PATHOLOGICAL STUDY OF TUMORS OF FIBROUS TISSUE AND THEIR RELATED CONDITIONS

Thesis

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By

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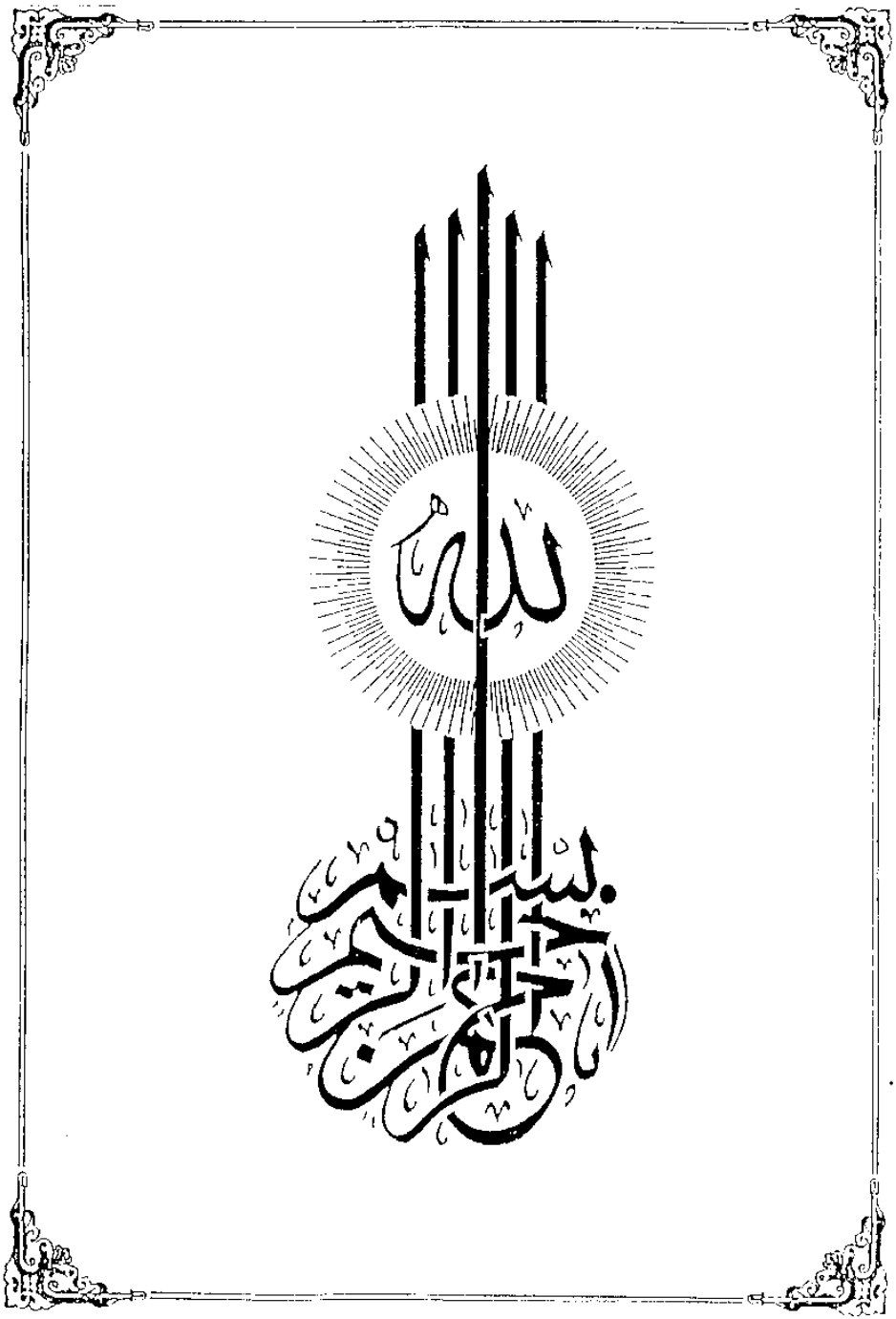

CONTENTS

INTRODUCTION

Fibrous tumors and tumor-like lesions form a large and diverse group of distinct entities that differ greatly in their behavior and at times cause considerable difficulties in diagnosis.

Some are perfectly benign lesions that remain localized and do not recur even after simple excision. Others are poorly circumscribed, grow in an infiltrative manner and tend to recur unless they are widely excised. Still others are frankly malignant tumors that recur and metastasize in a high percentage of cases.

Much new information about fibrous growths has been recorded in the last few years. A great deal has been learned about the true fibroblastic tumors. Very cellular non metastasizing fibroblastic tumors are often identified by the term fibrosarcoma. This leads to an entirely unjustifiable picture of the frequency of malignant fibrous tumors in as much as fibrosarcomas are actually relatively uncommon.

The study of the cellular origin of certain tumors has made it possible to reclassify a number of them and has facilitated the study of their behavior.

