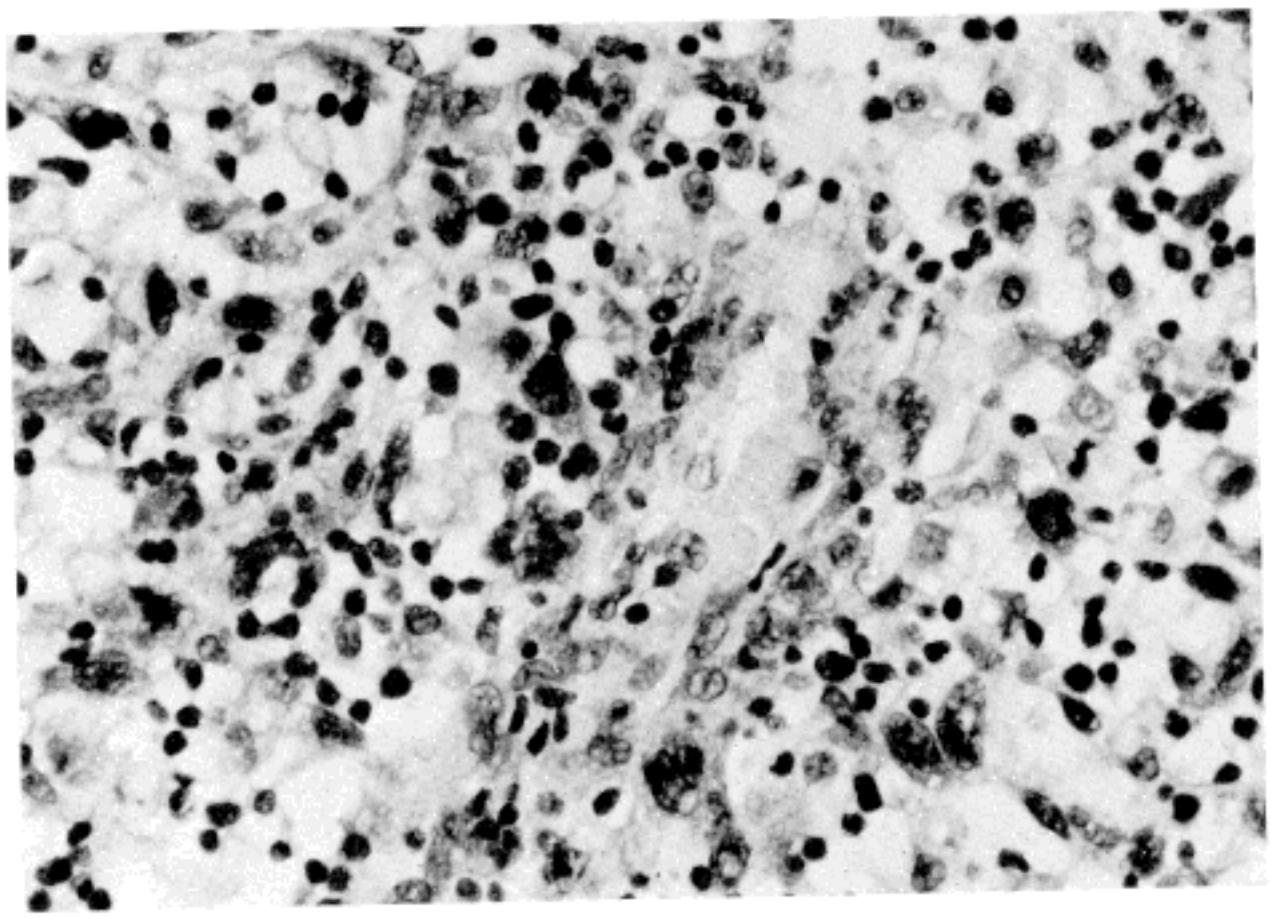

Fig. 5. Occasional vascular spaces containing red blood cells. (H&E, ×400)

