

## *Case Report*

# **Unusual Leiomyoma of the Vulva with Fibroma-Like Pattern and Pseudoelastin Production**

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**Summary.** A unique case of a 71-year old woman with an unusual fibroma-like leiomyoma of the vulva in the region of the left Bartholin's gland is reported. Light microscopically the tumor resembled a fibroma, but electron microscopically the cells corresponded to modified smooth muscle cells. The great number of vessels and their intimate relation to the tumor cells is remarkable. In the intercellular space granular and fibrillar pseudoelastin material was found and thus the presence of collagen type III in addition to type I is suggested. In the region of the right Bartholin's gland a mesenchymal proliferation with similar histological features was seen but there was more resemblance to a conventional leiomyoma. The histogenesis remains obscure so that the designation of these lesions should include light microscopic and electron microscopic appearances as well as the localization.

**Key words:** Vulva – Bartholin's gland – Leiomyoma – Fibroma-like-pattern – Smooth muscle cells – Pseudoelastin – Collagen

## **Introduction**

Leiomyomas of the lower female genital tract are rare growths (Riedel 1964, Kaufman and Gardner 1965, Tavassoli and Norris 1979). Their structure is like typical leiomyomas of the uterus (Hertig and Gore 1960).

We report on a unique tumor of the vulva of a 71-year old woman, which light microscopically resembled a fibroma, producing an elastin-like material. Electron microscopically the tumor cells clearly showed the structure of smooth muscle cells. We are not aware of a similar case in the literature.

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## Material and Methods

A 71-year old woman was admitted to hospital because of a tumor in the region of both Bartholin's glands. At operation a gray-white tissue specimen of 1.5 cm in diameter of firm-resilient consistency was removed from the left side of the vulva. From the right side two tissue specimens of 0.5 cm in diameter were obtained which consisted of interlacing bands of gray-white tissue and yellow adipose tissue.

The tissue material was fixed in 5% buffered neutral formol. The following stains and histochemical reactions were performed: H & E, elastica-Domagk, Goldner's trichrome stain, PAS, alcian blue at pH 0.5, 1.0 and 2.5, alcian blue (pH 2.5)-PAS, colloidal iron reaction after Hale and combined Hale-PAS reaction.

For the purpose of differentiating between elastin and pseudoelastin a staining with 0.5% paraldehyde-fuchsin in 70% ethanol and 1 ml glacial acid per 70 ml dye solution was performed. Furthermore, this procedure was preceded by a thiosulfation and an oxidation with acidified permanganate, resp. After the latter pretreatment and the following paraldehyde-fuchsin staining differentiation by HCl-ethanol was performed (Böck 1977).

For the purpose of electron microscopic examination a small example was taken from the paraffin-embedded material, deparaffinized, postfixed in 2.5% glutaraldehyde for 2 h (buffered with 0.1 M cacodylate buffer at pH 7.2) and  $\text{OsO}_4$  for 1. After embedding in Mikropal semithin sections (stained with toluidine blue) and ultrathin sections (contrasted with lead citrate and uranyl acetate) were made.

