Breast involvement in Wegener's granulomatosis

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SYNOPSIS Two cases of Wegener's granulomatosis are described in which the early and predominant lesions were found in the breast. The histological features of breast necrosis with granulomatous inflammation and widespread vasculitis in the breast in the absence of any demonstrable pathogens enabled a presumptive diagnosis to be made in one case before the development of the more characteristic clinical features in the respiratory system.

Wegener's granulomatosis is characterized by necrotizing granulomatous lesions of the respiratory tract with necrotizing vasculitis involving arteries and veins in many organs and necrotizing glomerulonephritis (Godman and Churg, 1954). Limited forms of the syndrome have been described (Carrington and Liebow, 1966). By 1967, only 138 unequivocal cases had been reported (Nielsen, Christiansen, and Jensen, 1967). The first case of Wegener's granulomatosis involving the breast was described by Elsner and Harper (1969). Two further cases with breast involvement are reported in this paper.

Case 1

A 40-year-old woman was admitted to hospital in February 1965 with a mass in the right breast. This mass had appeared a short time before admission and had grown rapidly in size. There was no history of trauma and the patient was otherwise well. Histology of the excised mass revealed necrotic breast tissue with granulomatous inflammation and vasculitis, (Figs. 1 and 2) but no bacteria, fungi, or protozoa were identified. The operation wound failed to heal. Following operation the patient developed a persistent influenza-like illness.

In May 1965 a mass developed in the right thigh and was excised. Histology revealed appearances similar to those of the breast lesion. This operation wound also failed to heal.

By August 1965 the patient had extensive purulent necrosis of the right breast and right thigh but no pathogenic organisms were obtained by repeated cultures of the lesions. At this time a hard goitre was noticed. A month later the patient became hoarse, developed signs of bilateral basal pneumonia and uraemia, and died.

Postmortem examination revealed extensive ulceration and necrosis of the right breast extending from the skin to the pectoral muscle with destruction of the nipple and areola. There was ulceration and necrosis of the right thigh with exposure of the underlying muscles. There was extensive destruction of the lower larynx and the upper trachea with fragmentation of the tracheal rings. Necrosis extended into the soft tissues involving the deep surface of both lobes of the thyroid gland. There were extensive areas of consolidation in the right lung and, to a lesser extent, in the left lung, with overlying pleurisy. Both kidneys were pale and swollen. In the spleen there were multiple infarcts although the splenic artery was normal.

Microscopy revealed irregular infarcts with surrounding granulomata in the larynx, trachea, thyroid, and adjacent voluntary muscle. In small arteries there was fibrinoid change (Fig. 3) and acute arteritis. The kidneys showed a proliferative glomerulonephritis with epithelial crescents (Fig. 4). There were thrombi in the glomerular capillaries and an active arteritis. The splenic infarcts were associated with an acute arteritis involving some follicular arteries and there was extensive necrosis of the myocardium with a vasculitis. In the lungs there was severe oedema, hyaline membrane, bronchopneumonia and thrombosis of the pulmonary artery and alveolar capillaries. The appearances were typical of Wegener's granulomatosis.
Case 2

A woman of 42, in previous good health, presented in November 1968 with bilateral deafness, with pain and discharge from the right ear. Treatment with antibiotics was not effective. A month later she developed a right facial palsy. The tympanic membrane was found to be oedematous and granular but there was no pus. The facial palsy recovered but the deafness persisted. In January 1969 a mass was palpated in the left breast. There was slight skin tethering and clinically the lesion was diagnosed as a carcinoma. Excision biopsy revealed a poorly circumscribed, firm mass, 2.5 cm diameter, with a variegated grey-white cut surface. Histology showed necrotic breast tissue heavily infiltrated by histiocytes, plasma cells, lymphocytes, and polymorphs together with granulomata containing Langhan’s giant cells (Fig. 5). Vasculitis was present in the breast, in some areas far removed from the main necrotic mass (Fig. 6). No pathogenic organisms were identified and the appearances suggested a diagnosis of Wegener’s granulomatosis. Since July 1969 there have been recurrent epistaxes. The nasal mucosa and the posterior wall of the pharynx became swollen and granular. There were crusts and fibrous adhesions on the turbinates, and granular areas on both sides of the nasal septum. Radiology revealed opacity of the left antrum. Swelling of the right orbit with involvement of the upper lid, lachrymal gland, and right side of the nose developed in August 1969. Biopsy of the lachrymal gland showed only atrophy with focal collections of histiocytes and polymorphs but no vasculitis. Culture of swabs from the conjunctival sac were negative. A month later a chest radiograph showed multiple round
discrete opacities in both lungs, consistent with Wegener’s granulomatosis (Fig. 7). The patient has since been treated with prednisone and to date there are no features of renal disease. However, the clinical and histological features so far evolved are characteristic of Wegener’s granulomatosis.