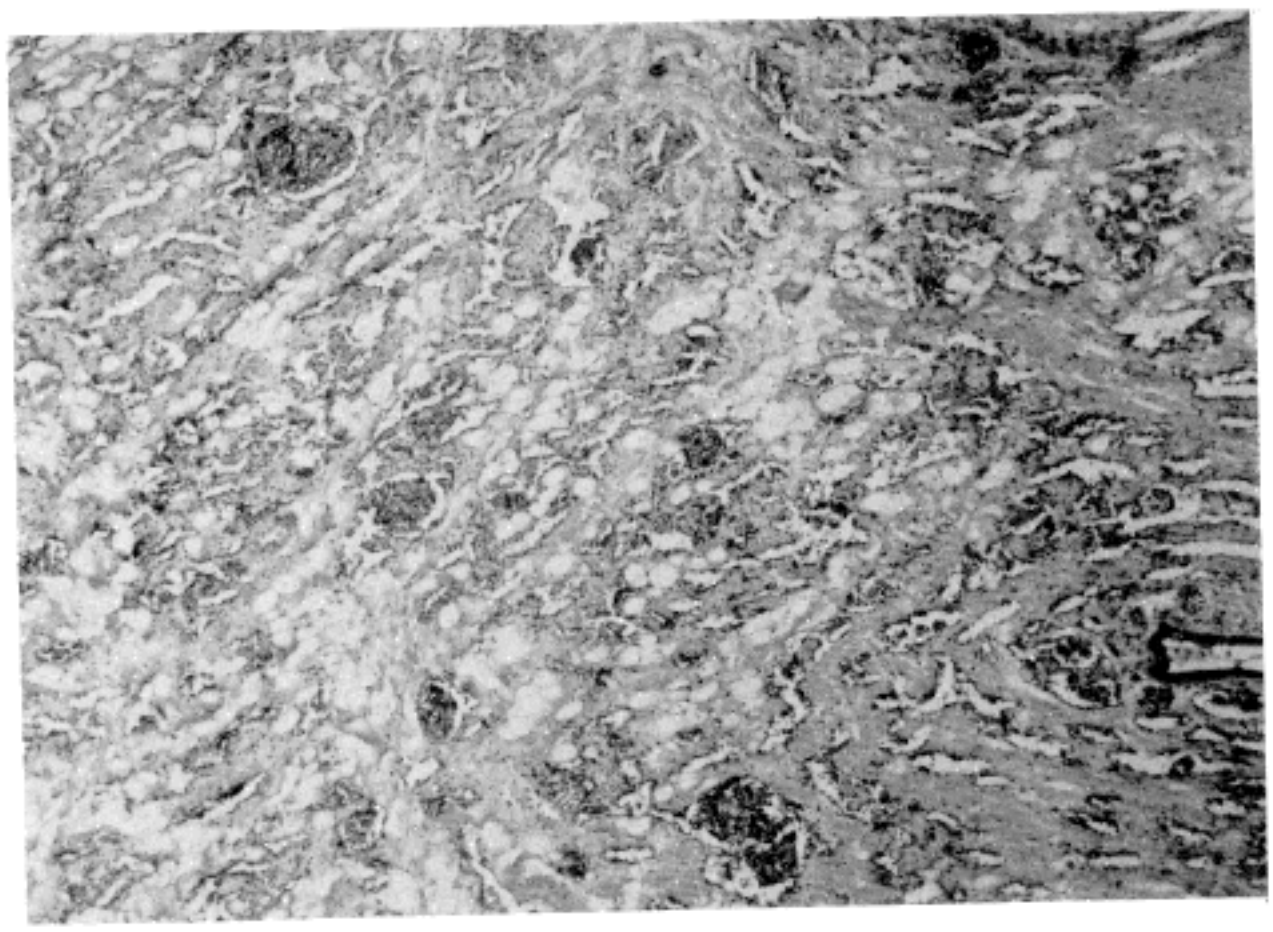

Fig. 6. Infiltrating vascular network of interanastomosing channels and prominent endothelial cells. (H&E, ×40)

tration raises a possibility of lymphatic differentiation.

In case 2 the tumor was entirely composed of infiltrating vascular network (Fig. 6). The vascular spaces were irregularly interconnecting and lined by a single layer of prominent hyperchromatic endothelial cells. The lumina sometimes contained red blood cells or cellular tufts of endothelial cells in which mitotic figures were often found (Fig. 7). Parenchymal duct structures and adipose tissue were found entrapped within the tumor (Fig. 8).

Case 3 showed similar appearance to that of case

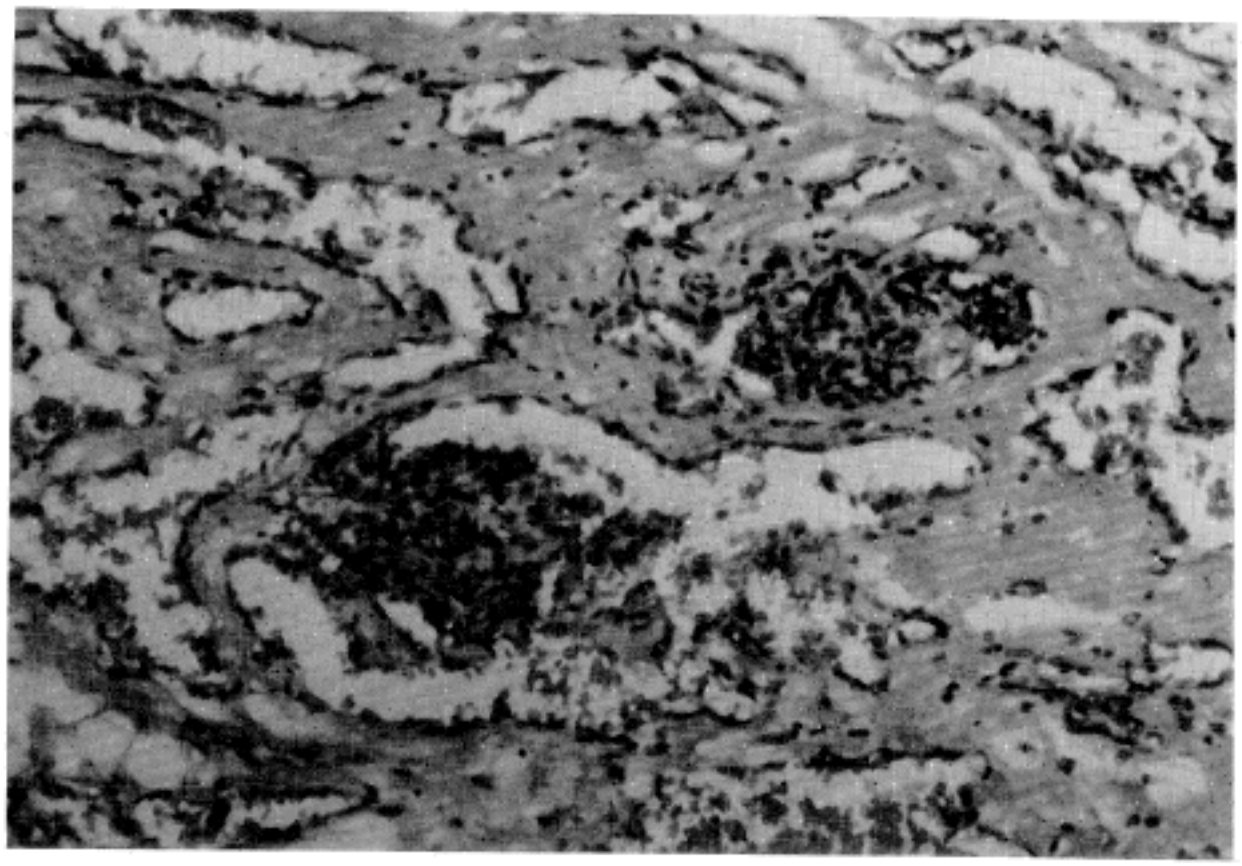

Fig. 7. Intraluminal endothelial tufting. (H&E, x200)

