Clinico-pathological study and management of Fibromatosis

Thesis

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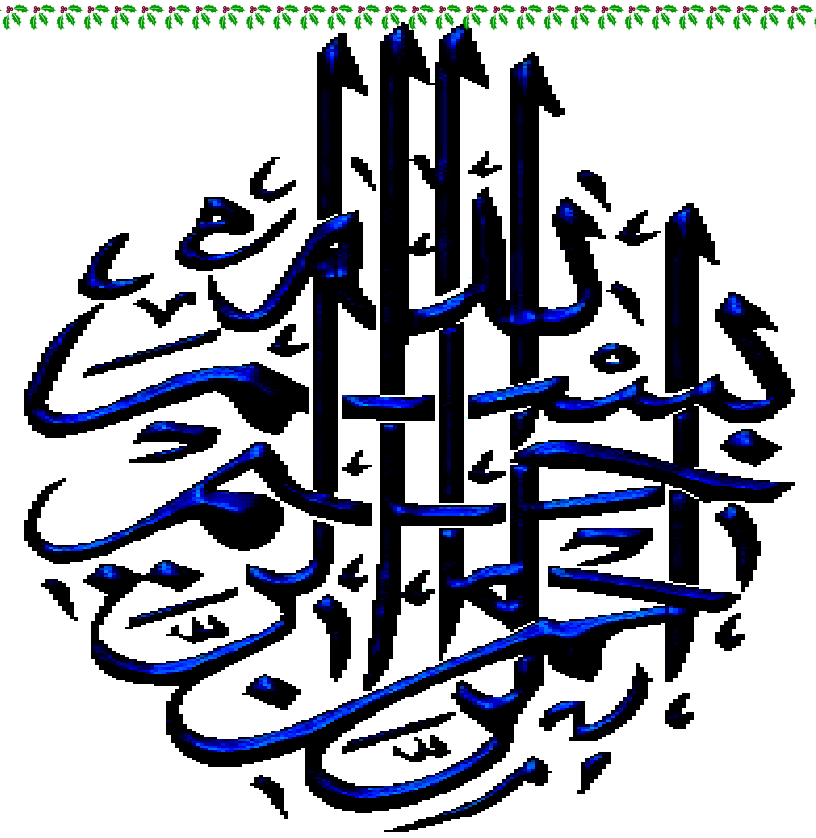

قَالُوا سُبْحَانَكَ لا عِلْمَ لَنَا إِلَّا مَا
عَلَّمْتَنَا
إِنَّكَ أَنْتَ الْعَلِيمُ الْحَكِيمُ

صدق الله العظيم
سورة البقرة:٣٢

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Abstract

Background: Aggressive fibromatosis (AF) is a rare soft-tissue tumour. It is histologically benign but locally aggressive and destructive. Surgery is the mainstay of treatment. This was a review of patients treated at the National Cancer Institute, Cairo University between 2005 and 2007.

Methods: Records of patients with extraabdominal (26) and abdominal wall(11) and pelvic(3) AF were studied to determine patient, tumour and treatment characteristics and outcome.

Results: Twenty three (70%per cent) of 33 patients developed recurrence (after surgical excision) after a median follow-up of 32 months. Recurrence was correlate with surgical margin. 96% of recurrence occurred in first three years after excision.

Conclusion: Wide excisional, function-preserving surgery is the goal in treatment of AF. Regular follow up is required after excision especially in first three years after excision.

