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Giant Cell Arteritis of the Breast

–A case report–

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Recently we experienced a case manifested by a lump in both breasts that mimicked carcinoma but was found to be a vasculitis of the giant cell type. Our case was characterized histologically by granulomatous arteritis involving small to medium sized arteries of the breast without involvement of the temporal artery.

We described the clinical and pathologic features of a case of giant cell arteritis of the breast and reviewed the eight previously reported cases in foreign literature.

Key Words: Giant cell arteritis, breast and temporal artery

Arteritis is generally a multiorgan disease but lesions in the breast have only rarely been reported. Giant cell arteritis is a systemic panarteritis affecting predominantly elderly patients (Fauci *et al.* 1978; Goodman 1979). Granulomatous giant cell arteritis has been observed to occur in virtually all the large to medium-sized and small systemic arteries of the body to (Hamilton *et al.* 1971; Ostberg 1973; Lie 1978) as well as in the pulmonary arteries (Wagenaar *et al.* 1981), and as the entity of isolated granulomatous angiitis of the CNS (Nurick *et al.* 1972).

Although small to medium-sized cranial arteries, most commonly the temporal arteries, may be involved, granulomatous arteritis may rarely present as an isolated finding involving small to medium-sized arteries in an extracranial site. Such involvement has been descibed in the breast and the female genital tract (Bell *et al.* 1979; Waugh 1950; Dega *et al.* 1974; Chaitin *et al.* 1981; Potter *et al.* 1981; Thaell and Saue 1983).

This form of vasculitis is a specific form of isolated granulomatous giant cell angiitis that selectively affects blood vessels of the breast or the feamle genital tract. This is usually without concurrent involvement of other organs or involving other organs to a less significant degree. The giant cell arteritis involving the breast was first described by Waugh in 1950, to date, only eight cases have been reported in the foreign literature.

A patient with giant cell arteritis, whose initial symptoms were due to involvement of arteries in the breasts, is reported.

CASE REPORT

A 69-year-old woman was admitted to the hospital because of painful lumps in both breasts of 4 weeks duraton. Also eczema and painful lumps of the right breast were found 1 momth before. These symptoms had developed in the left breast initially. Lumps of both breasts wer hard and fixed to the deep tissue but not to the skin.

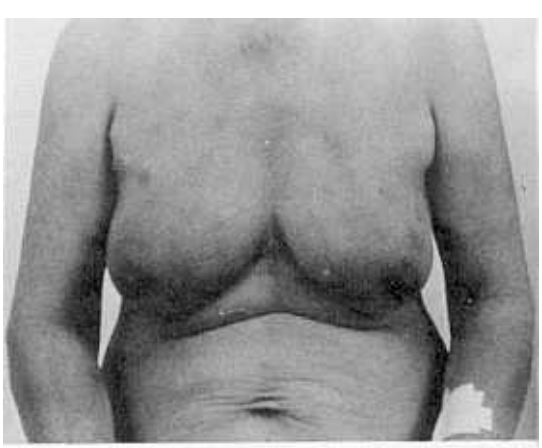

Fig. 1. Both breasts show multiple, plum-sized, tender, and eccymotic areas.

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Table 1. Reported cases of giant cell arteritis of the breast

