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Glomangiosarcoma in the shoulder of a 51-year-old man

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ABSTRACT Glomangiosarcoma is a very rare type of soft tissue neoplasm. Here we report such a case in the shoulder of a 51-year-old man.

Case report

A 51-year-old white Hispanic man presented with a pruritic lesion on the back of his left shoulder. He first noticed it about two months ago. The patient was otherwise well with no constitutional symptoms. His past medical history was unremarkable. Family history was significant for having older siblings with malignancy, one with multiple myeloma and another with thyroid cancer. Physical examination showed an overweight middle aged male with multiple small freckles, skin tags and nevi over his trunk and neck. A 1 cm pink, firm, rubbery, non-tender nodule was present on the back of left shoulder. There was no lymphadenopathy. A shave biopsy of the left shoulder lesion was performed.

Histopathological findings

Shave biopsy showed a cellular neoplasm with a multinodular growth pattern in the dermis (Figure 1A). The neoplasm was very cellular and consisted of round, ovoid and spindle cells with moderate nuclear atypia and frequent mitotic figures (up to 6 per 10 HPF) (Figure 1B–C). Focal areas showing histopathological features of glomangioma such as perivascular arrangement were noted (Figure 1D). Immunohistochemistry showed that the neoplastic cells were positive for smooth muscle actin (SMA), smooth muscle myosin heavy chain (SMMH), vimentin, and focally positive for caldesmon and negative for CD31, CD34, AE1/3, desmin, HMB45, Melan-A and S100. The neoplastic cells stained focally for p53. Based on these histological and immunohistochemical findings, a diagnosis of glomangiosarcoma was rendered.