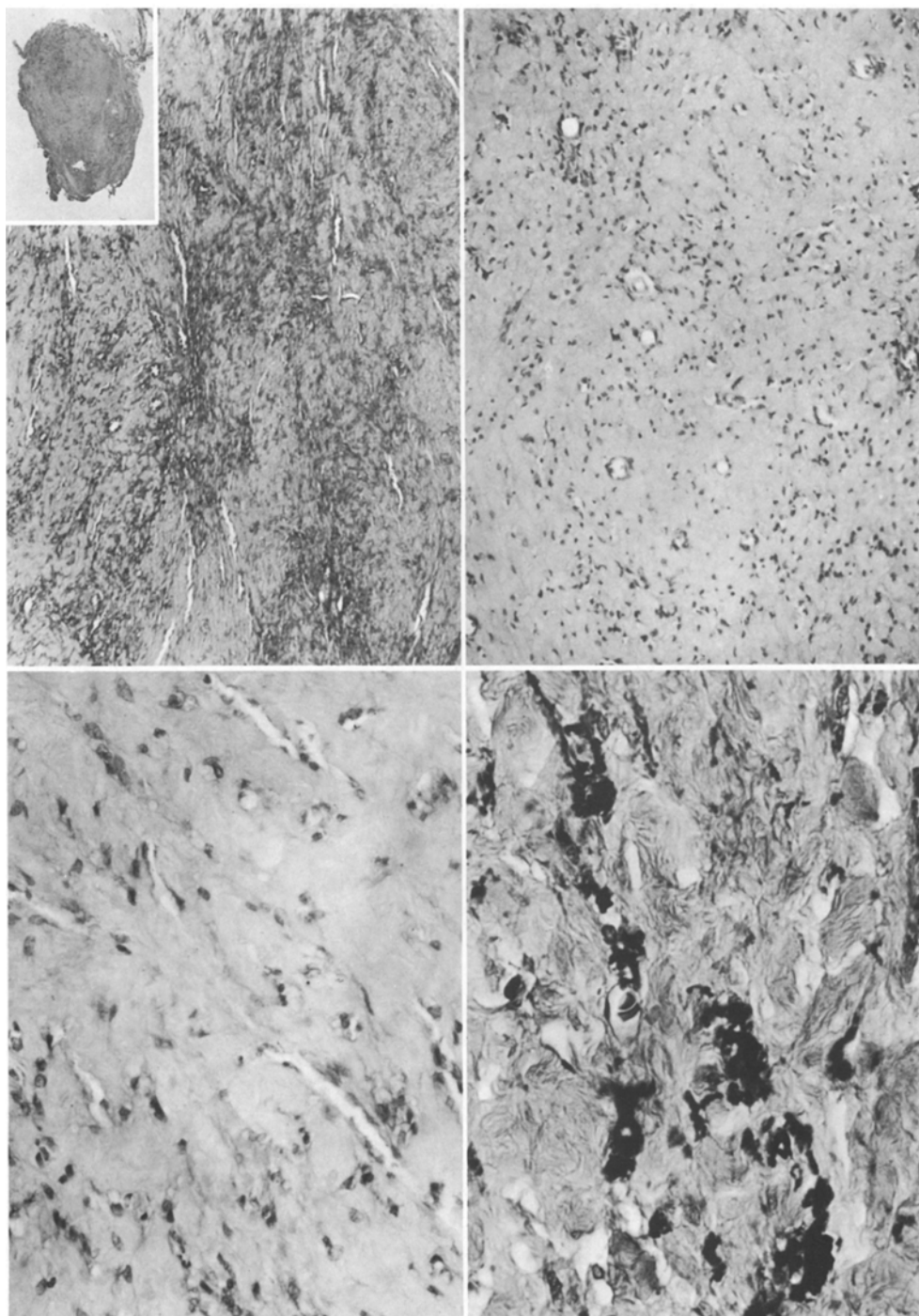
## Results

### *Microscopical and Histochemical Findings*

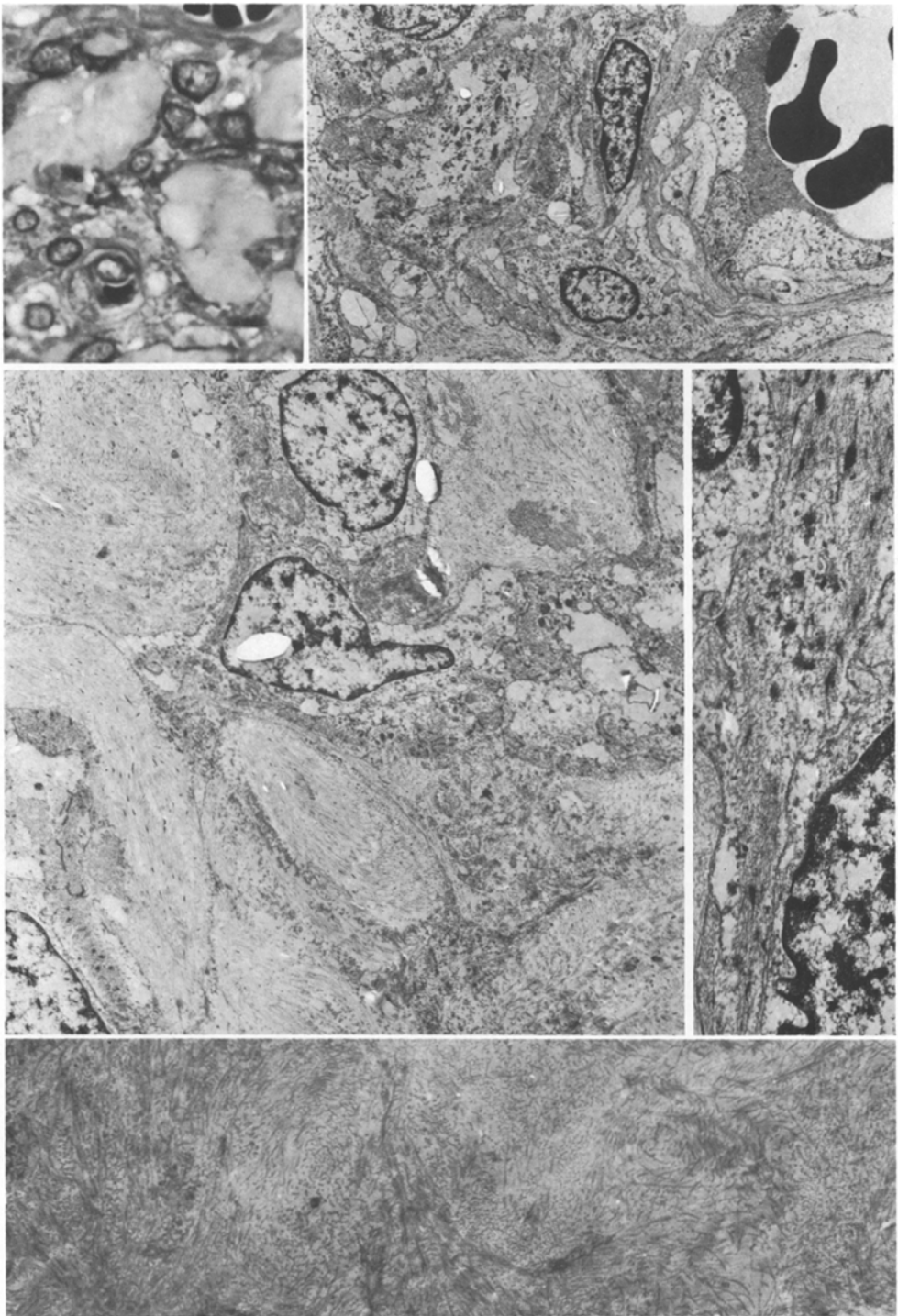
The tumor from the left side was not sharply demarcated from the surrounding connective tissue and thus without a pseudocapsule. The growth consisted of a fiber-rich tissue with a changing density of spindle-shaped cells and with a varying, but in general relatively high vascularity. The cells possessed oval nuclei with a moderate amount of chromatin and occasionally showed a slightly pink cytoplasm in the trichrome stain. However, the typical picture of smooth muscle cells was not present in most areas.