CLASSIFICATION OF TUMORS OF FIBROUS TISSUE AND THEIR RELATED CONDITIONS

Many classifications have been proposed.

The classification of the W.H.O. (1969) is as follows:

A) Fibromas:

1) Fibroma durum.
2) Fibroma molle (fibrolipoma).
3) Dermatofibroma (histiocytoma, sclerosing haemangioma).
4) Elastofibroma (dorsi).

B) Fibromatosis:

1) Cicatricial fibromatosis.
2) Keloid.
3) Nodular fasciitis (Pseudosarcromatous fibromatosis).
4) Irradiation fibromatosis.
5) Penile fibromatosis (Peyronie's disease).
6) Fibromatosis colli.
7) Palmar fibromatosis.
8) Juvenile aponeurotic fibroma (calcifying fibroma).
9) Planter fibromatosis.
10) Nasopharyngeal fibroma (juvenile angiofibroma).
11) Abdominal fibromatosis (abdominal desmoid).
12) Fibromatosis or aggressive fibromatosis (extra-abdominal desmoid).
13) Congenital generalized fibromatosis.

C) Dermatofibrosarcoma Protuberance:

D) Fibrosarcoma:

Ashley (1978) mentioned the following classification in his text-book "Evan's Histological Appearances of Tumors":

- Keloid fibromatosis.
- Desmoid tumor.
- Nodular fasciitis.
- Juvenile aponeurotic fibroma.
- Nasal and Nasopharyngeal fibroma.
- Fibromatosis: * Palmar and planter fibromatosis.
  * Congenital generalized fibromatosis.
  * Fibromatosis Hyalinica multiplex juvenalis.
  * Congenital subdermal (fibromatous tumors).
- Elastofibroma.
- Intra-mural fibroma of heart.
- Fibrous tumors of the dermis:
  * Histiocytoma and malignant histiocytoma.
  * Progressive Recurring Dermatofibroma (Dermatofibrosarcoma Protuberance).
- Fibrosarcoma.

The Armed Forces Institute of Pathology (1982) Classified tumors and tumor-like lesions of the fibrous tissue as follows:

- Fibroma
- Fibromatosis:
  - * Juvenile aponeurotic fibroma (calcifying fibroma).
  - * Fibromatosis colli.
  - * Penile fibromatosis.
  - * Progressive myositis fibrosa (hereditary polyfibromatosis)
  - * Pseudosarcomatous fasciitis (nodular fasciitis).
- Elastofibroma
- Fibrous histiocytoma
- Xanthomatoses
- Fibrosarcoma.

Enzinger and Weiss (1983) classified tumors of the fibrous tissue and related conditions as follows:

I. Tumors and tumor-like lesions of fibrous tissue

A) Benign:

1- Fibroma

2- Nodular fasciitis.

3- Proliferative fasciitis.

4- Proliferative myositis.

5- Fibroma of tendon sheath.

6- Elastofibroma.

7- Nasopharyngeal fibroma.

8- Keloid.

B) Fibrous tumors of infancy and childhood:

1- Fibrous hamartoma of infancy.

2- Infantile myofibromatosis (solitary, multicentric)

3- Fibromatosis colli.

4- Infantile digital fibromatosis.

5- Infantile fibromatosis (desmoid type).

6- Gingival fibromatosis.

7- Calcifying aponeurotic fibroma.

8- Hyaline fibromatosis.

C) Fibromatoses:

1- Superficial fibromatoses:

a- Palmar and planter fibromatosis.

b- Penile (Peyronie's) fibromatosis.

c- Knuckle pads.

2- Deep fibromatoses:

a- Abdominal fibromatosis.

b- Extra-abdominal fibromatosis.

c- Intra-abdominal fibromatosis.

d- Mesenteric fibromatosis (Gardner's syndrome)

e- Postradiation fibromatosis.

f- Cicatricial fibromatosis.

D) Malignant

1- Adult fibrosarcoma.

2- Congenital and infantile fibrosarcoma.

3- Postradiation fibrosarcoma

4- Cicatricial fibrosarcoma.

II. Fibrohistiocytic tumors:

A) Benign:

1- Fibrous histiocytoma:

a) Cutaneous (dermatofibroma).

b) Deep.

2- Atypical fibroxanthoma.

3- Juvenile xanthogranuloma.

4- Reticulohistiocytoma.

5- Xanthoma .

B) Intermediate:

1- Dermatofibrosarcoma protuberance.

2- Bednar tumor.

C) Malignant:

1- Malignant fibrous histiocytoma:

a) Stori-form-pleomorphic.

b) Myxoid (myxofibrosarcoma).

c) Giant cell (malignant giant cell tumor of soft parts).

PATHOLOGY OF FIBROUS TISSUE TUMORS AND TUMOR-LIKE CONDITIONS

A) FIBROMAS

The term fibroma was previously applied by Bartlett et al. (1961) to any benign fibrous growth whether or not it was a congenital malformation, reparative tissue, or a real neoplasm.

