Recurrent Fibromatosis

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ABSTRACT

Fibromatoses are a group of rare fibrous tissue proliferations of benign nature. They are characterized by an infiltrative growth with a tendency towards recurrence; however, they never metastasize. However, they do display local aggressiveness, with complete resection being the treatment of choice.

Key Words: Fibromatosis, Desmoid, complete resection

INTRODUCTION

The term "Fibromatosis" covers a broad spectrum of benign fibrous tissue proliferations; their biological behaviour is almost similar to that of benign fibrous lesions and fibrosarcomas. The most important strategy is to prevent their direct invasion into the adjacent tissues. Fibromatoss are known to arise from the connective tissue of muscles and the fascia or from the aponeurosis. The aetiology of fibromatosis is largely uncertain.

However, the lesions developing from previous scars and a definitive history of injury to the muscle, fascia and the aponeurosis are usually clinching clinical evidences. An association with familial polyposis coli and Gardner's syndrome is encountered frequently; particularly, the sporadic form is seen also with osteomas. The sporadic tumours are more frequent in women than in men from 2:1 to 5:1.

Fibromatoses are benign tumours that affect the connective tissues that grow rapidly in any part of the body, but do not metastasize. They also develop from muscles, fasciae and aponeuroses. The neoplasm is composed of spindle (fibrocyte-like) cells. As regards the site, fibromatoses, particularly the extra-abdominal ones in the areas of the shoulder and the pelvic girdle are characterized by a high risk of recurrence (25-65%) after surgical treatment.

The deep fibromatoses display locally aggressive behaviour; The superficial fibromatoses typically remain small and are less likely to recur, despite having an identical morphology. Somatic beta –catenin or APC gene mutations have been reported in more than 74% of the deep fibromatoses and in virtually 100% of the Gardner's syndrome cases which are associated with fibromatoses. Clonal chromosomal aberrations –trisomies of chromosomes8 and 20 and loss of 5q material has been seen.

In our case, resection and primary closure was performed and the patient was symptom free after surgery.

CASE REPORT

A 32 year old female presented to us with a swelling in the left lower back, with a duration of 1 and a half years. The swelling had rapidly increased in size since 6 months. There were no other relevant symptoms. She gave a history of a similar swelling in the same region 5 years back, which was operated twice in the past 2 years.

On examination, a single swelling in the left dorso-lumbar region, 2 cms from the midline was seen. It was 10x6 cm in size and 6 longitudinally oval in shape. The skin over the swelling was stretched and shiny, with a linear scar 8 cm in length and a transverse scar 5 cm in length. The swelling was non tender and the consistency was hard. The plane of the swelling was muscle. The spine was however normal.

Routine investigations did not show any abnormality. The diagnosis was confirmed by a tru-cut biopsy, which revealed recurrent fibromatosis. C.T. scan revealed a well defined soft tissue density mass involving the left para-spinal muscles, extending from the D11 to L3 – L4 vertebrae.

The operative procedure was done under general anaesthesia, with the patient in the prone position. A wide excision of the tumour and primary closure was performed. The tumour which was shaved off the para-spinal muscles, was situated close to the pleura. The procedure was uneventful. The histo-pathological examination revealed recurrent fibromatosis. The patient was discharged on the 12th post operative day. She was followed up for a period of one year and was found to be asymptomatic, with no evidence of recurrence.

DISCUSSION

Fibromatosis can occur in a variety of anatomical locations, the extremities, girdles, chest wall, abdominal wall, head and neck and very rarely in the retro peritoneum. The overall incidence is 0.4 to 1%. These tumours display local aggressiveness, but with no propensity to metastasize. The deep seated fibromatoses are more aggressive as compared to the superficial fibromatoses.
Our case was operated upon previously, but probably with a diagnostic outlook. The findings of CT scan and the tru-cut biopsy suggested a recurrence and therefore, an excision was planned. A 3-D excision was performed and the primary closure was done successfully. The histological picture revealed the proliferation of the well differentiated fibroblasts. Since the tumour was situated in the lower back, the patient sought medical attention early. Radiation therapy is an effective treatment in cases where complete resection is not possible.

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