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Roukema, J.A.; Leenen, L.P.H.; Kuizinga, M.C.; Maat, B.

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Angiosarcoma of the irradiated breast: a new problem after breast conserving therapy?

J. A. ROUKEMA, L. P. H. LEBENEN, M. C. KUIZINGA AND B. MAAT

Post-irradiation angiosarcoma of the breast is an uncommon neoplasm. The first patient with this disease after breast conserving therapy was reported in 1987 and only a few more patients can be traced in the literature since that time. Two more cases are presented in this paper: both women in the seventh decade of life. The tumour became apparent five and seven years after the primary treatment of breast cancer. The possible influence of irradiation and lymphoedema in inducing this malignancy is discussed.

Introduction

Angiosarcoma of the breast is a rare but highly malignant tumour of vascular origin.¹ Many angiosarcomas have been reported in postmastectomy scars and lymphoedematous extremities after radical mastectomy,² but it lasted until 1987 before the first case of angiosarcoma in the irradiated breast was published³ and a few more patients have been reported since that time.

We present the case history of two more patients.

Case reports

1. In 1982 a 60-year-old woman presented with a palpable mass in the lower inner quadrant of the right breast. An excisional biopsy was performed and pathological examination showed an infiltrating ductal carcinoma (diameter 2 cm) with clear excision margins. Axillary dissection revealed 22 lymph nodes without tumour involvement. Postoperatively, the patient received a course of whole-breast radiation therapy with a total dose of 50 Gy during five weeks. The primary site also received a 20 Gy boost in 10 fractions using 15 Mev electrons.

Follow-up was done at regular intervals. Oedema, telangiectasia and fibrotic changes of the treated breast were noted within six months after the primary treatment. In 1985 a punch biopsy of a discoloured and telangiectatic skin area of the right breast showed only lymphatic dilation, without atypical endothelium or carcinoma. The patient's condition remained stable and mammograms showed no abnormalities until October 1989. At that time, the patient had an ulcerating mass in the right breast.

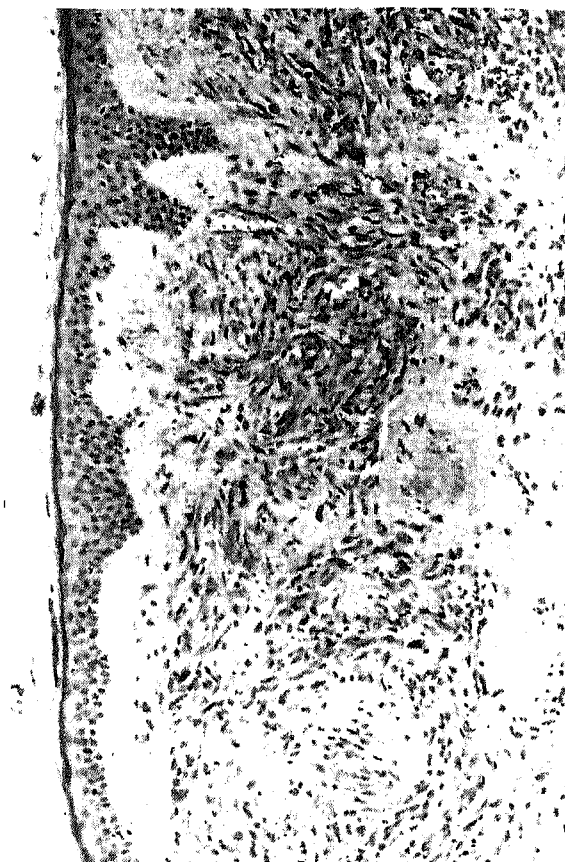


FIG. 1. Beneath the epidermis (top) solid parts of anaplastic spindle cells in addition to areas of vascular channels lined by atypical endothelial cells. H&E, original magnification 16 ×.

Advanced recurrent carcinoma was suspected and she was treated with tamoxifen 2 × 20 mg daily. After three months of treatment there was no tumour response and at last a biopsy was performed. Pathological examination revealed an angiosarcoma (Fig. 1). Simple mastectomy was performed for palliation of this angiosarcoma

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From the Departments of Surgery and Pathology of the Elisabeth Hospital and the Radiotherapeutic Institute Bernard Verbeeten, Tilburg, The Netherlands.

Correspondence to: Dr. J. A. Roukema, Department of Surgery, Elisabeth Hospital, P.O. Box 90151, 5000 LC Tilburg, The Netherlands.

