishing the diagnosis of glomangiosarcoma as the differential diagnosis may be broad based on the clinical presentation alone. In the skin and soft tissue, these lesions must be differentiated from other cutaneous round cell tumors such as Merkel cell carcinoma, eccrine spiradenoma, and melanoma as well as leiomyosarcoma and hemangiopericytoma.[1,3] Histologically, glomangiosarcomas have sheets of uniform, round to oval cells with eosinophilic cytoplasm, numerous vascular spaces, and cellular pleomorphism associated with frequent mitotic figures, features not present in benign glomus tumors.[4,5] On immunohistologic staining, glomangiosarcomas express many of the same antigens as their benign counterparts, including positive staining for vimentin, smooth muscle actin, and muscle-specific actin. However, malignant tumors stain more intensely for vimentin than their benign counterparts, which stain more intensely for actin and myosin.[6] Previous studies have also shown alterations in the expression of p53 and Bcl-2 when compared with benign glomus tumors.[7,8]

Wide local excision with negative surgical margins remains the mainstay of treatment for malignant glomus tumors, although the use of Mohs micrographic surgery has been previously described.[3,9] Adjuvant chemotherapy is not currently recommended for primary glomangiosarcoma.[9] Given the rarity of the tumor, sparse literature exists regarding the management of recurrent and metastatic disease. In the largest case series to date, 38% of patients with a pathologic diagnosis of malignant glomus tumor developed metastatic disease and 3 quarters of those died within 3 years.[4] Despite this seemingly dismal prognosis, local recurrence with simultaneous bilateral pulmonary metastases has been successfully treated with re-excision and platinum-based chemotherapy in 1 previous case.[1]

CONCLUSION

Excision of presumably benign skin and subcutaneous lesions are perhaps the most common surgical procedures performed today. This case serves as a reminder that essentially all "benign-appearing" lesions requiring excision should be examined histologically.

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