Discussion

Wegener’s granulomatosis is a syndrome which typically occurs in previously healthy, young or middle-aged adults of either sex. In about two-thirds of the cases persistent purulent rhinorrhoea is accompanied by nasal obstruction and crusting with antral pain and epistaxis. Otorrhoea, deafness, or ulceration of the gums is the initial symptom in some of the cases. In one-third of the cases attention is drawn to the lungs because of persistent cough, haemoptysis, or pleuritic pain. There may be temporary improvement following antibiotic therapy but lesions always persist. Extension of the inflammatory process in the nose and palate leads to mucosal ulceration and to cartilagenous or osseous destruction. In the lungs it leads to consolidation. Spread through the upper air passages is often followed by conjunctivitis, impairment of vision, increased lacrimation, and exophthalmos. There may be hoarseness or dysphagia due to ulceration in the pharynx or larynx. In the late stages fever, often of septic type, is almost constant, and albuminuria, haematuria, cylinduria, and pyuria is seen in most cases. Uraemia and bronchopneumonia are the two common causes of death (Walton, 1958).

These two cases of Wegener’s granulomatosis are of interest because of the parenchymal breast involvement. In both cases the breast lesions were
Fig. 5  Case 2: at the top of the photomicrograph necrotic breast tissue can be seen and nearby there is granulomatous inflammation with Langhan's giant cells × 125.

Fig. 6  Case 2: some distance from the necrotic breast tissue there is evidence of vasculitis. A vessel in the upper part of the photomicrograph shows fibrinoid necrosis. × 125.

Fig. 7  Case 2: chest radiograph reveals opacities in the right lower zone and in the left apex.
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noticed early in the disease. In case 1 the breast lesion was the presenting symptom and was a prominent feature of the patient’s illness. The lesions in both cases showed breast necrosis with a granulomatous reaction, vasculitis widespread in the breast, and no demonstrable pathogens. These histological appearances are typical of Wegener’s granulomatosis. Nevertheless diagnostic difficulties may arise when a lesion of Wegener’s granulomatosis is met in an unusual site or in a patient in whom the typical clinical features have not developed. In the second case the diagnosis was suggested from the histological appearances of the breast lesion, and in the subsequent evolution of the patient’s illness has confirmed this histological diagnosis.

The lesions of Wegener’s granulomatosis in the breast have to be differentiated from specific infective granulomatous diseases and from sarcoidosis, traumatic fat necrosis, Weber-Christian disease, and classical polyarteritis nodosa.

Although specific infectious granulomatous diseases such as tuberculosis, actinomycosis, and syphilis may produce a reaction similar to those of Wegener’s granulomatosis, none of them is likely to produce the characteristic triad—lesions involving the respiratory tract, widespread vasculitis, and glomerulonephritis (Godman and Churg, 1954). Serological tests or identification of the causative organism help to establish the diagnosis.

Sarcoidosis may produce a granulomatous angiitis (Meyer, Foley, and Campagna-Pinto, 1953) but the lesions of sarcoidosis are non-necrotizing.

Traumatic fat necrosis may be associated with many of the changes in the breast found in Wegener’s granulomatosis. However, in the latter the vasculitis is widespread and not confined to the necrotic granulomatous areas.

Lesions of Weber-Christian disease (relapsing febrile nodular panniculitis) have been reported in the subcutaneous tissues of the breast (Binkley, 1939). In one of the cases reported by Carrington and Liebow (1966) a subcutaneous lesion of Wegener’s granulomatosis had originally been interpreted as suggestive of Weber-Christian disease. But the lesions of Weber-Christian disease do not involve the breast parenchyma nor the dermis or the epidermis. In the case presented by Elsner and Harper (1969) and in the two cases reported here the breast parenchyma was involved. In this first case the dermis was also involved by a necrotizing granulomatous lesion with a vasculitis.

Classical polyarteritis nodosa involves small and medium sized muscular arteries. It does not involve the pulmonary arteries and usually does not involve the splenic follicular arterioles (Zeek, 1952). In Wegener’s granulomatosis smaller vessels are involved and granulomata are present, a feature not usually seen in classical polyarteritis nodosa.

Wegener’s granulomatosis, although a rare disease, is worthy of consideration in the histological diagnosis of necrotic breast lesions because of the serious implications attached to the diagnosis.

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References


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