Imagens em Oncologia

Síndrome de Stewart-Treves

Stewart-Treves Syndrome

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Resumo

Objetivo: Apresentação de caso raro e revisão das caraterísticas dos angiossarcomas da mama secundários a linfedema: síndrome de Stewart-Treves.

Métodos: Imagens de TC, RM e fotografia de recessão cirúrgica são apresentadas juntamente com discussão clínica do caso e breve descrição da patologia.

Resultados: Diagnóstico e cirurgia precoces oferecem possibilidade de melhorar o mau prognóstico deste tumor. Os sinais clínicos iniciais são variáveis incluindo máculas violáceas, nódulos cutâneos, massas subcutâneas ou escaras crónicas. Estudos imagiológicos como a RM auxiliam no diagnóstico e avaliação da extensão local da doença.

Conclusão: Linfedema crónico em que sinais clínicos, imagens de TC/RM demonstram nódulos ou espessamentos cutâneos/subcutâneos fora das áreas irradiadas num membro edematoso após mastectomia e radioterapia sugerem síndrome de Stewart-Treves.

Palavras-chave: Stewart-Treves, angiossarcoma secundário, angiossarcoma mama.

Abstract

***Goals**: To present and review the appearance of a rare case of lymphoedema secondary breast angiosarcoma (Stewart-Treves syndrome).*

***Methods**: Diagnostic computed tomography (CT) and magnetic resonance (MR) images and a specimen photograph are presented, with a clinical case discussion and a brief pathology description.*

***Outcomes**: Early diagnosis and prompt surgery are crucial to improve the poor prognosis of this tumor. Early signs of the condition vary, including purplish patches that develop into skin nodules, a mass under the skin, or a poorly healing eschar. Imagiological studies, mainly MR, aids in the diagnosis and disease extension assessment.*

***Conclusion:** Chronic lymphoedema in which clinical signs and CT/MR scans demonstrate nodules or enhanced skin thickening outside a radiated field in an oedematous limb after mastectomy and radiotherapy suggest the Stewart-Treves syndrome.*