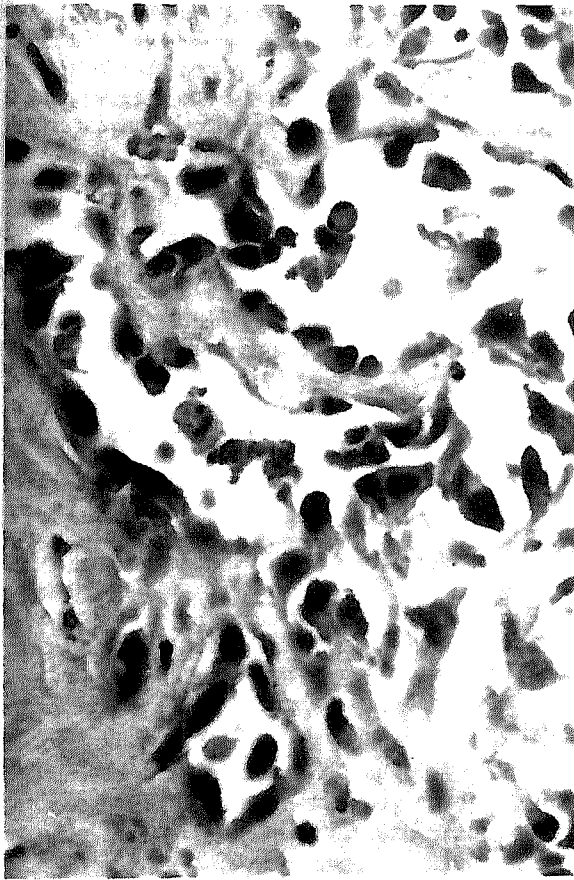


FIG. 2. Angiosarcoma showing irregular lumina lined by plump endothelial cells. H&E, original magnification 160 ×.

(diameter 9 cm). Histological examination showed free resection margins. After one year of follow-up she has no signs of recurrent disease.

2. In 1985 a 66-year-old woman underwent lumpectomy and axillary dissection because of an infiltrating ductal carcinoma (diameter 17 mm) located in the lower lateral quadrant of the right breast. No metastases were found in the axillary lymph nodes. She received a course of whole breast radiation therapy with a total dose of 50 Gy, followed by an iridium-192 interstitial implant boost of 25 Gy to the primary site. Oedema and fibrotic changes of the treated breast became apparent after six months with little progression during the next years. She remained well and had no recurrence until November 1990 when two areas of purple discoloration were noticed in the right breast. A biopsy showed a moderately differentiated angiosarcoma (Fig. 2). As a consequence mastectomy was performed. The postoperative course was uneventful.

Discussion

Angiosarcoma is a rare disease accounting for less than one per cent of all sarcomas, skin and superficial soft tissues being the most commonly affected sites.^{1, 2, 5} In 1948 Stewart and Treves described the first lymphangiosarcoma, also known as angiosarcoma, in the lymphoedematous upper extremities of women treated for breast cancer by radical mastectomy, mostly followed by radiation therapy.⁴ Since that time more than 200 patients with this condition have been reported. The development of angiosarcoma was generally attributed to chronic lymphoedema rather than to radiation-induced sarcoma. Cutaneous angiosarcomas arising on the chest wall after mastectomy and radiation therapy were described by Otis et al⁵, but these patients had no lymphoedema. In a review of the literature, Chen et al. traced 87 cases of primary angiosarcoma of the breast.² But it lasted until 1987 before others reported on the first patient with angiosarcoma of the breast after segmental mastectomy and irradiation.³ In that same year cutaneous angiosarcoma of the breast was reported in a 75-year-old woman six years after lumpectomy and radiation therapy to the remaining breast.⁶ Since that time, three more cases could be traced, two of them being elderly patients with lymphoedema of the irradiated breast.⁷⁻⁹

About 90 per cent of the vascular sarcomas complicating chronic lymphoedema are associated with post-mastectomy lymphoedema.^{2, 3} The majority of these patients have received radiation to the axilla in addition to surgery. However, some of the remaining 10 per cent occur in chronic lymphoedema or at other sites without previous radiation therapy. Early lymphoedema occurs in most irradiated breasts, especially if a complete axillary dissection has been performed. Lymphoedema that persists for more than three years is unusual.

The time interval for the development of angiosarcoma in a lymphoedematous region differs. After radical mastectomy the interval varies from four to 27 years,^{2, 3} but most sarcomas become apparent within about 10 years. In patients with congenital lymphoedema angiosarcoma generally develops after about 18 to 46 years.⁴ In the patients reported so far, angiosarcoma occurred within 2.5 to four years after primary treatment. On the basis of the experience with post-mastectomy angiosarcoma it seems reasonable to assume that it will take many years, and many patients, before it will be clear whether angiosarcoma in the irradiated breast is a rare phenomenon or a late complication of breast-conserving therapy.

Key words

angiosarcoma of the breast
post-irradiation angiosarcoma
lymphoedema

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