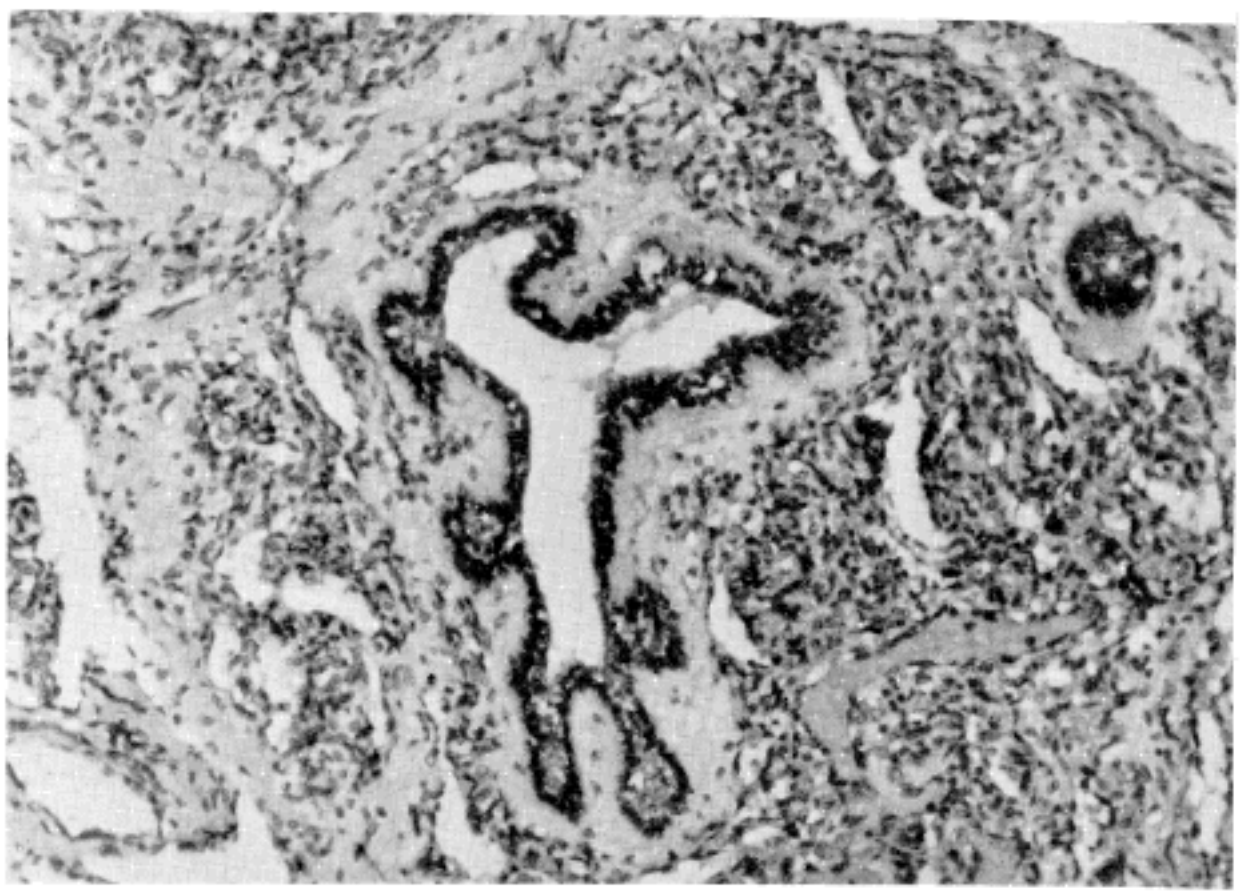

Fig. 8. Entrapped mammary duct within the tumor. (H&E, x200)

2, but more frequent and prominent endothelial tufting and papillary projection (Fig. 9), in which hyperchromatic anaplastic cells and a few mitoses were easily found (Fig. 10). This tumor was also highly infiltrative only sparing islands of normal ductal structures. Both case 2 and 3 were classified as moderately differentiated form (grade II).