Key words : Desmoid tumor , Aggressive fibromatosis

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ABBREVIATIONS

AF .....Aggressive Fibromatosis
APC……Adenomatous Polyposis Coli
BIRADS……Breast Image Reporting And Data System
cAMP……Cyclic adenosine monophosphate
CM……Centimeter
CUM……..Cumulative
CT……..Computed Tomography
DFS……..Disease free survival
DMSA……Dimercapto Succinic Acid
DT……..Desmoid Tumor
ER……..Estrogen Receptors
FAP……....Familial Adenomatous Polyposis
Fig……..Figure
FNAB……Fine Needle Aspiration Biopsy
GY……Gray
GIST……Gastriintestinal stromal tumor
HPF……...High Power Field
IFN……..Interferon
LIQ……....Lower Inner Quadrant
ILP……Isolated limb perfusion
MMP……Metallomatrix proteinase
MRI……...Magnetic Resonance Imaging
MSKCC……Memorial Sloan Ketering Cancer Center
NCI……...National Cancer Institute
NO=n……Number
NSAIDs……Non Steriod Antiinflamatory Drugs
ODC…….....Ornithine decarboxylases
PDGF-B……Platelet Derived Growth Factor- Beta
PR…….....Progesteron Receptors
RF……Radiofrequancy
RMS ……Rhabdomyosarcoma
RTH …… Radiotherapy
SE……..Stander error
SM……Safety Margin
SPSS……Statistical Package for Social Sciences
TC……..Technetium
TGFB……Tumor growth factor beta
TIMO……Tissue inhibitors metalloproteinase
TNF……Tumor necrosis factor
VS……..Versus
-ve……..Negative
+ve……..Positive
WHO……..World Health Organization

Fibromatosis are slow-growing, histologically benign tumors of fibroblastic origin with variable biologic behavior. Despite their benign appearance, they are locally aggressive and invasive to surrounding anatomic structures. Because of these characteristics, they are sometimes classified as low-grade fibrosarcoma. The fibromatosis can be divided into two major groups with several subdivisions: superficial (facial) and deep (musculoaponeurotic).

A-Superficial (fascial) fibromatosis:-

Palmer fibromatosis (Dupuytren's disease)
Planter fibromatosis (Ledderhose's disease)

Penile fibromatosis (Peyronie's disease)
Knuckle pads

B-Deep fibromatosis (musculoaponeurotic):-

Extra abdominal fibromatosis
Abdominal fibromatosis
Intra abdominal fibromatosis:-
Pelvic fibromatosis
Mesentric fibromatosis
Mesentric fibromatosis in Gardener syndrome

Superficial type: are slow growing and small in size that arise from the fascia or aponeurosis and rarely involve deep structures. The clinical course is usually divided to early, rather than cellular proliferative phase and late richly collagenous regressive or contractile phase**.**

Deep type (Desmoid tumor, Aggressive fibromatosis):- are rapidly growing tumors that often attain large size, their biological behavior tending to be more aggressive than superficial type, high recurrence rate and involve deep structures particularly the musculature of trunk and extremities.

Although precise etiology is unknown. They have been associated with trauma, 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.

Magnetic resonance imaging is the modality of choice for the diagnosis and the evaluation of the tumors. Current management involves a multidisciplinary approach. Wide margin surgical resection remains the main treatment modality for local control of the tumor. Amputation should not be the initial treatment, and function-preserving procedures should be the primary treatment goal. Adjuvant radiation therapy is recommended both for primary and recurrent lesions. Chemotherapy and non-cytotoxic drugs may be used for recurrent or unresectable disease.

Aim of this study

1- Retrospective evaluation of patients with fibromatosis presented and treated in the department of surgery at the National Cancer Institute, Cairo University in the period from January 2005 till December 2007.

2- Review the literature in management of fibromatosis.

3- Evaluation of morbidity of disease and treatment.

4- Quality of life of patients with and without treatment.

5- Management of patients with recurrent fibromatosis

(Superficial Fibromatosis)

Palmer fibromatosis (Dupuytren´ s disease)

Although it is named for French surgeon Baron Guillaime Dupuytren who reported this condition in 1831, Felix Plater was probably the first to describe the lesion in 1614. It is characterized by a nodular fibroblastic proliferation that occurs in volar surface of the hand and histologically closely resembles other forms of fibromatosis. The lesion appears to progress through a series of clinical and histological stages ultimately result in flexion contracture of fingers, a complication that usually necessitates surgical therapy (**Weiss and Goldblum 2001)**. There is no correlation with trauma or occupation and it lacks B catenin or APC mutation. It tends to be familial **(Murry et al 2008)**. Rarely similar lesion develops in dorsum of hand or fingers **(Fletcher 2007).**

