Acta Dermatovener (Stockholm) 57: 533–537, 1977

ELASTOSIS PERFORANS SERPIGINOSA WITH WIDESPREAD ARTERIAL LESIONS: A CASE REPORT

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Abstract. A case is presented of elastosis perforans serpiginosa (EPS) with unilateral dermal lesions, widespread arterial lesions with aortic rupture, and elastosis of the endocardium and bronchiolar walls. Other chronic skin disorders with lesions resembling EPS are discussed, and the arterial lesions compared with some arterial diseases. The findings support a concept of the disease as a focal affection of elastic tissue, not only in the skin, but also in arteries and other organs.

Key words: Elastosis perforans serpiginosa; Arterial changes; Aortic rupture

This unusual clinicopathological entity was first recognized by Lutz in 1952 and reported under the name of keratosis follicularis serpiginosa (15). The disease is characterized by a chronic eruption of circinate or arcuate peripherally spreading lesions, mainly on the neck, in children and young adults, or more disseminated lesions also on the arms and legs (20). They are usually unaffected by treatment but may heal spontaneously (10, 13, 20). The lesions consist of follicle-like pockets containing horny plugs surrounded by small papules (16). Miescher demonstrated that the dermal papillae adjacent to the follicle-like pockets were stuffed with elastic fibres, and that the elastic tissue in the corium perforated the epithelium and could be found inside the keratotic masses in the follicle-like pockets (22, 23, 24). He called the condition elastoma intrapapillare perforans verrucciforme, stressing the elastic tissue changes. More recently the term elastosis perforans serpiginosa (EPS) has been used, as first suggested by Dammert and Putkonen (6).

Anning (2) briefly reported the case of an 18-year-old youth with Marfan's syndrome (20) who died of a dissecting aneurism of the aorta. Histological examination of the aorta revealed degenerated elastic tissue. The association of Marfan's syndrome with such changes of the aorta is, however, well known. Lately we have seen a case of EPS in a young woman who did not show stigmata of Marfan's syndrome, but had widespread focal lesions of the elastic tissue of the larger arteries and aortic rupture.

CASE REPORT

Clinical history

A 30-year-old woman had since childhood suffered from a skin disease which was localized to the right side of the body, mainly on the neck, thorax, abdomen, and proximal part of the extremities. The lesions consisted of follicle-like pockets with central keratinous plugs (Fig. 1), occasionally bordered by small grey papules. She was treated in several hospitals because of recurrent abscesses, with antibiotics, drainage, local excision, and skin transplants. In Nov. 1974 she was admitted to the surgical department of this hospital because of acute abdominal pain radiating to the back. Clinical and X-ray examination suggested an abdominal aortic aneurysm and aneurysmal dilatation of some abdominal arteries. Laparatomy showed rupture of the anterior aspect of the aortic wall and a large haematoma. The abdominal aorta was resected and a Dacron Y-graft implanted. She was reoperated the next day because of bleeding from a lumbar artery. The patient later developed paralysis of the lower extremities. The course was complicated by bronchopneumonia and septicaemia and she died 25 days after the operation.

Post-mortem examination

Autopsy (O. 861/74) was performed 25 hours post mortem. The skin showed macroscopic changes as described above. The aortic wall was ruptured at the site of one of the sutures of the proximal anastomosis. This was as-

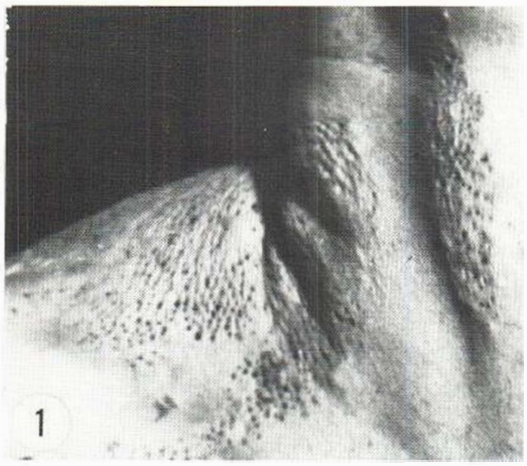

Fig. 1. Right anterior aspect of neck and shoulder showing the follicle-like pockets with central horny plugs, some of them (*right*) with bordering papules.

sociated with a large, partially organized retroperitoneal haematoma. The pelvic organs were embedded in a large haematoma contiguous with the operation scar, the lower end of which was open.

The larger arteries were diffusely dilated, with diminished wall thickness, and there were two saccular aneurysms of the splenic artery. The intima of the upper thoracic aorta showed circinate or polycyclical thickenings (Fig. 2). The thickness of the vessel wall was diminished inside these "rings". These changes were accentuated distally where the intima was rough and the vessel wall mainly thinner than normal.

Microscopical examination

The lungs showed bronchopneumonia and small peripheral lung emboli. Septic emboli were found in all organs, including the medulla spinalis.

