Haemangiosarcoma of breast

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SYNOPSIS This is a report of a haemangiosarcoma involving the breast, spleen, liver, lungs, and subcutaneous tissue. There appears to be evidence that the tumour arose in the breast with the other sites representing metastases rather than a malignant transformation of multiple haemangiomas. Histologically the primary breast tumour appeared deceptively innocuous.

Primary haemangiosarcoma of the breast is an uncommon tumour, which occurs more frequently in young women, although older age groups are not exempt (Steingaszner, Enzinger, and Taylor, 1965). These tumours have the worst prognosis of all malignant tumours of the breast (McDivitt, Steward, and Berg, 1968), despite the primary lesion’s possibly having a deceptively benign histological appearance (Mackenzie, 1961; Steingaszner et al, 1965). Metastases occur early through the blood stream and a short clinical course is common. In different series the time from the onset of symptoms to death has varied from 1-6 to 2-6 years (Mackenzie, 1961; McClanahan and Hogg, 1954; Steingaszner et al, 1965).

Several names have been applied to this tumour — benign metastasizing haemangioma, angiosarcoma, haemangiendothelioma, haemangiosarcoma (Steingaszner et al, 1965).

In all, 42 haemangiosarcomas have been reported since 1907 (Gulesserian and Lawton, 1969).

Case Report

The patient, a 57-year-old married woman without children, had a firm nodular lump removed from the right breast in January 1966. The nodule was first noticed some months previously and had been slowly enlarging since. The tumour was histologically diagnosed as a benign haemangioma. In October 1967 the patient was admitted with a recurrent mass of the right breast and a subtotal mastectomy was performed. Histological findings were again interpreted as benign haemangioma. In March 1968 the patient was readmitted with a recurrent tumour in the scar of the previous operation. A wide excision was performed. During 1969 the patient was readmitted several times with further tumours, one in the left breast, a second in the abdomen, a third in the left shoulder, and a fourth in the right side of the forehead. These tumours were considered to be metastases and the patient was treated by radiotherapy and later by chemotherapy (nitrogen mustard 5-3 mg x 4). In January 1970 radioactive gold and Indium scanning was performed, but no signs of hepatic involvement could be detected.

The patient’s general condition continued to deteriorate and she was readmitted in April 1970 because of abdominal pain.

She was severely anaemic (Hb 6-4 g%), and presented with signs of peritoneal irritation. An intra-abdominal haemorrhage due to rupture of a vascular tumour was suspected. In spite of repeated blood transfusions and other supportive treatment, the patient died.

Necropsy Findings

On postmortem examination the following pertinent changes were observed. The right breast area showed a tumour mass with a violet haematoxylike discolouration. On section it was about 12 cm at its greatest diameter, had ill-defined, irregular borders and a spongy, haemorrhagic appearance with vascular spaces. The tumour was adherent to the skin, but not to the fascia of the pectoral muscles. The liver was enlarged weighing 1,800 g. Beneath the capsule there were scattered multiple minute reddish-black vesicles. Sections showed a 'Swiss cheese' appearance with many vascular spaces of various sizes (Fig. 1). The spleen weighed 520 g and its external surface and sections disclosed vascular spaces similar to those in the breast and...
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Fig. 1  Liver section showing 'Swiss-cheese' appearance of the vascular tumour.

Fig. 2  Area of angiomatous breast tumour of benign histological appearance (haematoxylin and eosin × 40).

Fig. 3  Part of the breast tumour showing anastomosing vascular channels (haematoxylin and eosin × 100).

Fig. 4  Breast tumour with papillary growth into vascular spaces (haematoxylin and eosin × 100).
liver. Some of those had ruptured into the abdominal cavity, where about 3 l of a haemorrhagic fluid and a few blood clots were present. A few small similar metastases were also found in the lungs.

Histological examination of the first breast biopsy showed a tumour composed of blood spaces lined by benign-appearing endothelial cells (Fig. 2). A few anastomosing channels were lined by endothelial cells with somewhat larger and more hyperchromatic nuclei (Fig. 3). In the necropsy material atypical tumour cells had proliferated with mitotic activity and papillary projections narrowing the lumen of the vascular spaces (Fig. 4), while some foci still retained the appearance of a benign haemangioma. The cells were confined within a fine reticulin network. The final diagnosis was a typical haemangiosarcoma.

Discussion

Proliferating angiomatous lesions are very common, but it is doubtful if they are true tumours or vascular malformations. Some authors are reluctant to admit the existence of malignant angioblastic neoplasms, preferring to consider them as vasoformative variants of mesenchymoma (Tasker, 1958). Others believe that in the breast these lesions arise in perilobular angiomas (McDivitt et al., 1968). The behaviour of these lesions is variable. Haemangiosarcoma of bone runs a protracted clinical course and the patients have a long survival period after operation (Hartmann and Stewart, 1962). Haemangiosarcoma of the breast is a highly malignant tumour (Evans, 1968) producing a fatal outcome regardless of the mode of therapy. This tumour is more frequent in the reproductive period of life. This fact, and the relatively frequent association with pregnancy (Batchelor, 1958/59; Enticknap, 1946; McClanahan and Hogg, 1954; Tibbs, 1953), suggests local hormonal stimulation leading to malignant transformation of the vascular endothelium (Shore, 1957). A history of trauma preceding the development of the tumour has been described in several cases (McClanahan and Hogg, 1954; Patrick, Jarvie, and Miln, 1957; Steingaszner et al., 1965) but no definite relation has ever been established. Radioactive materials like Thorotrast are known to induce neoplastic changes, among them haemangiosarcoma (Casper, 1967).

Microscopically the haemangiosarcoma shows a network of multiple atypical capillaries and anastomoses lined by swollen anaplastic endothelial cells. In some cases (Mackenzie, 1961; Steingaszner et al., 1965) the tumour appears at the onset to be a benign haemangioma and the gradual transition to a malignant tumour is observed only in the recurrences. It is important to stress this behaviour because, as also happened in the present case, the tumour may be initially diagnosed erroneously as a benign lesion.

Another important question is whether multiple foci of this lesion are the result of metastases from a primary localized focus or whether these arise through a malignant transformation of multiple angiomatous malformations (Patrick et al., 1957). In the present case the angiomatous tumour in the breast definitely preceded all the other lesions. The clinical history and the scanning performed three months before death did not demonstrate any vascular lesion in the liver. Furthermore primary haemangiosarcoma of the liver is frequently associated with haemopoietic foci, possibly formed by differentiation of Kupfer cells into haemopoietic stem cells (Baker, Paget, and Davson, 1956). In the present case such haemopoietic foci were not found, and contributes to our assumption that the hepatic vascular tumours found at necropsy were metastases which occurred in the last months of the patient’s life.

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References