DISCUSSION

Since the early descriptions by Schmidt (1887) and Borrmann (1907), angiosaroma of the breast has shown quite unique feature. In addition to its occurrence in young women, presentation as a rapidly

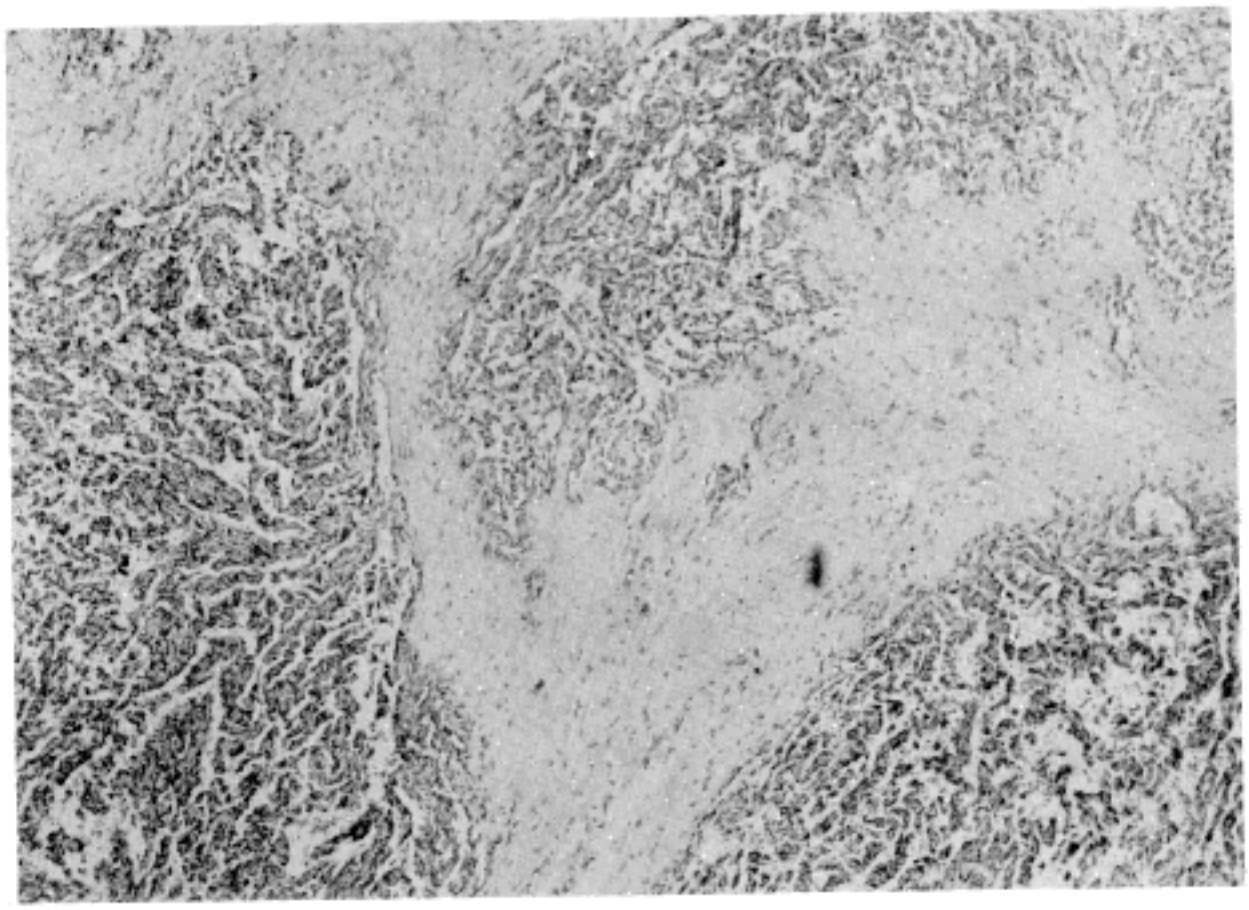

Fig. 9. Large irregular vascular spaces showing prominent intraluminal papillary projections. (H&E, x40)