Skin sections were taken from symmetrical regions of the body. The aorta and multiple large and medium sized arteries, including cerebral arteries, were sectioned. The material was fixed in 4% formalin and embedded in paraffin. Sections were stained with hematoxylin and eosin (HE), Elastin van Gieson, von Kossa, Masson trichrome, alcian blue at pH 1 and 2.5, alcian blue-cec (7), and periodic acid-Schiff (PAS).

Histology of the skin of the left side of the body showed no changes of the elastic tissue or any other abnormalities. In contrast, the right-sided skin sections showed follicle-like pockets open to the epidermal surface, containing keratotic masses, cell debris, and fibrillar structures staining red with HE and bluish-black with Elastin van Gieson. The rete pegs were elongated and almost surrounded the papillae, some of which were stuffed with elastic tissue (Figs. 3, 4). Elastic fibres were demonstrated inside the epithelium (Fig. 5). The elastic tissue in the corium was focally increased. von Kossa stain did not reveal any calcification of fibres. The form and size of

Fig. 2. The intima of the thoracic part of the aorta showing circinate and polycyclical ridges with central depressed areas (*upper right*) where the vessel wall is thin. Larger lesions are seen below.

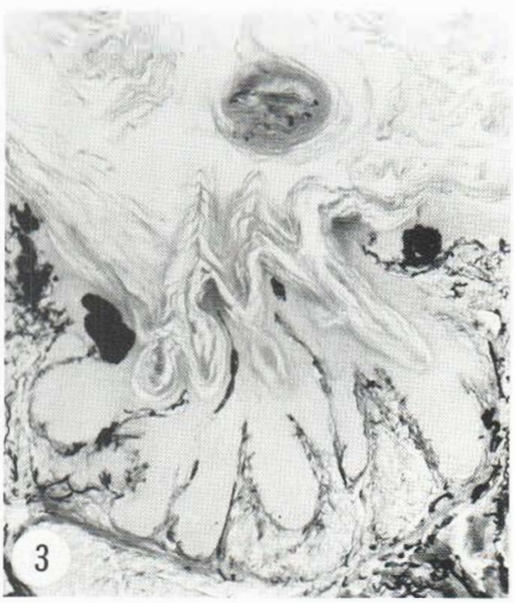

Fig. 3. Epidermal lesion containing keratotic material and elastic fibres. Note the accumulated masses of elastic fibres in the papillae bordering the lesion, and the elongation of the rete pegs (Elastin van Gieson, ×112).

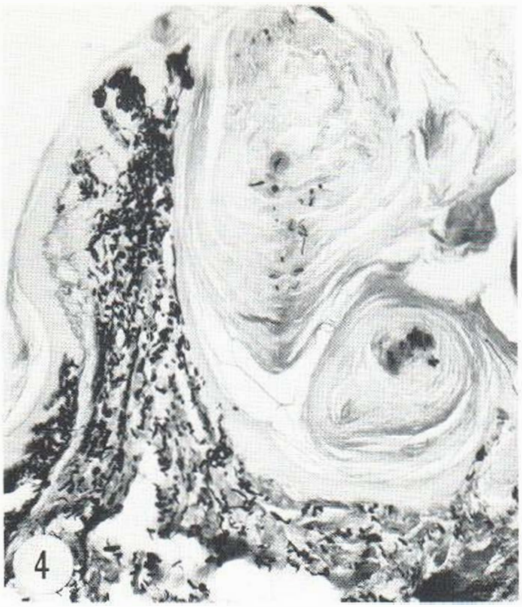

Fig. 4. Cup-formed lesion in the epidermis. The elongation of the rete pegs bordering the lesion is less pronounced (Elastin van Gieson, ×70).

Fig. 5. The elastic fibres seem to lie inside the epithelium of the skin (*arrows*) (Elastin van Gieson, ×280).

individual fibres were difficult to evaluate, but obvious abnormalities were not observed. Staining with alcian blue and PAS of sections from skin areas with elastic lesions unaffected by inflammatory processes did not show any increased positivity, as compared with the normal side.

Examination of the vessels revealed an aorta with focal fragmentation of elastic fibres with an increase in homogeneous basophilic material staining intensely with PAS and alcian blue at pH 2.5 and less so at pH 1.0. The alcian blue-cec procedure showed the strongest positivity at (Mg) 0.1 M. These results indicate the presence of weakly acidic sulphated mucosubstances and acidic mucosubstances carrying carboxyl groups (7). The changes were most pronounced in the distal part of the aorta, where large defects were seen in the media. Other areas showed focal elastosis, particularly of the intima and internal elastic lamina, with some fibrous tissue and smooth muscle cell proliferation (Fig. 6). The media was focally reduced in thickness, presenting as incomplete clefts in the inner vessel wall. Slight focal infiltration of lymphocytes, histiocytes, and a few multinucleated giant cells was present in the ruptured part of the lumbar aorta. The large arteries showed the same focal changes but they seemed to be less pronounced. Some regions of the arteries otherwise unaffected showed elastosis of the internal elastic lamina, with a rough, almost verrucose intima. The cerebral arteries were least affected.

