HAEMANGIOSARCOMA OF THE BREAST*

I. LISSOOS, F.C.S. (S.A.), A. SCHMAMAN, M.B., B.Ch., D.C.P. (LOND.), M.C. PATH. AND S. KLEINOT, F.R.C.S. (EDIN.), D.P.H., *Departments of Surgery and Pathology, Baragwanath Hospital, the University of the Witwatersrand and the SAIMR, Johannesburg*

The rarity of malignant tumours of undoubted vascular origin warrants the publication of each case.[1,2] One haemangiosarcoma of the breast is seen for every 2,000 carcinomata of the breast,[3] and according to Stout[4] the breast is the most frequent site of haemangiosarcomata. Forty-one cases of haemangiosarcomata of the breast are recorded in the literature. The first well-documented case was reported by Borrmann[5] in 1907. Batchelor[6] has previously reported a case in an African woman.

The disease has been called angiosarcoma,[7-10] benign metastasizing haemangioma,[5,11] haemangioblastoma,[6,12,13] haemangio-endothelioma[4] and haemangiosarcoma.[14,15] Haemangiosarcoma is the most favourable term, describing both the malignancy of the condition and its vascular origin.[16]

The following case report reveals some interesting features of the disease.

CASE REPORT

A Bantu schoolgirl, aged 15 years, was admitted to Baragwanath Hospital in March 1965, complaining of a swollen right breast. She said that the lesion had only been present for one week. There was no history of a nipple discharge or previous trauma to the breast.

On examination the right breast was 1½ times larger than the left breast. An area of erythema and induration surrounded the areola of the right breast. A mass about 3 in. in diameter could be felt below the nipple and areola. The breast was fully mobile and not fixed to underlying structures. Mobile, tender lymph nodes were palpable in the right axilla. Radiological examination of the chest was normal. A biopsy specimen of the tumour was obtained under general anaesthesia. The patient bled profusely from the biopsy site and required a blood transfusion.

Microscopically, the tumour showed the features of a haemangiosarcoma and consisted of proliferating oval and rounded cells with pleomorphic nuclei lining large and smaller blood spaces (Figs. 1 and 2). In some areas there were dense sheets of cells showing a moderate number of mitoses (Fig. 3).

In view of the malignant nature of the disease, a radical mastectomy and block dissection of the right axilla was performed. Pathological examination showed an enlarged breast in which there was a small fungating mass, 2 cm. in diameter, near the nipple (the biopsy site). A large area of skin around the nipple and extending as far as 4 cm. from the nipple was thickened, oedematous and reddish. Section through the breast showed a large (12 cm. diameter) haemorrhagic and fleshy mass of soft consistency occupying most of the breast (Fig. 4). Histological examination showed features similar to the original biopsy. The axillary lymph nodes showed well-marked follicular hyperplasia and sinus histiocytosis but no evidence of metastatic deposits.

*Date received: 21 March 1969.

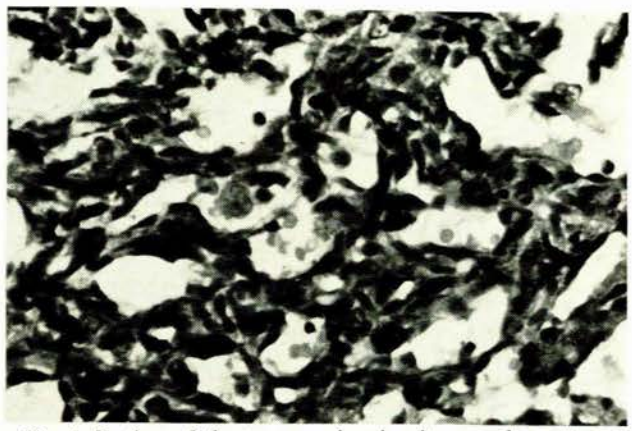

Fig. 1. Section of the tumour showing its vascular nature (H & E × 480).