***Key words:** Stewart-Tyreves, secondary angiosarcoma, breast angiosarcoma.*

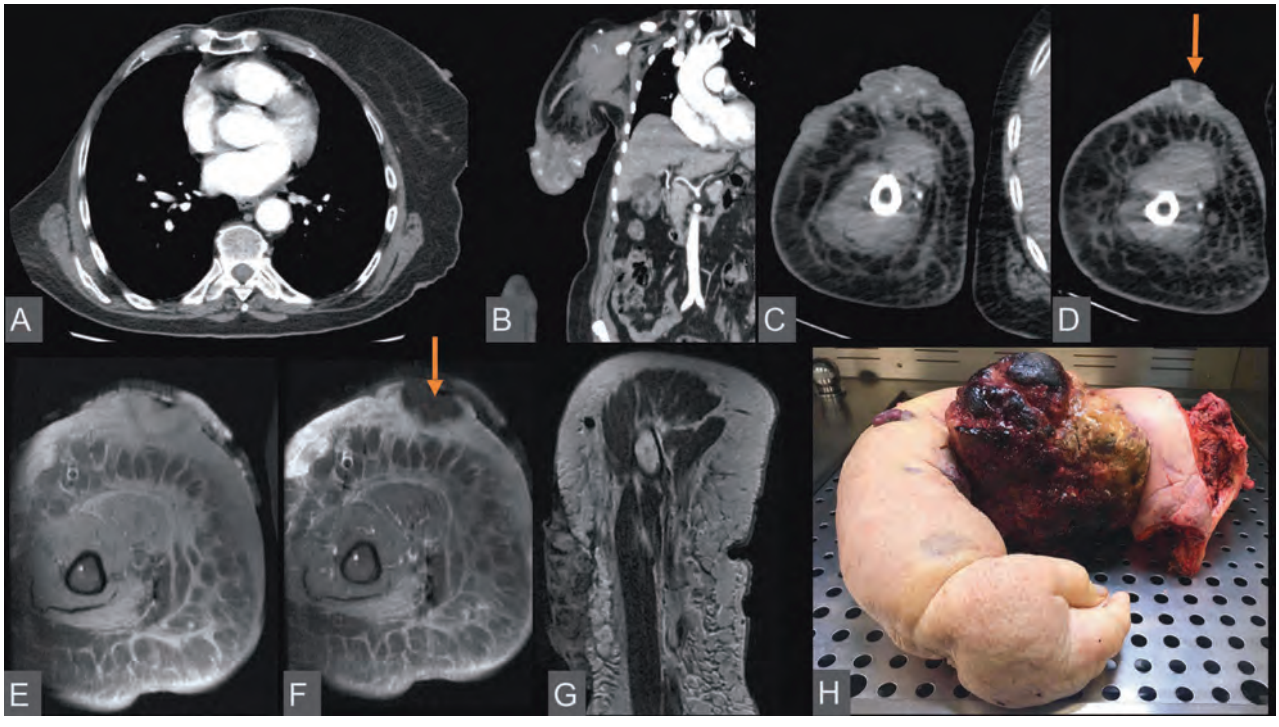

Figure 1. 62-year-old-woman with secondary breast angiosarcoma associated with lymphoedema.

Explicative text

We present the case of a 62-year-old-woman with high-grade secondary breast angiosarcoma associated with lymphoedema: Stewart-Treves Syndrome (Figure 1).

The patient had undergone a right mastectomy 6 years before due to an invasive ductal breast carcinoma. Follow-up thoracoabdominal computed tomography (CT) evaluation (Figure 1-A) performed 1 year after surgery showed no evidence of metastases; however, right arm lymphoedema had been present since the surgery. Now the patient presents purplish discoloration with multiple palpable skin nodules. Coronal (Figure 1-B) and axial (Figure 1-C,D) CT images show marked skin thickening with enhanced skin nodules and an area of ulceration (arrow). Axial T1 FS (Figure 1-E), sagittal T2 (Figure 1-G), and axial post-gadolinium (Figure 1-F) magnetic resonance (MR) images reveal severe right arm soft tissue oedema with marked skin nodular thickening and enhancement with non-enhancing ulcerated components. The gross specimen (Figure 1-H) obtained 3 weeks after the MR study demonstrates the rapidly growing partially ulcerated mass on the skin.

Secondary angiosarcoma of the breast is a rare tumor known to appear in older women following radiotherapy for breast cancer.[1] When associated with chronic lymphoedema and outside a radiated field, secondary angiosarcoma in an oedematous limb after mastectomy and radiotherapy is referred to as Stewart-Treves syndrome.[2] The underlying cause of the condition is poorly understood. Early signs of the condition vary and may include purplish patches that develop into skin nodules, a mass under the skin, or a poorly healing eschar with recurrent bleeding and oozing.[3] Its sometimes harmless appearance may contribute to a delay and neglect by both patients and physicians.[4] Imagiological studies, mainly MR imaging, aids in the diagnosis and disease extension assessment.[5,6]

Wide local excision is often reported, but whether or not all irradiated skin is removed is not specified. It is clear that aggressive treatment is necessary; however, the role of neoadjuvant, and adjuvant chemotherapy and radiotherapy remains ill-defined.[7]

This is a rare presentation as increased use of breast conservation therapy and sentinel lymph node sampling has lowered the incidence of treatment-related lymphoedema. A chronic lymphoedema in which CT or MRimaging scans demonstrate nodules or enhanced skin thickening outside a radiated field in an oedematous limb after mastectomy and radiotherapy suggest Stewart-Treves syndrome.

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This article does not contain any new studies with human or animal subjects performed by any of the authors. Further details on radiotherapy sessions (length, duration and size of the beam therapy) are not included in this manuscript due to lack of information at the stage of writing process.

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