One Case of Stewart-Treves Syndrome Successfully Treated at Two Years by Chemotherapy and Radiation Therapy in a 73-year-old Woman

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Lymphangiosarcoma of the upper extremity developed on lymphedema of the arm (usually after treatment for breast cancer) is a relatively rare tumour first described by Stewart & Treves in 1948 (1), although Lowenstein had already reported a case in 1906 (2). We report here a new case in a 73-year-old patient initially treated for carcinoma of the right breast 10 years before.

Case report. In July 1984, the patient who was then 63-year-old was treated by mastectomy and axillary lymphadenopathy for a 4-cm tumour of her right breast associated with massive axillary involvement. She received subsequently one course of CMF chemotherapy followed by radiotherapy up to a total dose of 50 Gy to the chest wall and the axillary node area and 45 Gy to the suspra-clavicular node area. Nine further courses of CMF chemotherapy were given after completion of radiotherapy.

Lymphedema of the arm started developing in August 1991. In July 1993, the difference in circumference between the two arms was 3.5 cm at arm level, 6 cm between the two forearms, and 4 cm at the wrists. No locap relapse or distant metastases were found at that time.

In July/August 1994, she developed bluish and purple lesions in her right forearm which could evoke lesions of ecchymosis but did not follow any trauma and subsequently tended to grow, coalesce and spread to most of the upper side of the forearm. Biopsy of the lesison showed angiosarcoma (with strong immunoreactivity for vimentin and factor VIII-related antigens, while keratin and EMA were negative).

Reassessment of the extension of the disease with magnetic resonance locally, thoracic and abdominal CT-scans and radioisotope scanning, found mediastinal pathological nodes and a stenosis of the superior bronchus of the inferior lobe. Biopsies at that site were in favour of a metastasis from the carcinoma of the breast and not from the lymphangiosarcoma.

Considering these metastases, radical surgery on the arm was excluded and the patient was treated with two courses of doxorubicin, ifosfamide and bleomycin, followed with 39 Gy in 13 fractions with photons on the whole forearm, and a boost of 16 Gy in 8 fractions with electrons on the cutaneal lesions, and finally with 6 more course of chemotherapy (the last one without bleomycin). Complete remission of the lesions of the forearm was obtained, whereas the mediastinal node had shrunk and the stenosis of the bronchus persisted but with negative biopsies. It is to be noticed, that the lymphedema of the upper limb had retroceded markedly in forearm and wrist.

In April 1996, there were no signs of relapse related to the lymphangiosarcoma of the upper arm, no mediastinal pathological node visible on CT-scan, but biopsies of the bronchus stenosis were found positive again (adenocarcinoma compatible with a metastasis from the carcinoma of the breast). No oher distant metastasis was found. Localized radiotherapy was delivered on the bronchus involved and corresponding part of the mediastinum with excellent tolerance, and the patient was put on tamoxifen.

Follow-up is only 2 years for the patient's Stewart-Treves sarcoma, but this 73-year-old woman does not show any sign of local relapse or distant metastases from her hemangiosarcoma of the upper-limb at present.

Discussion. Stewart-Treves syndrome arising in chronic or congenital lymphedema is uncommon. When developing on postmastectomy lymphedema, it usually occurs approximately 10 years afer loco-regional treatment with surgery and radiotherapy for a carcinoma of the breast (3) with an incidence of 0.07 (4) to 0.45% (5). It often presents with bluish or purple macular or papular lesions turning later into plaques at a more advanced stage, and must be distinguished from Kaposi sarcoma (context is different), metastases from the primary carcinoma of the breast, or even some malignant lymphomas. Immunohistochemistry using anti-vimentin, anti-factor VIII, ULEX-Europaeus Agglutinin I (positive), and anti-keratin KL1, anti-EMA and anti-desmosome (negative) antisera will confirm the diagnosis (6–8).

Radical surgery is the most commonly recommended treatment (3, 7, 9–11) as the best chance for cure, since wide excision is usually followed by loco-regional relapse and distant metastases (11). Nevertheless, some authors have obtained interesting results with radiation therapy (11–15), chemotherapy (essentially with 5-FU, methotrexate, or various combinations of vincristine, actinomycin D, cyclophosphamide, doxorubicin and/or dacarbazine with or without vincristine (11, 16), and more recently bleomycin alone (17) or actinomycin D combined with radiation therapy (15)), or even with immunotherapy (18).

Despite treatment, prognosis remains dismal, with a median survival of about 2.5 years (9), local recurrence is frequent, and

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most patients die within 2 years usually from metastatic disease, mainly to the lungs, less often to liver, bones, nodes, and exceptionally to the brain (3) with a very small number of long-term survivals (13). In a review of 129 patients reported in the literature, A. H. Woodland (19) found only 11 of them still alive at 5 years, while more recently, some other authors also reported a few cases of long-term survivors (11, 15).

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