BONE GRAFT SUBSTITUTES WITH BONE CURETTAGE FOR THE TREATMENT OF FIBROUS DYSPLASIA OF RIGHT TIBIA

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ABSTRACT

Fibrous dysplasia (FD) is a benign condition, with an ability to affect any bone in the body and has a wide spectrum of severity. It is a chronic condition of the skeleton where a portion of a bone develops abnormally. We report and discuss a case of six year old male patient suffering from fibrous dysplasia of right tibia, its diagnosis, and management including surgical intervention. Bone graft substitute with bone curettage was performed after thorough excision of bone tumour and bone wash. Procedure was carried out under general anaesthesia and C-arm guidance. Significant improvement was seen clinically and patient walked eight weeks after surgery. He was able to carry out routine activities normally with a significant improvement in quality of life. This was the latest surgical procedure available for the treatment of FD.

Keywords: Bone curettage, bone graft substitute with bone curettage, Fibrous dysplasia, quality of life

INTRODUCTION

Fibrous dysplasia (FD) is a rare clinical condition in which bone tissue is replaced by fibro-osseous lesions. These skeletal lesions representing about 5% of all benign bone tumors may involve one bone (monostotic) or multiple bones (polyostotic). The lesions can occur throughout the skeleton but tend to occur in long bones, ribs and craniofacial bones; they represent 5-7% of all benign bone tumours. [1] Symptoms usually appear between 5 and 20 years of age with an earlier onset is seen in more extended cases of dysplasia. [2] Exact pathogenic mechanism of fibrous dysplasia is unknown but recent studies indicate that genetic factors may be responsible, and the disease is linked to a postzygotic mutation in guanine nucleotide stimulatory protein (GNAS1) located on chromosome 20q13.2-13.3. [3]

Birth marks (cafe-au-lait spots) often can be the first presenting sign of FD and may be associated with other endocrinological disturbance such as precocious puberty, hyperthyroidism, hypophosphatemia, excess growth hormone. When a patient presents with birth marks or combination of symptoms, it is known as McCune-Albright syndrome.

Symptomatic treatment is the option in FD as there is no cure. Though medications such as bisphosphonates have been shown to be highly efficacious in relieving pain, surgery is still the mainstay of treatment for FD affecting long bones.

Many studies have been conducted and data is available on FD in developed countries, but limited data is available from developing countries including India, where only anecdotal cases are reported.

CASE REPORT

A six year old boy presented to our hospital, a tertiary care centre, with pain and swelling over right leg, associated with difficulty in walking, since one year. There was anterior bowing in the middle of the right leg. On admission, routine haematological and biochemical investigations including serum levels of calcium, phosphate, alkaline phosphate and ESR, serial X-rays – anteroposterior (AP) and lateral (LAT) view of right leg (fig 1a,b), tech 99m bone scan(fig 2a,b) and magnetic resonance imaging (MRI) scanning of right tibia were performed. Haematological and biochemical results were within normal limits.

Fig1a: X-ray right tibia anteroposterior and lateral view at initial stage of the disease

Fig1b: X-ray of right tibia anteroposterrial and lateral view at later stage of the disease
Patient's age, clinical history, and results of investigations indicated a probable diagnosis of fibrous dysplasia of right tibia.

C-arm guidance and tourniquet control surgery was performed under general anaesthesia (GA) i.e bone graft substitute with bone curettage after thorough excision of bone tumour and bone wash followed by bone biopsy.

Procedure

Longitudinal incision was given on the antero-lateral aspect of middle third right leg under GA; affected bone was identified after separating the tissues under c-arm guidance. Major lesions were located and identified, bone was drilled and three holes were made on the middle third of right tibia. Bone graft substitute with bone curettage was done after thorough wash. Wound was closed after confirming by C-arm. (fig3a,b,c,d,e,f,g)
development during the first decade of life, followed by spontaneous resolution. [10] Data on long term effects of the disease is not available in the literature and hence it is important to have long term follow up to observe and manage complications.

FD is seen in childhood and early adolescence, and our patient's age was within this age group. Usually it is asymptomatic, posing difficulty in diagnosis, but majority of cases present with bone pain, deformity and fractures, and our patient presented with bone pain, swelling and deformity. Though endocrinopathies are also associated with FD, our patient did not have any such feature. FD is usually diagnosed on the basis of characteristic radiological findings on X-ray, bone scans, and in this patient, a probable diagnosis of FD was made based on radiological findings.

Surgical intervention, which aims at symptomatic relief, is indicated in patients with disease that cannot be managed by conservative treatment, and who have an impending fracture or progressive deformity. We planned surgical treatment for this patient to achieve symptomatic relief and to improve quality of life. The surgery performed was the latest available surgical treatment in 2012.

Currently various surgical modalities are available for treatment of FD i.e minimal invasive plate fixation or intramedullary nailing, bone graft substitute with bone curettage; autograft is not advisable as it will be resorbed. Postoperatively bisphosphonates can be administered to the patient. Early surgical intervention prevents pathological fractures in these patients but requires customized plating; interference with wound healing is another issue which needs to be addressed. Though surgical treatment is available, but availability of experienced orthopedic surgeon that too in few selective centres, cost involved are major challenges faced by developing countries like India.

CONCLUSION

This technique is a useful tool in restoring the functions without compromising quality of life. Moreover, there was no surgery related complications. However, availability of an orthopedic surgeon with hands on experience in this technique is a challenge to be addressed.

Competing interest

The author(s) declare that they have no competing interests.

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