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High-Grade Undifferentiated Pleomorphic Sarcoma Masquerading as Acquired Lymphangioma Circumscriptum

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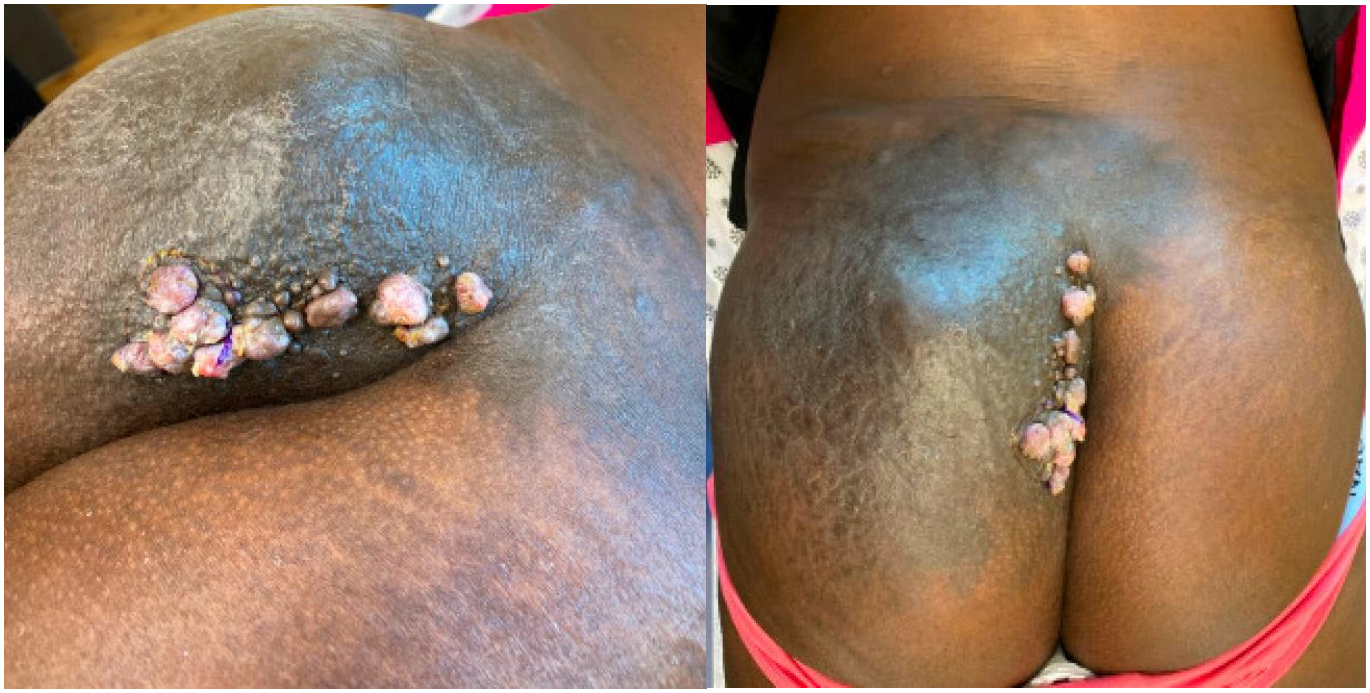

A previously healthy woman in her 40s presented to the dermatology clinic with a 1-year history of a tender, draining, enlarging mass on the left buttock. Physical examination revealed an extensively indurated hyperpigmented nodule with numerous overlying clustered exophytic verrucous papules on the L medial buttock (see figure 1). Initial skin biopsy of verrucous lesions was notable for lymphangioma with overlying condyloma, D2-40 positive. MRI abdomen and pelvis with contrast demonstrated a left gluteal enhancing subcutaneous soft tissue mass, 10.3 x 8.2 x 8.8 cm in size, overlying and involving the left gluteus maximus, along with satellite nodules present in the medial gluteus maximus and paraspinal muscles, with bilateral pelvic and inguinal adenopathy. Excisional deep tissue biopsy revealed a diagnosis of a high-grade, multifocal spindle and undifferentiated pleomorphic sarcoma. The patient subsequently underwent 4 cycles of neoadjuvant chemotherapy AIM

(doxorubicin, iphosphamide, MESNA), followed by radical resection of the left gluteal region, with positive margins, followed by repeat surgical resection and reconstruction with left-sided latissimus dorsi myocutaneous free flap. The patient has now completed radiation therapy.

Soft tissue sarcoma is a heterogeneous group of rare malignancies of mesenchymal origin, that represents 0.8% of all adult cancers. Prognostic factors associated with poor outcome include high histologic grade, large tumor size, presence of metastases, and unresectability[1]. Herein, we report a novel case of high-grade soft-tissue undifferentiated pleomorphic sarcoma, masquerading as acquired lymphangioma circumscriptum (ALC), as noted on initial skin biopsy of grouped flesh-colored verrucous papules overlying an indurated tender mass. ALC is a lymphatic malformation occurring as sequela to any process which interrupts previously normal lymphatic drainage. As such, subcutaneous lymphatic cisterns are unable to drain completely into the general lymphatic system, resulting in lymphatic dilation and subsequent development of vesicular and papular lesions on the trunk, extremities, and anogenital areas.[2] ALC can mimic common infectious growths including molluscum contagiosum and condyloma accuminata, and accordingly be mismanaged. Etiologic causes of ALC including malignancy, and has previously been reported in the setting of cervical carcinoma, vulvar carcinoma, endometrial carcinoma, Hodgkin lymphoma, rhabdomyosarcoma, rectal carcinoma, melanoma, and high-grade penile dysplasia[2,3,4]. Other etiologic causes can include surgery, trauma, radiation therapy, chronic inflammatory conditions including hidradenitis suppurativa, and inflammatory bowel disease.

To date, acquired lymphangioma circumscriptum has not been reported in the setting of underlying high-grade pleomorphic soft tissue sarcoma. As such, clinical presence of grouped frogspawn-like or verrucous papules, in an area of tender induration or lymphedema, should prompt a work-up to exclude an underlying malignant etiology as new onset ALC may be masquerading an underlying malignancy.

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