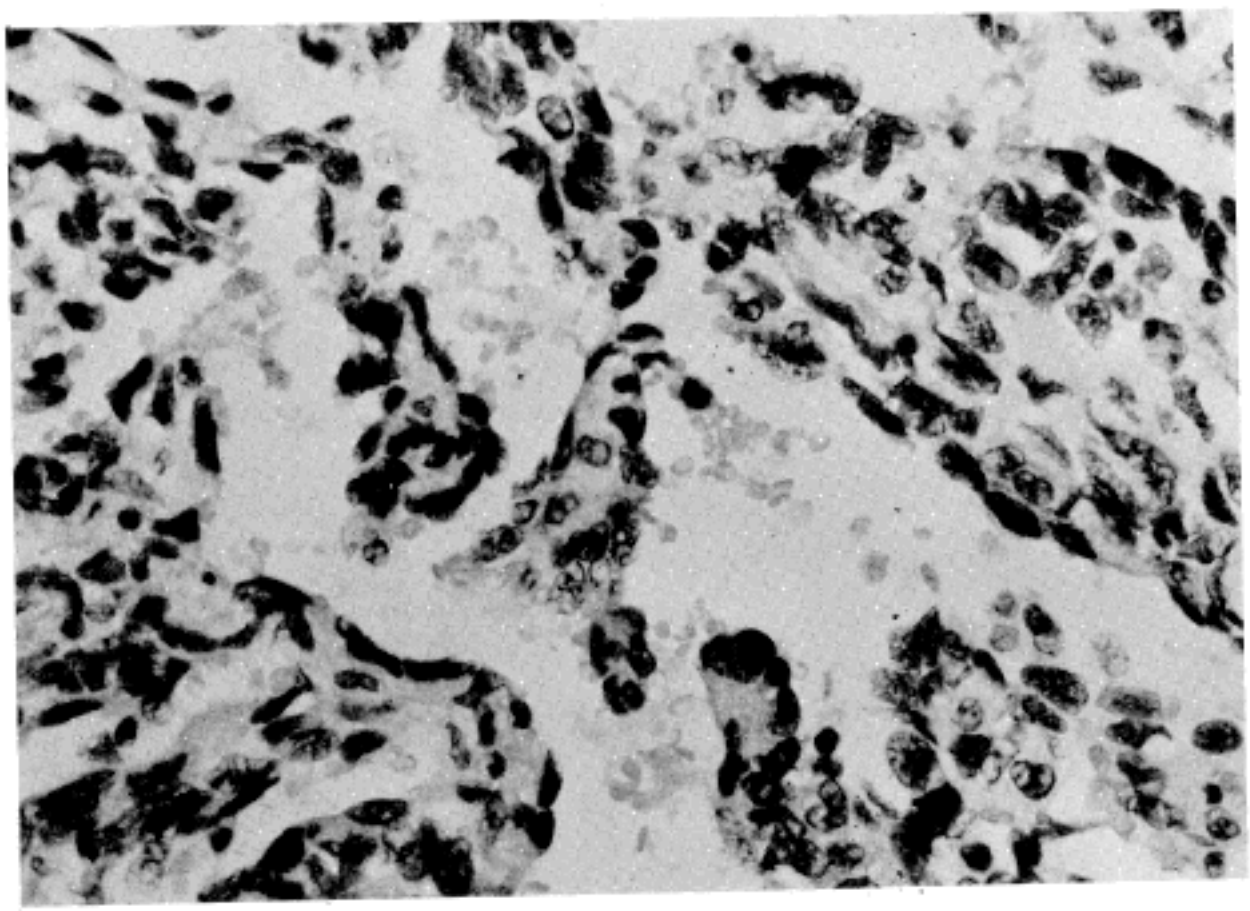

Fig. 10. Large pleomorphic endothelial cells showing occasional mitoses. (H&E, x400)

growing large mass and hightly malignant behavior including frequent involvement of contralateral breast, the histology does not greatly vary either. Our cases are not exceptions, though the age distribution shows some range (25, 46 and 66 years). The present three cases were encountered among about 1000 breast cancers experienced in the surgical pathology service of the Seoul Nat'l University Hospital during the last 25 years. This incidence is higher than that previously has been estimated (0.03 ~0.06%)[2~4].

Grossly the tumor is characterized as poorly cir-

cumscribed vascular mass and microscopically composed of interanastomosing vascular channels lined by hyperchromatic endothelial cells which have potential to become more anaplastic. Merino et al [5] classified it into three groups. Well differentiated form showed no hemorrhage, necrosis, pleomorphism or anaplasia, rare mitoses and occasional endothelial tufting and thrombosis. Moderately differentiated group had foci of solid tumor comprising less than 20%, and poorly differentiated form was composed predominantly of solid tumor showing hemorrhage, necrosis, cellular pleomorphism and abundant mitotic figures. Hunter et al[6] classified the tumor as grade I if it is characterized by previously described vascular channels without mitoses, necrosis and papillary projections, grade II with endothelial tufting and papillary projections, and grade III with the same criteria as the above poorly differentiated form.

The only differential diagnosis is benign vascular lesion of the breast, which is rarer than angiosarcoma. Steingaszner et al[7] had already described larger and hyperchromatic lining cells, endothelial piling up and intraluminal papillary projections and freely anastomosing and irregular intercommunicating vascular channels as criteria of malignancy. Jozefczyk et al[8] reviewed 100 vascular lesions of the breast, comprising 62 angiosarcomas, 11 perilobular hemangiomas, 14 hemangiomas, diffuse vascular proliferation or angiomatosis and venous hemangioma. The atypical forms of perilobular hemangioma and hemangioma showed some degree of cellular atypism and focal anastomosis, but remained benign. They emphasized small size (less than 2.5 cm) and good circumscription of the benign vascular tumors. There is no evidence to prove angiosarcoma arises from atypical hemangioma.

The prognosis is known to be related to the histologic degree of differentiation and Merino et al concluded that well differentiated tumor tend to have indolent course and late recurrences. However, angiosarcoma is the most malignant tumor of the breast and all the present cases revealed metastasis within postoperative 6 to 10 months and one with contralateral breast involvement. The axillary lymph node metastasis is very rare in angiosarcoma of the breast, and one such a case has been reported in 1985 on a monthly conference of the Korean Society of Pathologists.

The treatment of angiosarcoma of the breast has been in controversy. Gulesserian[9] and Fernandez[10] recommended radical mastectomy, but Chen[11] and Savage[12] reported that simple mastectomy was apropriate and axillary lymph node dissection would be superfluous. Adjuvant chemotherapy or radiotherapy has been ineffective in most of the patient.

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＝국문초록＝

유방의 맥관육종

— 3예 보고 —

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66세, 46세 및 25세 여성의 유방에 발생한 맥관육종 3예를 보고하였다. 제 1 예는 좌측유방의 고형종괴로 비정형 과염색성 혈관내피세포의 군집으로 이루어져 나쁜 분화도를 보였으며 환자는 절제술 6개월후에 연부조직 전이를 나타내었고 14개월에 사망하였다.

제 2 예와 3예는 우측유방의 혈관성 대형종괴로 조직학적으로 서로 교통하는 불규칙한 혈관강과 혈관내피세포의 강내증식과 유두상 돌출을 보여 중등도의 분화도에 속하였다. 제2예는 근치적 유방절제술 10개월후에 좌측유방과 하복부에 전이성 종괴가 생겨 약물치료를 받았으나, 계속 악화중이고 제3예는 단순유방절제술후 6개월에 간으로 전이를 보였다. 본 종양은 유방암중 가장 나쁜 예후를 가지며, 전유방암의 약 0.3%를 차지하여 외국의 집계(0.03～0.06%)보다 높은 빈도를 보였다.

