Fibromatosis of the Extremeties

Essay for partial fulfillment of master degree in orthopedic surgery

Submitted by

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Abstract

Desmoid's tumours (DTs) are rare neoplasm arising from fascial or deep musculo-aponeurotic structures.

They are localized in the abdominal wall, the bowel, and the mesenteryn (associated with familial adenomatous polyposis) or in extra-abdominal sites, such as the trunk and the extremities. The incidence of DTs ranges from 2% to 4% per million; almost Optimal management has not been clearly defined. Surgery is generally considered the treatment mainstay.

Key words

Desmoid tamour (Dts)

Fibraomatosis

management

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LIST OF CONTENTS

- **ARABIC SUMMARY .....**

LIST OF FIGURES

Clinical types of fibromatosis

Introduction

The fibromatoses are a diverse group of soft-tissue lesions that occur at different ages and anatomic locations and that have common histopathologic features. They are composed of spindle-shaped fibrous cells that are separated and surrounded by abundant collagen material with rare mitoses. The term 'Fibromatosis' was introduced for the first time by Arthur Purdy Stout [(1)].

Their biologic behavior is intermediate in aggressiveness between benign fibrous lesions and fibrosarcoma. There is a strong tendency toward local recurrence; however, these lesions never metastasize. The lesions have a well-circumscribed or infiltrative margin and the degree of cellularity is widely variable[(1)].

The term **desmoid** is derived from the Greek "desmos" (meaning tendon-like). Desmoid tumors (also known as aggressive Fibromatoses) were first described as arising from the anterior abdominal wall. In 1923, Nichols reported on extra-abdominal desmoid tumors that had developed at other sites, and

it is now recognized that desmoid tumors can arise from any mesenchymal tissue [2,28].

Desmoid tumours are relatively rare and, hence, they are seen infrequently by most physicians. They account for about 0.03% of all neoplasms, and less than 3% of all soft tissue tumours. The estimated incidence in the general population is 2 to 4 per million populations per year, almost half of them occur in the extremities and trunk [3].

Although aetiology is poorly defined, a connective tissue growth disorder has been supported [4].

Several etiologic factors has been implicated in the pathogenesis of desmoid tumours, among them, genetic abnormality, steroid sex hormones, and trauma. Desmoid tumours have been associated with hereditary syndromes (Gardner's syndrome), pregnancy, especially second pregnancy, and endogenous/exogenous female sex hormones in adults. However in children, these associations are difficult to establish[3].

ψ *Clinical types of fibromatosis*

Based on X-chromosome inactivation pattern, desmoid tumours appear to be monoclonal disorders, indicative of a neoplastic rather than an inflammatory fibrous reactive process[(5)].

Desmoid tumors are observed in nearly every part of the body. Numerous sites in the trunk and extremities have been identified, and two forms of the disease are distinguished; an extra-abdominal and an intra-abdominal form [(6)] .

Generally, desmoids usually occur in three locations; the extremities (commonly around the limb girdles or the proximal extremities); the abdominal wall (commonly found in women, especially during and after pregnancy); and the bowel wall and mesentery (often associated with Gardner syndrome) [(6)].

In approximately 10% of the cases, multicentric disease has been reported. Multicentricity is defined as the presence of disease in more than one location at the time of diagnosis [(6)].

Multicentric lesions tend to be in the same limb or anatomic region, and it is possible that many cases of distant or local recurrence of primary lesions within the same region represent multicentric disease that were not clinically apparent at the initial diagnosis [(6)].

These tumors are extremely difficult to treat. Because they are locally aggressive, some physicians have considered them to be low-grade sarcomas. Such an assumption may lead to overtreatment. In contrast, some tumors may be undertreated, and this may lead to problems with local control [(2)].

The fibromatoses are commonly divided into two major groups: superficial (fascial) and deep (musculoaponeurotic). The superficial fibromatoses are usually small, slow-growing lesions that arise from fascia and aponeuroses. The deep fibromatoses are larger and may grow rapidly. They are more aggressive and likely arise from the deep fascia, muscle and aponeurotic tissue[(7)].