Case	Age (yrs)	Presenting Features	Hemoglobin (g/dl)	ESR (mm/h)	Treatment and Follow-up
Waugh	64	Hard 1.3cm mass in right breast; 2 mth later, tender, firm, fixed 3.0cm mass in left breast	Anemia	40	Elevated ESR at 14 mth; no evidence of disease at 18 mth
Dega and Hunder	72	Firm, tender 3.0cm mass in left breast; weakness, pain in extremities, night sweats, fatigue, anorexia, weight loss	10.5	112	Muscle biopsy at 2 mth revealed polyarteritis; steroid therapy x 3yr; no evidence of disease at 8 yrs
Chaitin *et al.*	54	Two tender nodules, 0.2 and 1.5cm, in left breast, associated with hyperemia of overlying skin	NS	NS	No evidence of disease at 8 mth
Potter *et al.*	59	Hard, fixed 4.0cm mass in right breast with retracted nipple and palpable axillary nodes; similar mass in left breast; malaise, fever, diffuse aches and pain, swollen ankle	Normal	98	No evidence of disease at 20 mth
Thaell and Saue	72	Multiple discrete tender 1-to 2cm masses in both breasts; violaceous skin, enlarged left axillary node, fever, anorexia, weight loss	11.1	124	Steroid therapy x 6 mth; breast lump and elevated ESR recurred; steroids x 18 mth; no evidence of diease at 24 mth
Nirodi *et al.*	68	Painful 4cm mass in left breast; 4 yrs prior had rash and ESR>100 that regressed spontaneously	12.4	NS	Elevated ESR (60 mm/h); otherwise no evidence of disease at 1 mth
Stephenson and Underwood	62	Tender masses in both breast; 2.0cm mass in right breast biopsied	Normal	85	Steroid therapy x 2 mth; thyroid lobectomy at 2 wks revealed adenoma and vasculitis; no evidence of disease at 2 mth
Clement *et al.*	68	0.5cm mass in right breast; shoulder and neck pain, leg stiffness	Normal	65	Steroid therapy x 3 d; no evidence of disease at 14 mth
Present case	69	Firm, tender masses in both breasts	Normal	60	Steroid therapy x 15 d; no evidence of disease at 18 mth

Abbreviations: ESR, erythrocyte sedimentation rate; NS, not stated; mth, month.

On admission, she was chronically ill. Her temperature was 37°C and her pulse was 65 beats/min with a murmur. Three lumps in the right breast were tender, small, hard, somewhat irregular, and fixed. The largest lump was about 2.5cm in diameter. There was a similar lump in the left breast, measuring 1.5cm in diameter. Laboratory investigations revealed a hemoglobin level of 13.4 g/dl, a white blood cell count of 9470/mm³ with 63.2% polymorphous leukocytes and 20% lymphocytes, and an erythrocyte sedimentation rate(ESR) of 60 mm/hr (corrected ESR of 38 mm/hr). Liver function values were normal. A sonogram of the breast disclosed, that there was no definite evidence of a mass lesion, but several enlarged heterogenous mixed echogecities without any definite fluid collection were shown. The computer tomogram of the chest revealed mitral valvular disease with pulmonary edema probably due to heart failure. Under the provisional diagnosis of inflammatory carcinoma, lumps in both breasts were sampled for biopsy, and all the specimens were submitted for a frozed-section examintion. After the final diagnosis,

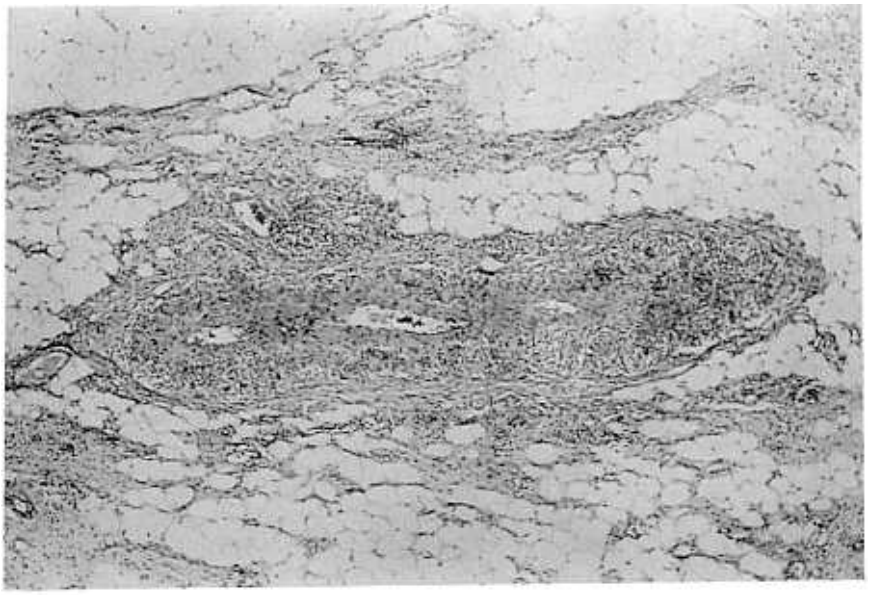

Fig. 2. Florid granulomatous arteritis involves aged, atrophic and fibrous parenchyma and surrounding fat tissue. (H & E ×100)

