

Fibromatoses

Introduction

These are proliferative lesions composed of cells resembling fibroblasts, associated with the production of a variable amount of collagen and ground substance.

They have ill defined margins and tend to recur locally.

Some may undergo spontaneous regression; others continue to grow progressively.

Juvenile subcutaneous fibromatosis (Fibrous hamartoma of infancy)

This presents in the first decade with a subcutaneous nodule fixed to the skin and underlying fascia.

The lesions tend to be multiple.

They may occur anywhere on the body, but have a predilection for the upper limbs.

Most of the lesions resolve completely.

Aggressive fibromatosis (desmoid tumour)

This is an uncommon condition, which may occur anywhere. Associated with Gardner's syndrome.

More common in females, from 2nd to 4th decade.

Characterised by fibrous proliferation in deep tissues that shows continuous and relentless growth and a marked tendency to recurrence and destruction of local structures. It tends to grow along muscle planes.

The term is used when the histology is bland but the behaviour aggressive. If there is atypia the diagnosis is likely to be fibrosarcoma.

Excision with a wide margin is necessary, and recurrences may require amputation.

Plantar fibromatosis

This is usually a disease of young adults, but has been reported at any age from birth to 70 years.

One or more firm nodules are felt in the sole, attached to the plantar aponeurosis. Bilateral in 10%.

Some lesions regress spontaneously and can be watched.

If there is ongoing pain or progression the lesion can be excised, but with a wide margin, preferably including the skin with a graft. Methotrexate can be used as adjuvant treatment.

Plantar fibromatosis of the heel

This is a separate condition, seen in young adults, where there is a lesion on the medial aspect of the heel, which has been referred to as fatty heel lumps, but in fact contains fibrous tissue.

It usually resolves spontaneously.

Congenital generalised fibromatosis (Stout's disease)

Rare form of fibromatosis where there are multiple subcutaneous and visceral nodules present at birth, generally fatal.

Bone lesions may occur, typically lucent defects in the metaphysis.

Recurring digital fibroma of Reye

These lesions tend to occur in children under one, and present as firm subcutaneous nodules on the dorsum or sides of fingers or toes. Multiple lesions in over 50%.

Recurrence is noted in over 75% of excised lesions.