The two major groups are:

1- Superficial (fascial) fibromatosis:

features: 1.slow growing tumour; 2.small size; 3.arise from fascia or aponeurosis; 4.less aggressive

A. palmar fibromatosis (Dupuytren`s contracture)

B. planter fibromatosis (Ledderhose`s diseasee)

C. Penile fibromatosis (Peyronie`s diseasee)

D. Knucle pads.

2-Deep(musculoaponeurotic)fibromatosis:

features:1.rapidly growing tumour; 2.usually attain large size; 3involve deeper structures (musculature of trunk and extremities).

A. Extraabdominal fibromatosis (extraabdominal desmoid)

B. Abdominal fibromatosis (abdominal desmoid)

C. Intraabdominal fibromatosis (intraabdominal desmoid) [(8)].

Superficial Fibromatoses

Palmar Fibromatosis;

Palmar fibromatosis (Dupuytren disease) is the most common type of fibromatosis, with a prevalence of 1%–2% in the general population [(9)].

Older patients are most frequently affected, with 24% of people over the age of 65 years demonstrating such lesions. Palmar fibromatosis is bilateral in 42%–60% of cases [(9)].

These lesions are three to four times more common in men and are rarely seen in children [(9)].

The aetiology of palmar fibromatosis still remains obscure. However, there is little doubt as to the existence of a hereditary disposition. Ling (1963) suggested that a single gene, behaving as a Mendelian dominant, was likely to be involved [(9)].

Although several investigators mentioned that palmar fibromatosis was more common in patients having diabetes mellitus , chronic alcoholism, chronic liver disease or epilepsy , the exact significance of these coexisting diseases was unknownThe other causative factor may be minor trauma

producing partial rupture of palmar fascia, followed by reparative fibroblastic proliferation [(10)].

The clinical presentation is subcutaneous nodules on the palmar surface at the level of the distal crease of the hand (Fig 1). These nodules slowly progress to fibrous cords or bands that attach to and cause traction on the adjacent flexor tendons, resulting in flexion contractures of the digits. The fourth and fifth rays are most commonly affected, followed by the second and third rays [(9)].

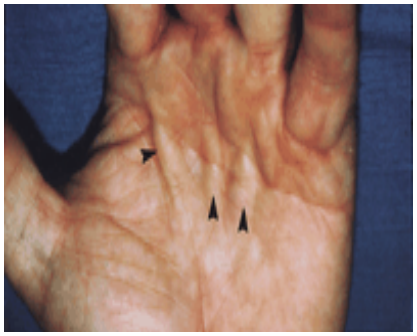

Figure 1. *Palmar fibromatosis. Clinical photograph shows multiple subcutaneous cords and nodules (arrowheads) at the bases of the second through fifth digits, which resulted in flexion contractures.(1)*

Plantar Fibromatosis;

Plantar fibromatosis is a benign fibroblastic proliferative disorder of the plantar fascia that may present as an isolated fibroma, desmoplastic fibroma, juvenile aponeurotic fibroma, or

generalized fibromatosis [11]. Plantar fibromatosis (Ledderhose disease) occurs most frequently between the ages of 30 and 50 years, with bilateral involvement seen in 20%–50% of cases. Concomitant palmar fibromatosis is seen in 10%–65% of patients[11].

these lesions tend to be asymptomatic and to be discovered by palpation. Clinically, the diagnosis is more obvious if the nodules are multiple, either unilaterally or bilaterally [11].

Previous reports have cited a greater prevalence in male, white, and middle-aged patients, but the literature is inconsistent. Patients who have epilepsy have been thought to be predisposed to the condition [11].

These lesions develop most commonly in the medial aspect of the plantar aponeurosis. They usually manifest as one or multiple firm, fixed, subcutaneous nodules, which can extend to involve the skin or invade the deep structures of the foot [11].

Unlike the lesions of Dupuytren contracture, the lesions of plantar fibromatosis seldom have an inflammatory proliferative phase and tend to remain asymptomatic throughout