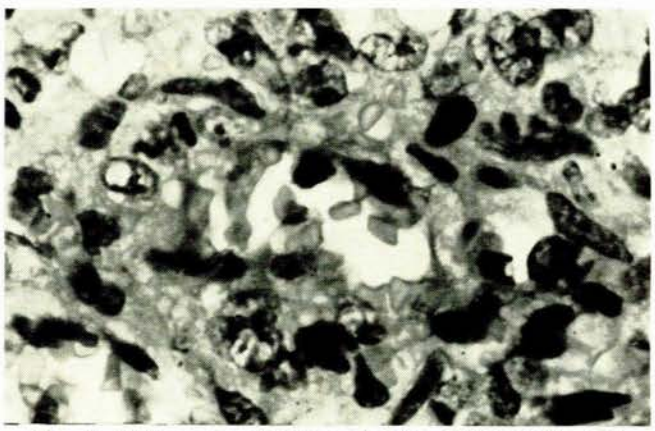

Fig. 2. Higher magnification showing pleomorphic and actively dividing cells lining the vascular spaces (H & E × 1,200).

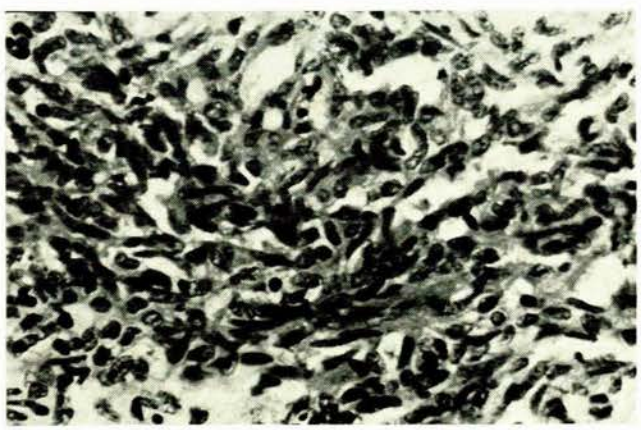

Fig. 3. An area where the tumour is more solid and its vascular nature not so obvious. Several mitoses are present (H & E × 480).

The patient made an uneventful recovery and was discharged from the hospital but told to return for follow-up examination. She did not, however, return to hospital until June 1967 (2 years later), when she complained of a lump in her left breast which she claimed had been present for only 2 weeks. Her left breast was grossly enlarged; peau d'orange of the whole breast was present and there were numerous septic ulcers over the breast. The breast was hot, red, tender and fixed to the underlying structures. A large mobile lymph node was present in the left axilla. On auscultation of her chest numerous rhonchi and crepitations were heard. Her liver was palpable and tender and she was anaemic (haemoglobin concentration 6·2 G/100 ml.). Her chest X-ray showed multiple pulmonary opacities strongly suggestive of metastatic deposits.

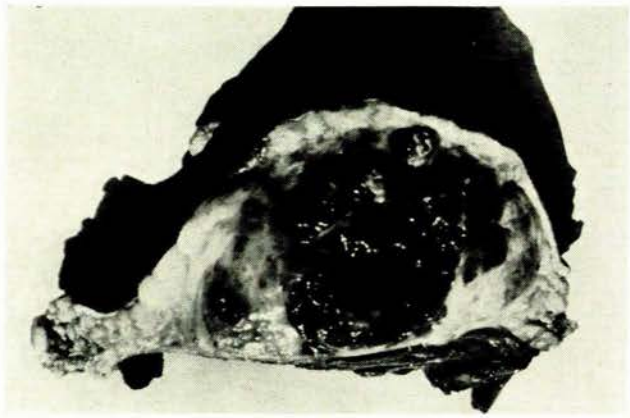

Fig. 4. Cross-section of the breast, showing the tumour occupying almost the entire breast.

A punch biopsy specimen of the lesion was obtained, using local anaesthetic. The histological features of this tumour were similar to those seen previously in the right breast.

The patient's condition deteriorated steadily and after a series of convulsions she died a fortnight after admission. Her relatives refused to allow an autopsy to be performed.

DISCUSSION

Young women are affected more frequently than older ones, the average age of the reported cases being 35 years. Thirteen of the 41 patients reported presented with a painful or tender swelling in the breast,[2,7,10,11,16-18] while the remainder complained of a painless swelling. Bilateral involvement of the breasts occurred in 11 of the cases. This once more raises the question whether the involvement of the second breast is a metastasis or a second primary lesion, a question difficult to answer.[6,16-19]

An interesting feature of the disease is the high incidence of haemorrhage following any operative procedure or as a terminal event.[9,15,17] Our case required a blood transfusion following the first biopsy.