Within cell-rich regions a moderate amount of acid and neutral mucopolysaccharides (glycosaminoglycans) was found. Regions with reduced numbers of cells exhibited a partly hyalinized, fibrous intercellular substance. In these areas collagen fibers were the main constituent of this intercellular substance. Acid and neutral mucopolysaccharides were present in relatively small amounts. In all tumor regions a varying amount of randomly deposited granular and fiber-like material was characteristic in the intercellular space. It could be stained by elastic dyes. In places these fiber-like structures were swollen or clumped. Granular deposits of an elastic stain positive material predominated over fiber-like structures. The latter elements were irregular and could be described as fragmented or thickened, often curved fibers. Pearl-like thickenings along the fiber length were barely detected. Within cell-rich areas elastic stain positive material was observed in greater amounts than in regions with reduced cell numbers. Histochemical methods showed an almost exclusive presence of pseudoelastic material.

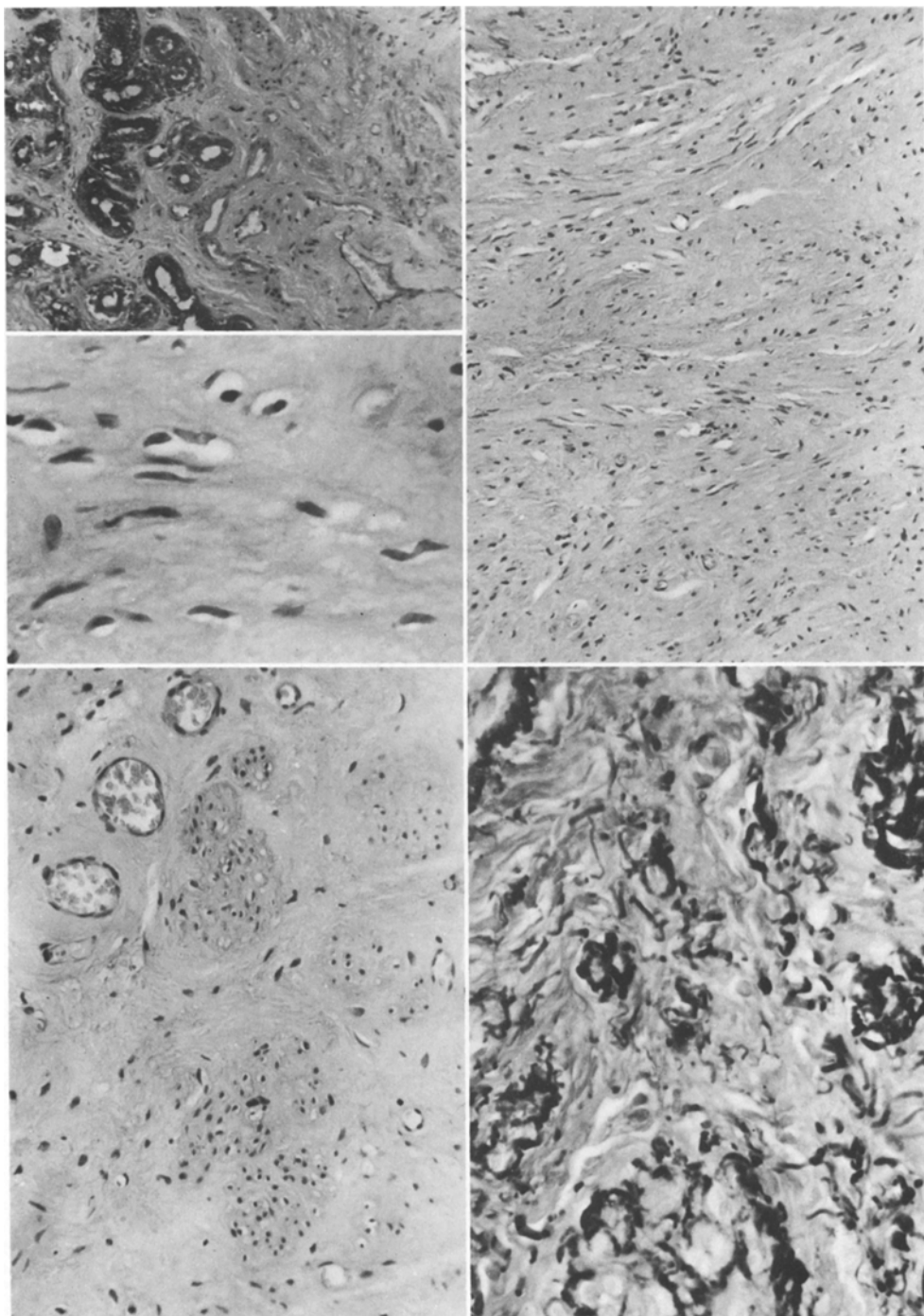
Most of the vessels were of the capillary type, but some arterioles and small arteries could also be seen. A synopsis of the histological features is given in Fig. 1 (cp. also Fig. 2a).



**Fig. 1 a–d.** Histological structures of the left-sided fibroma-like leiomyoma of the vulva. **a** Structure of this lesion at low magnification. Fibrous tissue with varying density of mesenchymal cells and with numerous vessel-like slits is visible (HE, 50:1). Inset: The round tumor without a pseudocapsule. **b** This area shows reduced numbers of mesenchymal cells and a partly hyalinized fibrous intercellular substance. Several capillaries are also present (HE, 120:1). **c** The cells have oval blunted nuclei, cytoplasm is hardly seen. Note the numerous capillary vessels (HE, 260:1). **d** In the intercellular space fiber-like material is encountered which can be stained by elastic dyes. The fiber-like structures are partly curved, irregularly thickened and “fragmented”. Granular elastic positive deposits are inconspicuous (elastic-Domagk, 260:1)



**Fig. 2a-e.** Ultrastructural features of the left-sided fibroma-like leiomyoma of the vulva. **a** The cells are arranged in little complexes and strands. Note the capillaries (semithin section, toluidine blue, 960:1). **b** The capillary has swollen endothelial cells. The vessel is surrounded by proliferated mesenchymal cells which show some characteristics of pericytes and muscular cells (5,600:1). **c** Cellular strands and numerous collagen fibers in interlacing arrangement. Neighbouring cells have plasma membranes which in part ran parallel to each other in a slightly wavy fashion. The intracellular filaments are not clearly visible at this magnification (7,350:1). **d** Detail of cell with some characteristics of a smooth muscle cell. Myofilaments with scattered dense bodies are the prominent organelles (11,300:1). **e** Collagen fibers in an irregular arrangement. Elastic fibers are absent (10,500:1)



**Fig. 3a-e.** Histological features of a leiomyoma from the right side of the vulva. **a** Structures of Bartholin's gland and bundles of smooth muscle cells are visible (PAS, 120:1). **b** The cells show oval nuclei. Some cells possess an epithelioid appearance. Abundant intercellular substance is present (HE, 380:1). **c** There are interlacing bundles of smooth muscle cells. This picture is typical of a leiomyoma (HE, 120:1). **d** Only some bundles of smooth muscle cells are visible. Between them a fibroma-like pattern can be seen (HE, 120:1). **e** Fibres are seen in the intercellular space, stained by elastic dyes. They are localized in the region of muscle bundles (→) and in fibroma-like areas (elastica-Domagk, 260:1)

The material removed from the right side of the vulva revealed similar histological features, but the occurrence of interlacing fascicles of spindle-shaped smooth muscle-like cells was more pronounced. Epithelial remnants of Bartholin's gland were demonstrated, enclosed in this mesenchymal proliferation (Fig. 3).