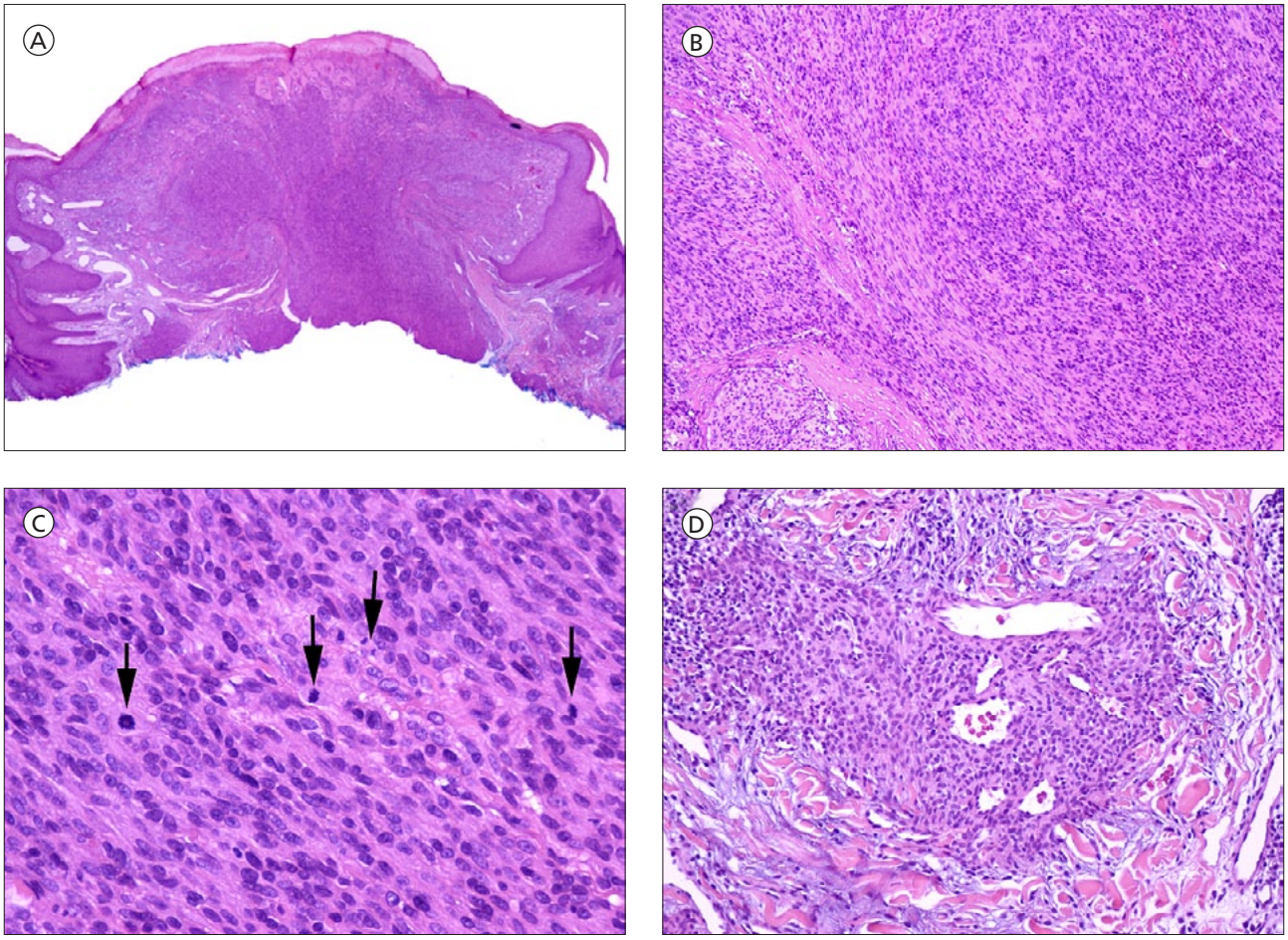

Figure 1. Shave biopsy shows a cellular multinodular neoplasm with surface ulceration (A) (H&E (hematoxylin and eosin, 2X). The neoplasm consists of sheets of ovoid-spindle cells (B) (H&E, 10X). The neoplastic cells are ovoid, spindle and epithelioid with frequent mitotic figures (C) (arrows indicating mitotic figures, H&E, 40X). Focal glomangioma-like areas are noted in the peripheral of the lesion (D) (H&E, 20X). [Copyright: ©2012 Rishi et al.]

Imaging studies, including whole body PET scan revealed no evidence of metastatic lesions. Subsequently wide excision was performed and showed a 0.9 cm tumor involving the dermis and subcutis. Cytogenetic analysis showed no chromosomal abnormality.

Discussion

Although glomangioma and glomus tumor and glomangiosarcoma and malignant glomus tumor have been used interchangeably in the literature, we recommend the use of glomangioma and glomangisarcoma for benign and malignant glomus neoplasms, respectively, simply for consistency of terminology. For example, we do not call leiomyosarcoma malignant leiomyoma or angiosarcoma malignant hemangioma.

Glomangiosarcoma is very rare. The first case was reported by Lumley and Stansfeld in 1972, who used the term of malignant glomus tumor to described a lesion located deep to the Achilles tendon in a 24-year-old female with severe and persistent right distal lower extremity pain that finally required an above-knee amputation [1]. The first case series of glomangiosarcoma was reported by Gould et al in 1990. The authors presented six cases of locally aggressive glomus neoplasm and proposed a three-tier classification for these lesions, namely, locally infiltrative glomus tumor, glomangiosarcoma arising in a benign glomus tumor and de novo glomangiosarcoma [2]. Aiba et al suggested using the term glomangiosarcoma when a sarcomatous component arises in the background of a pre-existing glomangioma [3]. Rodríguez-Justo et al reported a clinico-pathological review of 19 cases of glomangiosarcoma and de novo glomangiosarcoma, including their case of glomangiosarcoma arising in benign glomus tumor [4]. Kreutz et al reported the first metastatic glomangiosarcoma in 1987 in a 33-year-old male with a large lesion found superficially on the thigh with metastasis to maxilla [5]. Since then, a few more cases with metasta-

sis have been reported and outcome in most of them was lethal [5-7]. In 2001, Folpe et al, after analysis of 52 cases of atypical and malignant glomus tumors, proposed an empirical classification and three diagnostic criteria for malignancy in glomus neoplasm, namely, (1) deep location and a size of more than 2 cm; or (2) presence of atypical mitotic figures; or (3) moderate to high nuclear atypia with five or more mitotic figures/50 HPF [8]. Based on the proposed criteria, 32 out of the 52 cases were classified by Folpe et al as malignant glomus tumor, while the rest either as symplastic glomus tumor, glomus tumor of uncertain malignant potential, or glomangimatosis. This empirical classification as well as the proposed diagnostic criteria, are adopted in the current WHO publication on tumors of soft tissue and bone [9], although further validation and refinement are clearly needed.

In the case we presented here, although the lesion was small (0.9 cm in size) and superficially located, it was cellular with ovoid-spindle morphology, moderate nuclear atypia, and many mitotic figures. In our opinion, these features are those of glomangiosarcoma. However, we understand that observation of nuclear atypia is subjective. One may argue that the current lesion showed only mild nuclear atypia and thus, according to the criteria proposed by Folpe et al, it would be classified as glomus tumor of uncertain malignant potential. Actually the latter is not a diagnostic entity, but rather a descriptive term used for those lesions when one is not sure if it is gloomangiosarcoma or not. In the present case, even in the absence of significant nuclear atypia, we believe the histopathologic findings, namely, high cellularity, ovoid-spindle morphology and increased mitotic activity, are best interpreted as glomangiosarcoma.

Microscopically glomangiosarcoma varies depending on the degree of differentiation. Well differentiated ones have histology similar to glomangioma except that there are frequent mitotic figures and presence of cytological atypia. Poorly differentiated ones resemble high-grade sarcoma and a definite diagnosis based solely on morphology can be challenging. Histological differential diagnoses include epithelial, myoepithelial, melanocytic and smooth muscle tumors. Immunohistochemistry can be helpful in differential diagnosis. Immunohistochemically, glomangiosarcoma is positive for vimentin, SMA, SMMH, and collagen IV and negative for epithelial, vascular, melanocytic and neural markers. Although myoepithelial lesions express SMA, they are also positive for other markers, such as epithelial markers and S100, which are negative in glomangiosarcoma. Leiomyosarcoma expresses desmin and caldesmon, which are usually negative in glomangiosarcoma.

The treatment for glomangiosarcoma is complete surgical excision. No radiation or chemotherapy is recommended at this time for primary, recurrent or metastatic disease, although their use had been reported by several authors [8,9].

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