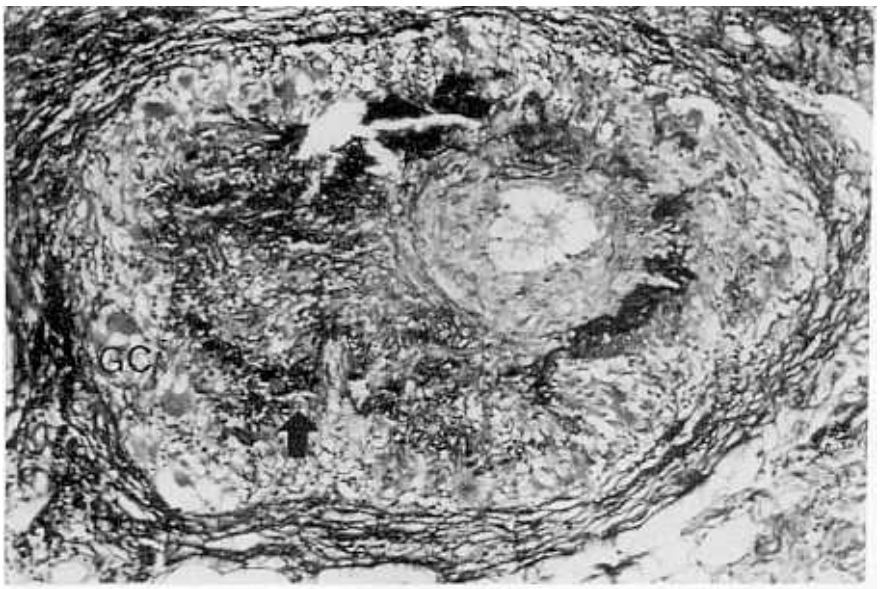

Fig. 3. Fragmentation of the internal elastic lamina (arrow) with giant cells(GC) closely apposed to the elastic fiber fragments. (Elastic Van-Gieson, ×200)

the evidence of the involvement of the temporal artery was not found.

PATHOLOGIC FINDINGS

Histologically the specimens from both breast lumps were similar. Typical fibrous atrophic parenchyma with florid granulomatous arteritis with surrounding infiltrated fat tissue was seen. The affected artery was a small to medium sized artery. It was a perforating branch of the internal mammary artery which supplies the medial portion of the gland. The walls

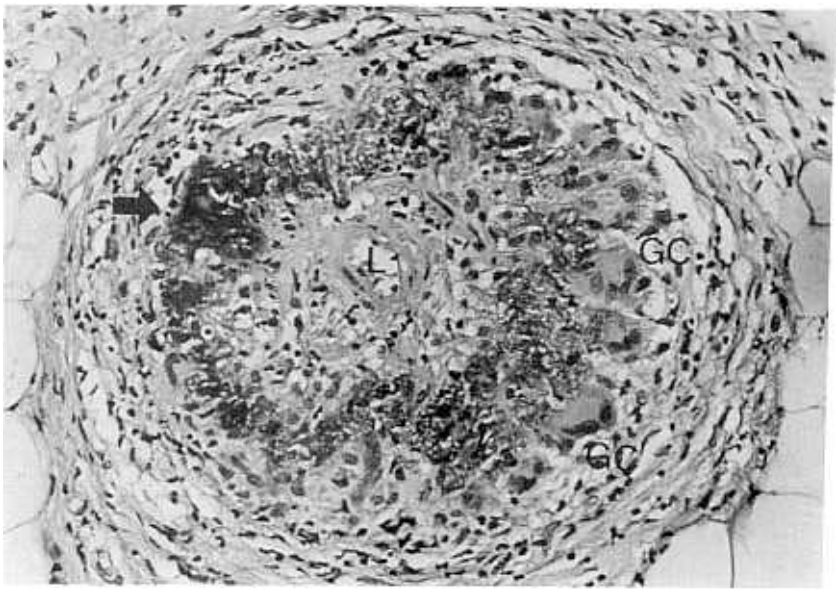

Fig. 4. The media of a medium-sized artery shows focal calcification (arrow), numerious inflammatory cells, predominantly histiocytes and giant cells(GC). The lumen(L) is narrowed due to intimal fibrosis. (H & E ×200)