대한병리학회지 : 제22권 제1호
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Angiosarcoma of the Breast

— Three cases report —

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Three cases of angiosarcoma of the breast are described. One case in a 66 year old woman was a solid mass and histologically showed poor differentiation leading to a death 14 months after the diagnosis. Another two in 46 and 25 year old females were large vascular masses showing moderate differentiation with endothelial tufting and papillary projections, and abdominal and contralateral mammary, and liver metastasis occurred within postoperative 10 months and 6 months in each case.

A radical mastectomy and adjuvant chemotherapy in case 2 were not lifesaving. Although histologic degree of differentiation seems prognosis-related, angiosarcoma is the most malignant tumor of the breast.

Key Words: Angiosarcoma, Breast, Hemangioma

INTRODUCTION

Angiosarcoma of the breast is a rare malignant tumor of vascular origin, being a distinct clinical and pathologic entity. It frequently occurs in young women, presents as a rapidly growing painless mass and is highly malignant in spite of its characteristically innocuous appearance. The prognosis is known to be related with the histologic degree of differentiation and the most effective therapeutic modality is not conclusively settled yet. In Korean literature only one case was recorded in 1980[1]. This paper describes three new cases of angiosarcoma of the breast and discussion about their estimating incidence, histopathologic features, prognosis and treatment with a review of the literature.

* 본 논문의 요지는 1986년 5월 23일 대한병리학회 제11차 춘계 학술대회에서 발표되었음.

CASE HISTORY

Case 1.

A 66 year old female was found to have a left breast mass in February 1985. She had been suffered from known liver cirrhosis since 1975. An excisional biopsy of the breast mass was done and the specimen was interpreted as angiosarcoma, but no further treatment was given. Six months later another mass developed in the right buttock, but biopsy was not performed. Her physical condition deteriorated continuously and she died of multiple organ failure in April 1986.

Case 2.

In May 1985, a 46 year old female visited a physician for a right breast mass which had been initially noticed 11 months previously. Physical examination revealed a 10 cm sized slightly movable firm mass with intact overlying skin. A needle aspiration showed bloody content. A radical mastectomy was

done and histologic examination revealed a relatively well differentiated angiosarcoma without any axillary nodal metastasis. A new mass was found in her contralateral breast 10 months after the operation, and another adjacent nodule and a lower abdominal mass subsequently developed. Thereafter she received 2 cycles of chemotherapy with Cytoxan, Vincristine and Adriamycin, but has been suffering from multiple masses in left breast, lower abdominal pain and distension of abdomen till now, as of the time of this report.

Case 3.

In April 1986, a 25 year old female visited her physician for a right breast mass which had been found 14 months previously. The mass initially was small, measuring 2 cm in diameter, and grew continuously accompanying purplish discoloration of the overlying skin. A simple mastectomy was done under the impression of hemangioma and the mass was histopathologically proven to be angiosarcoma. No further treatment was done and the patient visited another hospital 6 months later for hepatic rupture which turned to be due to metastasis of angiosarcoma. She has been lost from our follow-up since then.

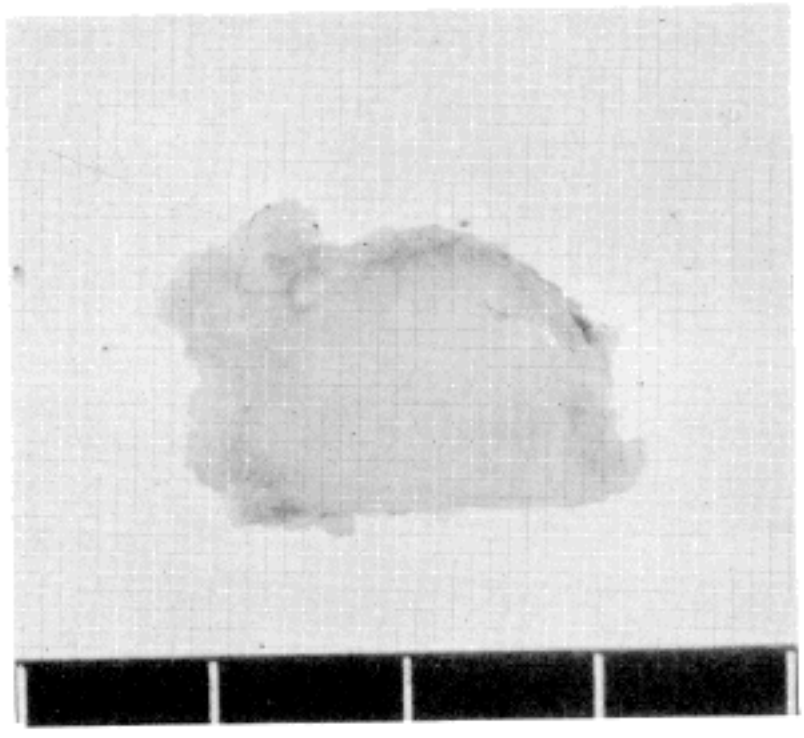

Fig. 1. A homogeneous pale pink solid mass in case 1.

PATHOLOGIC FINDINGS

Grossly, the excised tumor in case 1 was an ovoid pinkish, rather firm mass, measuring 3 cm in diameter. Cut surface was solid and homogeneous, pale pink in color and rubbery firm in consistency. Vascular spaces were not identifiable (Fig. 1). In case 2 the tumor measured 10×5×5 cm and was a poorly demarcated rather solid hemorrhagic mass. Cut surface showed numerous blood filled spaces with hemorrhagic nodules (Fig. 2). Case 3 showed a poorly demarcated hemorrhagic and cavitary mass, measuring 8×5×3 cm. Cut surface was purplish and spongy in appearance with various sized hemorrhagic spaces (Fig. 3).