The average survival period after diagnosis of the disease was 2½ years. Only 4 cases have survived for longer than 5 years.[10,16,20] The most frequent sites of metastasis were the skin, lungs, skeleton and abdominal viscera, particularly the liver and ovaries.

An outstanding feature of the reported cases is the frequency with which the misdiagnosis of a benign lesion was made on histological examination. Steingasner *et al.*[16] suggest that, if the following points are borne in mind, recognition of the malignant nature of the disease should be possible:

1. The lining of vascular spaces by endothelial cells with larger and more hyperchromatic nuclei than are found in haemangiomata.

2. The finding of at least a few foci where the endothelial cells are piled up and form papillary projections into the lumen of the vascular channel.

3. The presence of freely anastomosing and irregular intercommunicating vascular channels.

4. The great variation in cellularity in the different parts of the tumour.

Haagensen[21] describes the use of infrared photography as an aid to the diagnosis of this condition, the marked vascularity of the tumour being well demonstrated. Mackenzie[5] described the use of arteriography through the subclavian artery to demonstrate the vascularity of the tumour.

The ideal form of treatment is a local mastectomy with the incision well clear of the tumour. Local excision of the lesion results in a high incidence of local recurrence. Axillary lymph-node dissection, although frequently performed, is unnecessary as the lesion rarely metastasizes to lymph nodes.

SUMMARY

Haemangiosarcoma is a rare form of breast malignancy which affects mainly young women. A case is reported of a 15-year-old Bantu schoolgirl who, after 2 years, again presented with a similar lesion in the other breast and pulmonary metastases.

We should like to thank Dr W. H. F. Kenny, Medical Superintendent of Baragwanath Hospital, for permission to publish this case; Prof. J. H. S. Gear, Director of the South African Institute for Medical Research, for granting facilities; and Mr M. Ulrich for the photographs.

REFERENCES

1. Gill, W. and McGregor, J. D. (1968): J. Roy. Coll. Surg. Edinb., **13**, 155.
2. Raven, R. W. and Christie, A. C. (1953 - 54): Brit. J. Surg., **41**, 483.
3. Stewart, F. W. (1950): *Atlas of Tumor Pathology*, fasc. 34. Washington, DC: Armed Forces Institute of Pathology.
4. Stout, A. P. (1943): Ann. Surg., **118**, 445.
5. Borrmann, R. (1907): Beitr. path. Anat., **40**, 372.
6. Batchelor, G. B. (1958 - 59): Brit. J. Surg., **46**, 647.
7. Barber, K. W., Harrison, E. G., Clagett, O. T. and Fratt, J. H. (1960): Surgery, **48**, 869.
8. Mackenzie, D. H. (1961): Brit. J. Surg., **49**, 140.
9. McCarthy, W. D. and Pack, G. T. (1950): Surg. Gynec. Obstet., **91**, 465.
10. McClanham, B. J. and Hogg, L. jnr. (1954): Cancer (Philad.), **7**, 586.
11. Robinson, J. M. and Castleman, B. (1936): Ann. Surg., **104**, 453.
12. Edwards, J. A. and Strouth, B. P. (1956): Northw. Med. (Seattle), **55**, 788.
13. Patrick, R. S., Jarvis, J. and Miln, D. C. (1957 - 58): Brit. J. Surg., **45**, 188.
14. Mallory, T. B., Castleman, B. and Parris, E. E. (1949): New Engl. J. Med., **241**, 241.
15. Shore, J. H. (1957): J. Path. Bact., **74**, 289.
16. Steingasner, L. C., Enzinger, F. M. and Taylor, H. B. (1965): Cancer (Philad.), **18**, 352.
17. Edwards, A. T. and Kellett, H. S. (1968): J. Path. Bact., **95**, 457.
18. Enticknap, J. B. (1946): Brit. Med. J., **2**, 51.
19. Tibbs, D. (1952 - 53): Brit. J. Surg., **40**, 465.
20. Menville, J. G. and Bloodgood, J. C. (1933): Ann. Surg., **97**, 40.
21. Haagensen, C. D. (1956): *Diseases of the Breast*, p. 343. Philadelphia: W. B. Saunders.