Clinical findings:

It is a relatively common condition that tends to affect adult men, with a rapid increase incidence with advancing age. It has been estimated that almost 20% occur in old age >65years. Patient younger than 30 years is rarely affected. It affects men more than women (3-4 times). It occurs commonly in northern Europe and those parts of the world that setteled by northern Europeans. It is rare in black population **(Sladicka et al 1996, Urban et al 1996).**

There is slight predilection for right palmer surface but in 50% may occur bilaterally. It is most prominent on the ulnar surface of the palm affecting 4^{th} and 5^{th} fingers. The thumb and index fingers are least often affected. The onset of disease is insidious and the initial manifestation usually occurs as an isolated asymptomatic firm nodule in palmer surface of the hand (because lack of symptoms in this stage, many patients ignore the nodule and seek medical advice). Although clinical progression does not invariably occur, several months or years after the original nodule, cord-like induration(Fig 1) or bands develop between nodules and adjacent fingers, often causing puckering and dimpling of overlying skin. With increase severity of contracture, normal function of the hand impaired and at this point therapy is sought **(Weiss and Goldblum 2001).**

Imaging:

MRI may be useful for determining the extent of disease and appropriate therapy (**Yacoe et al 1993).**

Treatment:

Surgical extirpation remains the treatment of choice in patient with sever flexion contracture that impair normal hand function **(Weiss and Goldblum 2001)**.

Fasciotomy (subcutaneous division of the fibrous bands) leads to good immediate improvement of contracture of metacarpophalangeal joint. It has no effect on the progression of disease but may cause injury to digital nerves and vessels. It is preferable in those who can tolerate only minor surgical procedure or who have only limit palmer involvement **(Rodrigo et al 1976).**

Subtota or total fasciectomy associated with lower local recurrence and better long term effect than fasciotomy(**Shaw et al 1996).**

Dermofasciectomy followed by skin graft associated with lowest local recurrence <10% , this done in patient with recurrence require re - operation and for patient who his primary disease involved the skin **(Hall et al 1997).**

Non surgical methods includes: radiotherapy, **Keilholz et al (1996)** found radiotherapy effected in prevent disease progression and helping in therapy to avoid unnecessary surgical procedures for advanced disease. The use of collagenase and calcium channel blockers in treatment of palmer fibromatosis have only been tested in vitro **(Starkweather et al 1996, Rayan et al 1996).**

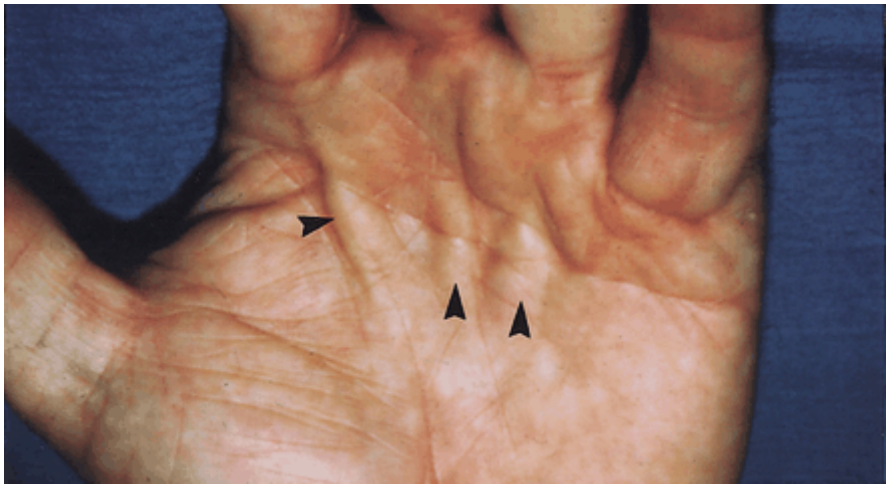

Figure (1) *Palmer fibromatosis*

(Mark et al 2001)