of the involved vessels were thickened by transmural inflammatory infiltrates that were composed of lymphocytes, plasma cells, histiocytes, and multinucleated giant cells within the inner media. Elastic stains revealed focal disruption and loss of the internal elastic lamina, with numerous epithelioid histiocytes and giant cells which lay adjacent to the fragmented elastic fibers. There were varying degrees of intimal fibrosis resulting in luminal narrowing or obliteration. The arterioles, venuels and veins were unremarkable.

DISCUSSION

Giant cell arteritis is a disease of the elderly that generally involves the carotid arteries or their branches, especially the temporal arteries. However, it may rarely be present as an isolated finding with clinical evidence of the involvement of small to medium sized extracranial arteries in 10-15% of the cases (Huston *et al.* 1978; Klein *et al.* 1875). Histologic features are characterized by the following; 1) thickening of the vessel walls with marked lumenal narrowing, 2) an inflammatory infiltrate of lymphocytes, histiocytes, giant cells, eosinophils, and plasma cells, especially in the outer intima and inner media, and 3) the internal elastic lamina is lost or fragmented; histocytes and giant cells often are closely apposed to the elastic tissue fragments. Waugh (1950) reported a remarkable case of a 64-year-old women who presented with a symptomatic lump in her breast. This was the first case of giant cell arteritis of the breast that resulted in a simple mastectomy, despite failures to confirm carcinoma at the time of the intraoperative frozen section. Giant cell arteritis of the breast occurs predominatly in elderly women (mean, 65 years; range, 54 to 72), who show a high ESR and are usually febrile. Anemia and discoloration of the breast skin were noted in some patients. Four patients of eight cases showed bilateral involvement as in our case (Waugh 1950; Potter *et al.* 1981; Thaell and Saue 1983; Stephenson and Underwood 1986). In addition to the breast mass, four patients had synchronous systemic manifestations including one or more of the following: malaise, fever, anorexia, weight loss, weakness, muscle and joint pain, and stiffness (Dega and Hunder 1974; Potter *et al.* 1981; Thaell and Saue 1983; Clement *et al.* 1987). However, none of the patients had clinical manifestations related to any temporal artery involvement.

The cause of giant cell arteritis remains obscure. There is conflicting evidence on the involvement of immune complexes and cell-mediated hypersensitivity (Gallagher and Jones 1982). A possible role from environmental and genetic factors has been suggested (Goodman 1979; Liang *et al.* 1974; Hazleman *et al.* 1977).

Spontaneous regression (Potter *et al.* 1981) or steroid therapy result in a drop in the elevated ESR to normal levels and in the disappearance of systemic symptoms. One case of the eight patients had a coincidental, in situ, and infiltration ductal carcinoma, in the same biosy specimen (Clement *et al.* 1987).

The differential diagnosis of mammary giant cell arteritis includes other rare forms of vasculitis involving the breast. Six cases of Wegener's granulomatosis (Elsner *et al.* 1969; Pambakian and Tighe 1971; Kuhne 1983; Deininger 1985; McCarty *et al.* 1968) and four cases of polyarteritis presenting in the breast have been described (Elsner 1970; Nielsen and Lorentzen 1985). Other lesions in the differential diagnosis include: thrombophlebitis of the superficial veins of the breast (Mondor's disease), spontaneous infarction of breast tissue during pregnancy or lactation, and hemorrhagic necrosis of the breast with the complication of anticoagulant therapy. In our case, giant cell arteritis presented in the breasts of a postmenopausal woman without involvement of the temporal artery and it mimicked carcinoma on clinical examiantion.

We believe our case is the first report of giant cell arteritis involving both breasts, in Korea.

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