

LYMPHSPIRATION PERIPHERAL LYMPHEDEMA AND FIBROSCLEROSIS OF THE LYMPHATIC WALL

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Whereas deposition of collagen in the soft tissues as brawny edema or in the extreme as elephantiasis with lymphostatic disorders is well known, less attention has been directed at ongoing fibrosclerosis of disturbed peripheral lymphatic vessels themselves. Accordingly, we examined by light microscopy cross sections of dorsal pedal lymphatics of 402 patients with primary lymphedema. These included 59 with lymphangiographically demonstrated hyperplasia (i.e. large tortuous channels) and 343 with hypoplasia. 267/402 had secondary lymphedema (84 in the upper extremity). In patients with peripheral lymphedema, lymphatics had much thicker walls than healthy individuals (Fig. 1). The

thickening was transmural with abundant fibrosis in the tunica media, displacing the muscularis (Fig. 1, 2). These fibrosclerotic lymphatic changes were present in both primary lymphedema—either hypoplasia (Fig. 1) or hyperplasia (Fig. 2)—and in severe secondary lymphedema (Fig. 2 inset). Prominent metaplasia of flat endothelium into cubical or cylindrical endothelium was also seen (Fig. 3).

Extensive intramural lymphatic fibrosclerosis probably restricts motility of lymphatics and thereby limits their transport capacity. Such defunctionalization also accounts for limited success of lymphovenous shunts to alleviate peripheral lymphedema in management of “hyperplastic

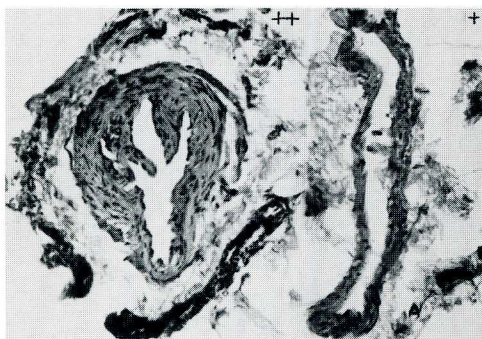


Fig. 1: Hypoplastic lymphatic (left) in patient with peripheral lymphedema demonstrating transmural thickening of the vessel wall with abundant fibrosis in the tunica media. Normal lymphatic (right).

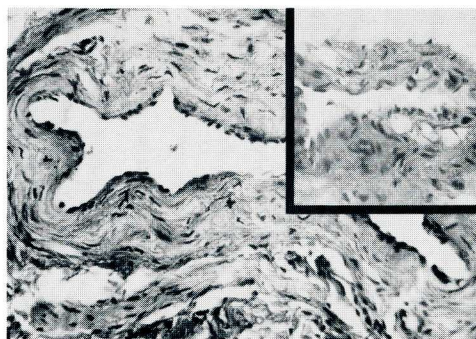


Fig. 2: Hyperplastic lymphatic in primary lymphedema and secondary lymphedema of the arm (inset) demonstrating extensive fibrosclerotic wall thickening.

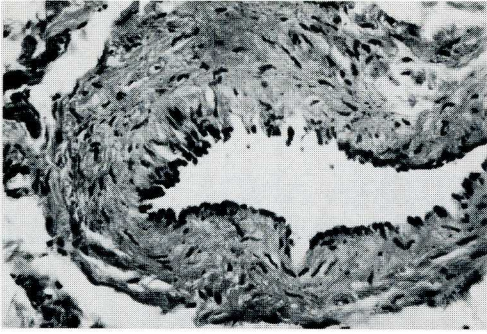


Fig. 3: Hyperplastic lymphatic demonstrating not only dense desmoplasia but also prominent cuboidal metaplasia of ordinarily flat endothelium lining the luminal surface.

lymphedema". Lymph nodal and lymphatic vessel scarring with tissue fibrosis also restricts peripheral lymph flow which further contributes to the onset and progression of extremity edema.