Microscopically three cases of present series showed some variation pertaining to their degree of

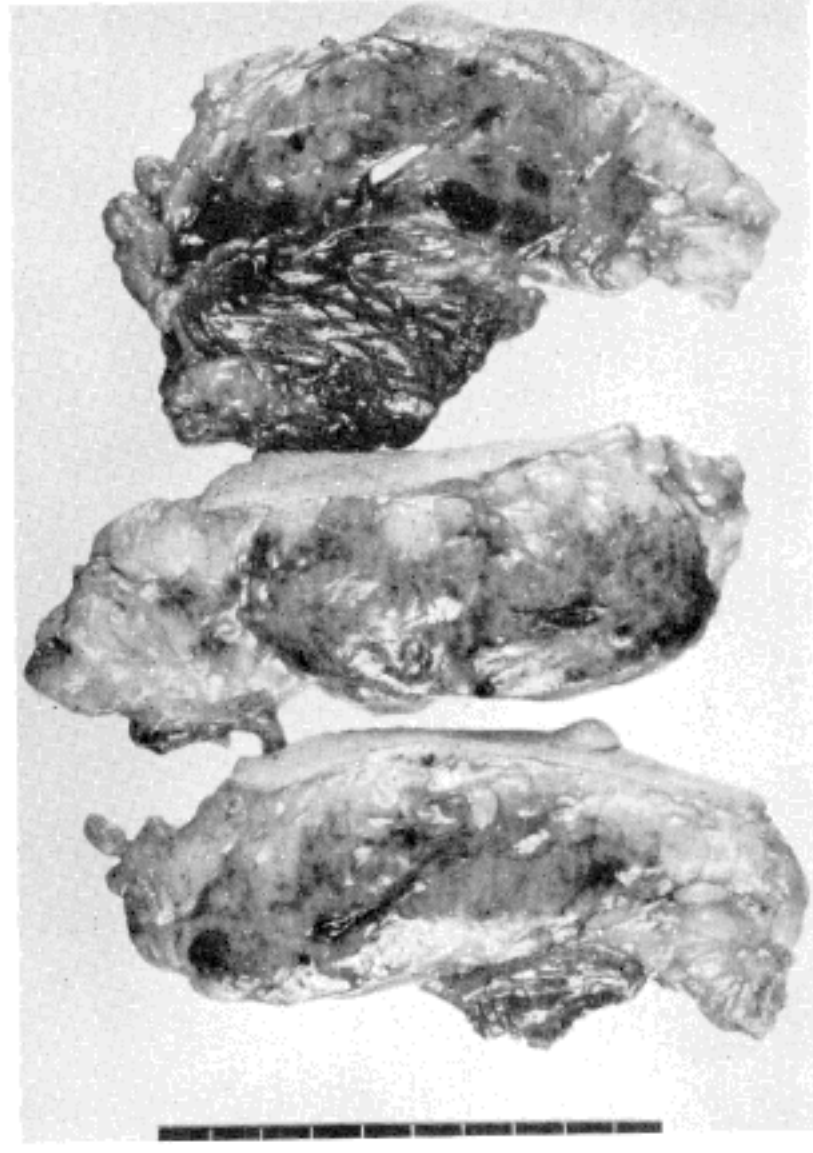

Fig. 2. A poorly demarcated large vascular mass in case 2.