Only slight calcification of the intimal and medial fibres was seen.

Examination of the lung showed elastosis in bronchiolar walls. In the heart, focal fibroelastosis of the endocardium was present.

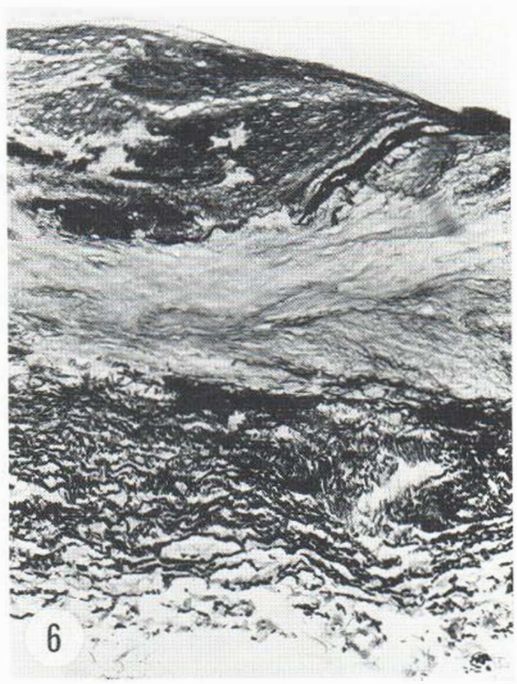

Fig. 6. Distal part of the aorta showing elastosis of the intima and adventitia. The internal elastic lamina is coarse and frayed and melts with the fibroelastic tissue in the intima, which is thicker than normal. The media is partly thinner than normal and almost devoid of elastic tissue (Elastin van Gieson, ×70).

DISCUSSION

The histopathological findings in the skin, with accumulation of elastic fibres in the dermal papillae adjacent to lesions consisting of follicle-like pockets containing keratotic material and elastic fibres, are characteristic findings in EPS (22, 23, 24). In addition, this case shows distinct lesions of elastic tissue of blood vessels, resulting in aortic rupture, and elastosis in some bronchiolar walls and in the endocardium.

Several other chronic skin diseases with similar clinical and histopathological lesions could be excluded. Kyrle's disease (Hyperkeratosis follicularis in cutem penetrans) (1, 14) does not show such changes in dermal and epidermal elastic tissue (11, 28). Widely disseminated skin lesions may be seen in porokeratosis Mibelli, and unilateral lesions have been described (8). However, even if the parakeratotic plugs in this disease may clinically remind one of the lesions of EPS, the histopathological differences are obvious (16). Perforating folliculitis as described by Mehregan (21) does not show any increase in the elastic fibres in the corium, and the dermal papillae do not contain accumulated masses of elastic fibres, in contrast to the present case. The staining for calcium (von Kossa) of the dermal sections was negative and excluded pseudoxanthoma elasticum (9).

Rupture of large arteries is seen in some hereditary conditions affecting the connective tissue, such as Ehlers-Danlos syndrome, Marfan's syndrome, and osteogenesis imperfecta, and the aorta may show generalized degenerative changes with accumulation of acid mucosubstances (4, 18, 19). Our case did not show clinical stigmata of any of these disorders, and the proximal part of the aorta was normal by light microscopy. The focal fibrosis and elastosis of the arteries in the present case are reminiscent of the lesions of fibromuscular hyperplasia (dysplasia) of arteries, but although this disease has been shown to affect multiple arteries (26), it is mainly a constrictive disease of the renal arteries provoking renovascular hypertension (5). Furthermore, the arteries in our case were dilated and thin-walled. Extraordinary elongation and dilatation of arteries is seen in advanced forms of arteriosclerosis (25) but our findings are not compatible with that process. The inflammatory lesions of the lumbar aorta were too slight to represent a primary inflammatory disease such as giant cell aortitis (12), and could have been a reaction to the aortic rupture, as they were not seen elsewhere in the aortic wall or medium-sized arteries.

In a discussion of the pathogenesis of EPS, Miescher suggested that the "intrapapillary elastoma" in the dermal lesions underwent necrobiosis with expulsion of necrotic tissue and elastic fibres through the epithelium, provoking hyper- and parakeratosis, with secondary formation of follicle-like pockets (22, 23). Other authors do not accept the material in the papillae as elastic fibres but believe it could be elastoid degenerated collagen (16, 17, 28). However, enzymatic studies with elastase (27), and electron microscopic examination (3) have confirmed that the fibres are elastic. Miescher's concept that the elastic tissue changes are of primary importance and not secondary phenomena is supported by the present finding of multiple elastic tissue lesions in the arterial and bronchiolar walls as well as in the endocardium.

In EPS the production of connective tissue fibres is focally changed. It does not seem justified, however, to compare it to genetic disorders of the connective tissue, such as Ehlers-Danlos or Marfan's syndrome, as any evidence of heredity or generalized affection of the connective tissue in EPS is lacking.

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Received December 2, 1976

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