### *Electron Microscopic Observations*

The cells of the left-hand tumor were of an elongated, but often clearly irregular form. They formed small complexes or lattice-shaped cellular strands with abundant intercellular connective tissue fibers. The cell membranes of neighbouring cells often ran parallel to each other leaving only a narrow intercellular space. Vascular (capillary) channels were frequently visible, they were closely surrounded by the tumor cells and seemed to be an integral part of the tumor (Fig. 2b).

The cellular nuclei were partly indented and often equipped with an activated nucleolus. The cytoplasm contained myofilaments which corresponded to actin filaments and measured 50–60 Å in diameter. The filament bundles also showed changing amounts of scattered small dense bodies (Fig. 2d). Numerous attachment sites of myofilaments with localized thickenings of the plasma membrane were visible. Free ribosomes and some slender tubules of rough endoplasmic reticulum were also found. A basement membrane enveloped some tumor cells especially in the vicinity of vessels, but mostly incompletely. Around the vessels pericyte-like cells could be seen interdigitating with smooth muscle-like cells (Fig. 2b).

The intercellular fibers varied in their thickness between 200 and 1,000 Å. Partially cross-banding of mature collagen fibers (600–700 Å) could be detected. But the most fibers were without clear periodicity and strongly interwoven (Fig. 2e). Typical elastic fibers or fiber-like material was not to be observed ultrastructurally.

### **Discussion**

The tumor of the left labium majus presented in this paper bears light microscopic resemblances to a fibroma, but is ultrastructurally composed of smooth muscle-like cells. Their electron microscopic appearances are within the range of their morphological variation of other sites, e.g. in leiomyomas of the uterus (Fritzsche et al. 1978). This evidently benign growth has a relatively rich vasculature. In spite of the localization in the region of the left Bartholin's gland no epithelial elements are detectable. On the right side of the vulva a similar tissue proliferation took place, but here the myoma-like aspect was more obvious and epithelial components of the Bartholin's gland could be seen.

The abundant intercellular substance can only have been produced by the tumor cells themselves. The major part of the extracellular fibrils are collagen fibers. The fiber-like and granular material visualized by elastica stains is not

of true elastic nature. From our histochemical investigations the main part of these substances is clearly a pseudoelastic material. In places these structures remind of the intercellular material in elastofibromas (Järvi and Saxen 1961), but the arrangement and, in particular, the amount of this material do not correspond to a real elastofibroma (Järvi 1979).

It is well known that several fiber types and mucopolysaccharides can be synthesized by different connective tissue cells (Rhodin 1967). On the basis of our histochemical results and considering the electron microscopic picture it appears that both collagen type I and collagen type III is present. Collagen type III may be a product of smooth muscle cells (Scott et al. 1977) or myofibroblasts (Gabbiani et al. 1976). The latter cells have intermediate features between fibroblasts and smooth muscle cells (Gabbiani et al. 1971). Electron microscopically we found cells which seemed to be somewhat modified smooth muscle cells, so that the vulva tumor presented here cannot be named a "myofibroblastoma" (Wirman 1976; Ramos et al. 1978; Bhawan et al. 1979).

The histogenesis of this tumor-like proliferation remains obscure. Three general explanations seem to be possible:

1. The tumor develops from the muscoli arrectores pilorum as in certain leiomyomas of the skin.
2. The tumor arises from cells of vascular walls in analogy to angiomyomas or from smooth muscle cells of the erectile tissue, and
3. The stem cell for this tumor is localized in the Bartholin's gland; it could be either a smooth muscle cell of the glandular stroma or a myoepithelial cell.

The topographical localization and epithelial cells in the lesion of the right side appear to favour the last possibility. On the other hand, the numerous vascular channels and their intimate relation to the tumor cells point to a multipotent vascular cell as the precursor cell, as with uterine leiomyomas (Honoré 1977).

Because our findings do not allow a definite histogenetic conclusion and the lesion does not fit into the usual classification of soft tissue tumors (Enzinger et al. 1969) we can only suggest a descriptive classification. The preliminary designation should include the light microscopic and electron microscopic appearances as well as the localization.

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