In order to avoid this loose application of the term; Mackenzie (1964) defined it as a pedunculated or filiform congenital malformation composed of normal fibrous elements of the corium covered by epidermis. These are harmless, seldom removed except for cosmotic reasons, and therefore are not often seen by the pathologist. Allen and Enzinger (1970) described fibroma as a benign simple tumor arising from fibrous tissue. It appears as an oval or rounded well-encapsulated tumor. It may be "hard" if it contains much collagen fibres or "soft" if it is very cellular.

The sites of fibromas as mentioned by Lazarus and Trombetta (1982) are mainly the skin, subcutaneous tissue, fibrous sheaths of muscles, internal organs e.g. ovary, kidney, gastrointestinal tract and nasopharynx. Nathan and Zocholl (1983) added that fibroma may be found rarely in the ureter.

Microscopically; the cells of the tumor are fibrocytes; their nuclei are long, narrow and densely stained, the cytoplasm so scanty as to be hardly seen, and mitoses are very rare. Bundles of dense collagen separate the cells.

Fibroma durum:

Kauffman and Stout (1961) defined fibroma durum as a benign frequently pedunculated, well-circumscribed dense growth of fully matured and richly collagenous fibrous connective tissue occuring on the body surface and mucous membranes. They are found mainly in hands and feet but Mackenzie (1964) added that it may be found attached to tendons, tendon sheaths, joint capsules or in muscles and deep fibrous tissues.

According to Rios-Dalenz et al. (1965), the microscopic appearance of fibroma durum is quite variable, depending upon the amount of fibroblasts and multinucleated giant cells of foreign body type (histiocytes) with or without lipids or hemosiderin. Mehregan (1981) added that there may be occasional inflammatory cells.

Fibroma molle (Fibrolipoma):

Enzinger et al. (1969) defined it as a benign and usually pedunculated growth made up of a mixture of mature

fibrous connective tissue and adult-type fat occuring on the body surface.

Intramural fibroma of the heart:

This is a fibroma which usually occurs in the left ventricle and causes sudden death in infancy and early childhood (Clay and Shorter, 1957).

Dermatofibroma:

Tellem et al. (1965) defined dermatofibroma as a benign, non-encapsulated, superficial lesion, characterized by an intimate mixture of histiocyte and fibroblast-like cells associated with varying amounts of collagen and thin-walled blood vessels. Frequently, lipid macrophages and siderophages are prominent features of the lesion. It includes:

A) Benign histiocytoma:

Histiocytoma is now the common term used to indicate certain benign dermal fibrous tumors believed to be of histiocytic origin. Kauffman and Stout (1961) mentioned that the histiocyte can act as a facultative fibroblast and may conceal its true nature by appearing to be a simple fibroblastic tumor with no obvious foamy cytoplasm containing lipids. Hudson and Winkelmann (1972) stated

that the tumor may arise from primitive mesenchymal cells acquiring certain morphological features.

The vascular nature of this type of lesion has been stressed even by earlier investigators as Dawson (1948) who mentioned that the histiocytoma results from a succession of events resulting in obliteration of a capillary angioma. However, Fu et al. (1975) suggested that the blood vessels are only a secondary feature and not an integral component of histiocytoma. Evidence of a vasoformative process is lacking and it is more likely that the spindle cells and connective tissue fibres of the lesion indeed represent histiocytes which function as facultative fibroblasts.

The tumors are common in adults between the ages of 20-40 years but rare in children, though Orzello et al. (1963) have reported 39 examples in subjects less than 16 years of age.

Most commonly this tumor occurs in the dermis and superficial subcutis mainly on extrimities, buttocks, shoulders and face. Webb et al. (1974) added that it may be found in soft tissue and sporadically in parenchymal organs.

The tumors occur as slowly growing nodules which may be multiple but usually are solitary. They are well demarcated, non-encapsulated, rarely exceeding 2 to 3 cm. in diameter. They vary in colour and may be black, purple, red or yellow, or any mixture of these colours, depending on their vascularity, lipid and hemosiderin content (Kempson and Kyriakos, 1972).

Orzello et al. (1963) described histiocytoma as a mixture of fibroblastic and histiocytic cells that are often arranged in a cartwheel or storiform pattern showing uniformity of size and shape containing a variable amount of lipid with minimal or absent mitotic activity. The cells are separated by delicate collagen network surrounding individual cells and accompanied by varying numbers of inflammatory cells, foam cells, and siderophages. Jacobs et al. (1975) added that the tumor may also show multinucleated giant cells of foreign body or touton type as a typical feature or rarely osteoclast-type giant cells. They noticed, as well, that repeated hemorrhage and ulceration are common, and with occurrance of hemorrhage; hemosiderin accumulations are seen. Carstens and Schrodt (1974) noticed that in some histiocytomas, or in different parts of the same histiocytoma, the vessels and stroma exhibit a striking hyalinization, a feature that has led to use of the misnomer "sclerosing haemangioma".