Case 25

Panties mass

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Patient: A 67-year-old Thai woman from Nakhonratchasrima

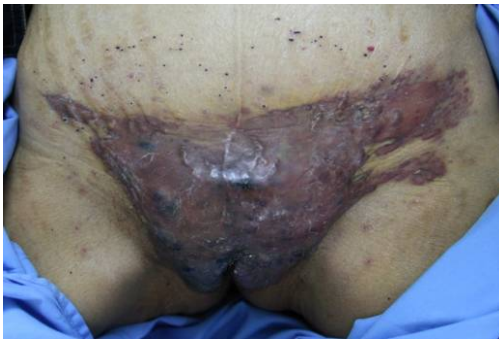
Chief complaint: Enlarging mass on pubic area for 2 months

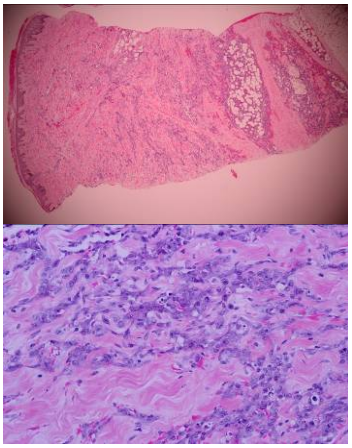
Present illness:

She was diagnosed with endometrium carcinoma stage IC. She underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy, followed by pelvic radiation in 2004. The total radiation dose was 50 Gray (Gy).

In 2014, she presented with a gradually enlarging mass, at the pubic area extending to mons pubis for the past 2 months. It also was painful on palpation and bled easily. The affected site was within the field of prior radiation.

Past history: as above





Skin examination:

- A large well-defined firm violaceous plaque, 12x25 cm in size, and several bluish nodules on mons pubis and labia majora, in the shape of a pair of panties
- Pelvic examination was not performed due to the obstructing mass

Histopathology: (S14-3350, pubic area)

- Cords and strands of atypical cells, some forming anastomosing vascular lumens (dissecting collagen
- Atypical cell showing nuclear pleomorphic, few in mitosis in the dermis and subcutaneous tissue
- Immunostains:
  - Positive for CD31
  - Negative for CD68, AE1/AE3, HHV-8 and VEGFR-3

Diagnosis: Radiation-associated angiosarcoma (RAAS)

Treatment:

- Paclitaxel at 70 mg/m2/dose every 2 week

Discussion:

Angiosarcoma (AS) is a rare aggressive malignant mesenchymal tumour. The majority of patients develop angiosarcoma at advanced age on sun-exposed skin, with predilection for the scalp. The known risk factors for developing secondary angiosarcoma include chronic lymphedema and prior radiation. The frequency of radiation-associated angiosarcoma (RAAS) is 17-25% of all angiosarcoma cases.[1] The diagnostic criteria for radiation-induced sarcomas include previous history of radiotherapy with a latency period of more than 3-4 years, development of sarcoma within a previously irradiated field or in the tissues adjacent to the field, and histologic confirmation.[2] The most frequent clinical setting is the treatment of breast carcinoma, predominantly after radiotherapy in breast conservative therapy, followed by gynaecological malignancies.[3]

Almost all of the patients are female. The median age at presentation was 68 years; the median radiation dose is 40-60 Gy. The latency between radiation and diagnosis of RAAS is approximately 5–6 years.[3] The clinical presentation included a large violaceous or erythematous patches, plaques, nodules, or bruise-like discolouration, overlying a painless area of induration. Most are multifocal lesions and the size varies from a few millimeters to more than 20 centimeters.[3] The histological features range from well to poorly differentiated. These lesions are poorly circumscribed, involve the dermis and sometimes subcutaneous tissues. They consist of irregular interanastomosing channels that infiltrate surrounding tissue, diffusely dissecting the dermal collagen. The atypical endothelial cells are usually multilayered; sieve-like architecture is another common feature. The immunohistochemistry shows a positive staining with antibodies against endothelial markers including CD31, CD34, FVIII-related, and von Willebrand factor.[3] CD31 is the best marker with high sensitivity and specificity.

Angiosarcoma is a very aggressive tumour. Standard therapy is surgery with complete tumour resection. However, when surgery is not possible, chemotherapy has also proved to be beneficial. Anthracyclines alone or in conjunction with iphosphamide have led to disease control after several months.[4] Over the past few years, paclitaxel has been used in advanced stage and/or metastatic AS.[5] In our patient, the large tumor could not be resected; chemotherapy was started with paclitaxel at 70 mg/m^2/dose every 2 weeks. After completing the fifth cycle of paclitaxel, the lesion was markedly decreased in size and the symptoms previously described were also completely resolved.

References:

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3. Brenn T, Fletcher CD. Radiation-associated cutaneous atypical vascular lesions and angiosarcoma: clinicopathologic analysis of 42 cases. Am J Surg Pathol. 2005; 29(8): 983-96.
4. Scott MT, Portnow LH, Morris CG, et al. Radiation therapy for angiosarcoma: the 35-year University of Florida experience. Am J Clin Oncol. 2013; 36(2): 174-80.
5. Perez-Ruiz E, Ribelles N, Sanchez-Munoz A, Roman A, Marquez A. Response to paclitaxel in a radiotherapy-induced breast angiosarcoma. Acta Oncol. 2009; 48(7): 1078-9.