CASE REPORT

Lymphangiosarcoma of Mons Pubis Following Radiotherapy for Carcinoma Cervix

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Background

Post-irradiation lymphangiosarcoma/angiosarcomas are rare soft tissue sarcomas which pose considerable diagnostic dilemma with atypical vascular lesion, a premalignant vascular tumor that commonly follows radiotherapy. Here we present a case of a 42 year female with lymphangiosarcoma of mons pubis occurring after 15 years following radiotherapy for carcinoma cervix stage IIA. To the best of our knowledge this is the first case of post-irradiation lymphangiosarcoma of mons pubis. The case is being documented for its rarity.

Introduction

Common post-irradiation vascular tumors are post-radiation angiosarcoma, atypical vascular lesions, lymphangiomatosis, and lymphangiosarcoma. While atypical vascular lesions and lymphangiomatosis behave in a benign fashion, post-radiation angiosarcoma and lymphangiosarcoma have a

significantly high mortality. Post-radiation angiosarcoma/lymphangiosarcoma usually occurs after an interval of 5–11 years following irradiation for carcinoma of the breast, ovary, uterus, and cervix [1]. This condition is often difficult to diagnose due to its rarity, relatively bland appearance, difficulty in differentiation from radiation-induced changes in the skin, and close mimickers like atypical vascular proliferation and lymphangiomatosis. Meticulous morphological assessment remains the cornerstone to differentiate between angiosarcoma and lymphangiosarcoma from their benign mimickers like atypical vascular proliferation and lymphangiomatosis. It is difficult to differentiate malignant vascular tumors of blood vessel and lymph vessel origin solely on morphological grounds. However, recently, D2-40, a new selective monoclonal marker of lymphatic endothelium, has gained importance in differentiating angiosarcoma from lymphangiosarcoma [2]. Lymphangiosarcoma of the mons pubis following radiotherapy is an extremely uncommon entity. Recently, we have seen such a case where the patient had received radiotherapy for carcinoma cervix 15 years prior to the present illness. To the best of our knowledge, this is the first case of post-irradiation lymphangiosarcoma of the mons pubis in the English literature, and hence it is being reported for its rarity.

Case Report

The index case is of a 42-year-old female who underwent Wertheim's hysterectomy followed by radiotherapy for

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Fig. 1 Tumor showing ramifying network of vascular channels infiltrating dermal collagen (A), endothelial cells showing cellular atypia, hyperchromatic nuclei and mitoses

carcinoma cervix stage IIA. A total of 50 Gray was given in 28 fractions, 5 fractions per week. After that, the patient received intravaginal brachytherapy (IVBT). She was kept on regular follow-up. After 15 years of uneventful follow-up, she presented with complains of an indurated nodular growth over the mons pubis. There was no history of lymphedema or any preceding traumatic event in this area. There was no associated lymphadenopathy. The routine laboratory examination was unremarkable except for

moderate anemia. Abdominal and trans-vaginal ultrasound revealed no evidence of any recurrence. Chest X-ray was normal. An incisional biopsy from the lesional area was sent for histopathologic examination. However, this biopsy was inconclusive as it included a very small part of the lesional area. Following the biopsy, a wide excision of the lesion with adequate margins was done and sent for histopathologic evaluation. The histopathologic examination revealed a diffusely infiltrating vasoformative tumor in the

Fig. 2 Tumor cells are positive for CD34(A), CD31(B), D2-40(C) and MIB-1 labelling index is high (D)

dermis and subcutis. The tumor showed a ramifying network of vascular channels lined by endothelial cells with cellular atypia, hyperchromatic nuclei, and the presence of mitoses. At places, there was multilayering of the endothelium. Mature lymphocytes were seen within and around some of these vascular channels. At the periphery of the lesion, the channels were dissecting the collagenous plane (Fig. 1). There was mild dermal sclerosis. Immunohistochemically, these cells were positive for CD34, CD31 and D2-40. The proliferation index (MIB-1 immunoreactivity) was high (10–15 %) (Fig. 2). On the basis of the above histomorphology, a diagnosis of lymphangiosarcoma was made.

Discussion

Over the last few years, there has been significant improvement in the treatment of various cancers due to a multimodality approach which has led to increased patient survival. As the patients are living longer, they encounter more treatment-related complications among which radiation-induced secondary malignancy (RISM) is a serious problem. Criteria for RISM include the following:

1. There should be a history of previous irradiation.
2. The new malignancy should arise within the previously irradiated field.
3. Histology of the second neoplasm should be different from the first.
4. There should be a long latent period between the tumors(>5 years).

Of the radiation-induced secondary malignancies, post-radiation vascular tumors deserve special mention because of their varied clinical and histologic spectrum. Also, it is difficult to differentiate tumors of endothelial and lymphatic origin purely on morphological grounds. However, a newly described antibody against lymphatic endothelium D2-40 is helpful to resolve the issue in such cases.

According to the literature, there are two cases of post-irradiation angiosarcoma of the mons pubis that occurred following irradiation for carcinoma cervix [3]. To the best of our knowledge, the index case is the first bona fide case of post-irradiation lymphangiosarcoma in this region.

Conclusion

Post-irradiation lymphangiosarcoma, though rare, can pose a diagnostic challenge as it may simulate angiosarcoma or atypical vascular lesion. An immunohistochemical panel including D2-40 is crucial to establish the correct diagnosis.

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