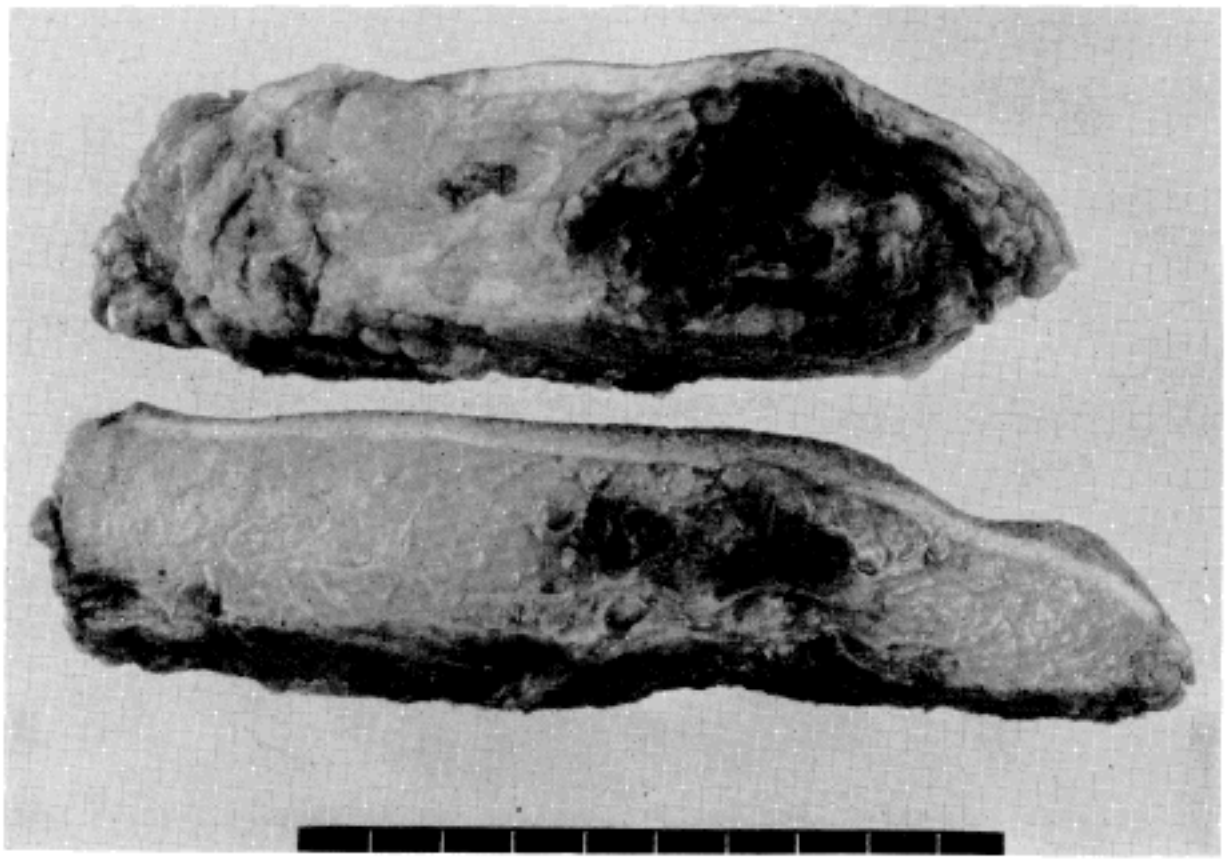

Fig. 3. A poorly demarcated vascular and spongy mass in case 3.

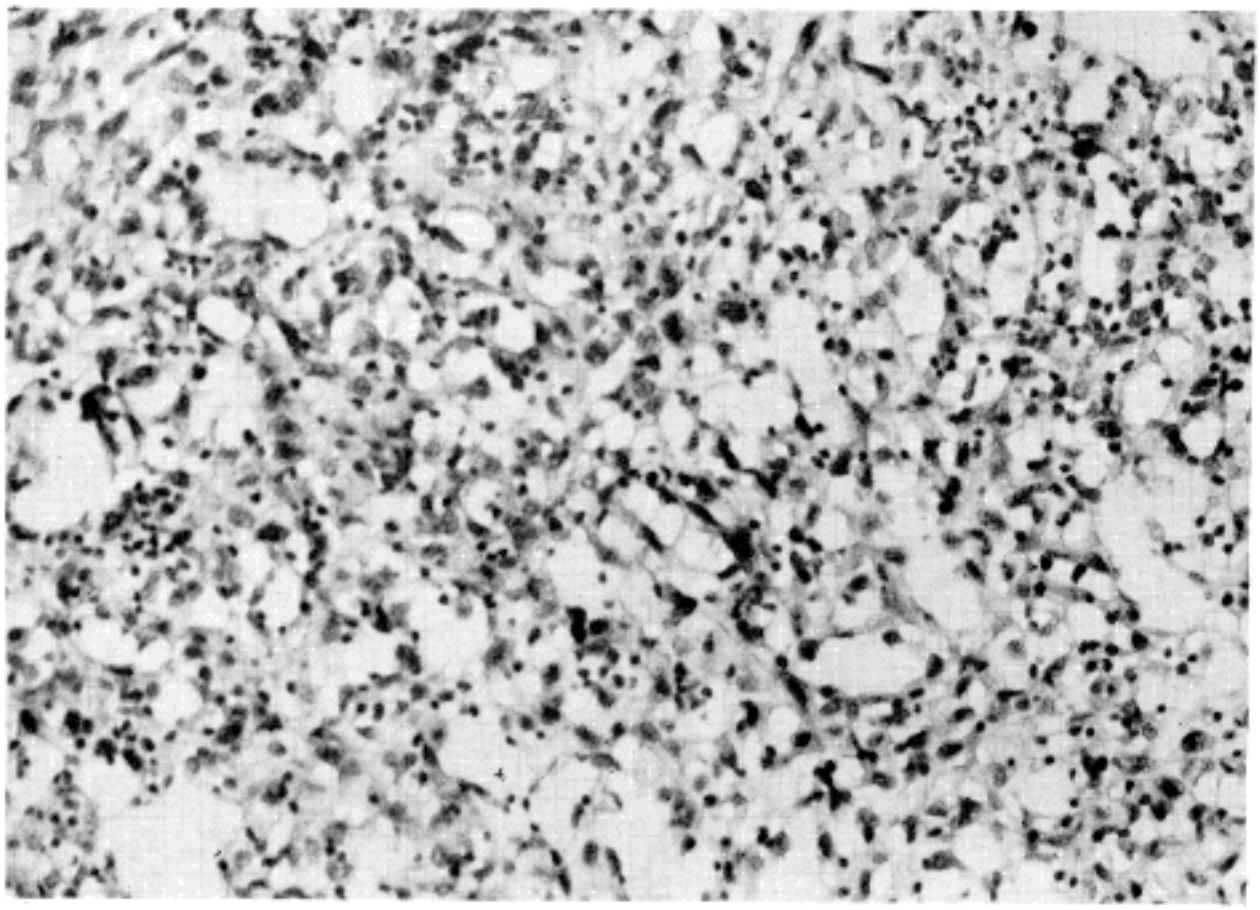

Fig. 4. Loosely arranged pleomorphic spindle cells and intervening lymphocytes in case 1. (H&E, x100)

differentiation. In case 1 the tumor was composed of pleomorphic spindle cells, which were rather loosely arranged without definite organoid pattern or intervening stroma (Fig. 4). The cells occasionally ran in rows or formed vascular like spaces containing red blood cells. Mitotic figures were frequently seen and there were many scattered lymphocytes in-between the tumor cells (Fig. 5). These findings were compatible with a poorly differentiated form (grade III) of angiosarcoma. The patient had no history of lymphedema or lymphangiomatosis, but lack of hemorrhagic nature and associated lymphocyte infil-