Nonossifying Fibroma: A Literature Review and Case Study

Jason Grove, BS and Carly Robbins, BA

Abstract

Nonossifying fibromas or fibrous cortical defects are developmental defects usually found in the metaphysis of long bones in the lower extremity. Radiographically, they are well delineated multi or uniloculated lesions with sclerotic margins. Based on radiographs alone, one can usually make an accurate diagnosis of nonossifying fibroma. Commonly found in children and adolescents, nonossifying fibromas are the most common fibrous bone lesions.[1] Therefore, it is important to recognize the radiographic and clinical characteristics of these benign lesions. A case of nonossifying fibroma is presented here with a review of the literature.

Introduction

Fibrous cortical defects and nonossifying fibromas are benign lesions occurring most often in the distal femur, proximal and distal tibia, and fibula.[2] Histologically, the lesions are identical but classified separately based on the extent of osseous involvement. Fibrous cortical defects are small, intracortical lesions that are asymptomatic, whereas nonossifying fibromas are larger, actively growing lesions that involve the medullary cavity.[3,4] Collectively, the two lesions are referred to as fibroxanthomas or histiocytic xanthogranulomas.[1] We will refer to both lesions as nonossifying fibroma (NOF).

Literature Review

The incidence of nonossifying fibromas is 30-40% of all children over the age of 2 with the highest incidence between ages 4-8.[3] Most studies have shown a male to female ratio of 2:1. The etiology of NOF is still unknown. The current theory is that these lesions occur as a result of developmental aberrations at the epiphyseal plate. Hatcher described NOF as a lesion that does not undergo osseous metaplasia, suggesting that these lesions are not neoplasms, but developmental defects.[5,6] It is also thought that since these lesions tend to occur after the age of

2, a muscle pull and periosteal injury may be a contributing factor.[7] Histologically, the lesions contain whorled bundles of connective tissue cells admixed with foamy histocytes, hemosiderin, hemorrhage, collagen, multinucleated giant cells, and bone trabeculae.[2,8,9]

Nonossifying fibromas have been found to be associated with several diseases, although they can occur in otherwise healthy individuals. A correlation between multiple nonossifying fibromas and neurofibromatosis has been identified. Although these disorders are not mutually exclusive, it has been speculated that multiple NOF are another variation of mesodermal abnormalities.[10] Nonossifying fibromas have also been found to be associated with Osgood-Schlatter disease, osteochondritis of patella, osteochondritis dissecans of medial femoral condyle, and Legg-Calve-Perthes disease.[3,5]

Clinically, the majority of all NOF are asymptomatic and are discovered incidentally on radiographs. Symptomatic lesions may present with mild pain and swelling of short duration. On physical exam, they may have bone tenderness with palpation. However, the physical exam is usually unremarkable. Patients who recall a history of trauma, have often suffered trauma of moderate severity such as a sprain, kick, or fall.[8] Pathologic fractures through NOF are rare, but can occur if the lesion is large enough. If symptoms are present, pathologic fracture should be ruled out. These fractures have been shown to successfully heal in a normal length of time after cast immobilization.[7] However, in most cases the lesion persists after the fracture has healed.[7]

On plain film radiographs, NOFs appear as eccentric, multi or uniloculated, ovoid lesions in the metaphysis of bone with sclerotic margins.[8] The lesions may also extend into the medullary cavity. The long axis of the NOF is most commonly seen parallel to the long axis of the bone and are usually located medially.[3] Jaffe stated that radiographs are sufficient to make a diagnosis of NOF.[8] Others went as far as to say that a radiographic diagnosis of nonossifying fibromas has been shown to approach an accuracy of 100%.[10,11,12,13,14]

The defining characteristics of an NOF can also be found with ancillary imaging. Magnetic resonance imaging can be very helpful in the proper diagnosis of NOF. On a T1

weighted image, the lesion will show a decreased signal intensity. NOF on T2 weighted images may appear hypointense or hyperintense. Hypointensity is thought to be related to extensive hypercellular fibrous tissue. Hyperintensity may be due to a lack of fibrous tissue and abundant foamy histiocytes. Sclerotic margins are seen on MRI as a peripheral hypointensive rim. Infusion of gadopentetate dimeglumine may be helpful for examination of the lesion because it enhances contrast.[9]

Treatment varies depending on the size and severity of the nonossifying fibroma. Surgery is often not required to treat NOF due to a high rate of spontaneous regression and a lack of symptoms.[2,3,4] For non-symptomatic lesions, the only treatment indicated is yearly or bi-yearly radiographs until the lesion has ceased growth and appears stable.[15] Special attention should be given to lesions measuring larger than 33mm and involving more than 50% of the bone's diameter.[7] This is especially true for lesions found in young children due to the risk of further growth.[15] The need for prophylactic curettage of large, growing lesions that are not painful is in debate. However, most authors state that surgery should not be done in asymptomatic patients. Symptomatic lesions should first be treated conservatively. Conservative care consists of limited activity and immobilization, in addition to yearly or bi-yearly radiographs. If symptoms persist with conservative treatment, then wide surgical curettage with autogenous bone graft is indicated.[4,16,17,18] Simple curettage is not recommended because it may result in an increased recurrence rate.[4,16,17,18] Tiedeman suggests using a mixture of autogenous bone graft with demineralized bone matrix powder because it may be difficult to obtain a sufficient amount of autogenous bone in children.[4] If a pathologic fracture is identified, it should first be treated with reduction and cast immobilization.[2,7] If the lesion persists after fracture healing, then the treatments previously discussed should be administered. The recurrence rate of an NOF is low after adequate curettage and subsequent bone grafting has been done.[15]

Case Report

The patient was an 11 year old African American female who presented with pain in her feet and ankles. She admitted to suffering an inversion ankle sprain to her left ankle within the past year.

Her mother related that she had gained 100 lbs and had a major growth spurt within the past year. The physical exam revealed pain on palpation of the left lateral malleolus, pain with resisted eversion, and mildly decreased muscle strength of the everters of the left foot. There was a decrease in dorsiflexion of both ankles, with all other joints having normal range of motion. The patient had a pronated foot type on and off weight bearing.

Anterio-posterior, lateral, and medial oblique radiographs were taken bilateral to assess the patient's flatfoot. Upon examination of the radiographs, the distal extent of what appeared to be a lytic lesion was discovered on the left distal tibial metaphysis. Subsequent lateral ankle and ankle mortise views were taken of the left ankle to further evaluate the lesion (figures 1, 2). Both ankle radiographs revealed a 2.6cm x 4.9cm intramedullary lesion. Tibial cortices were intact, and there was some change in the shape of the posterior cortex. The lesion was multi-loculated with sclerotic, scalloped margins and appeared to be non-aggressive and benign in nature. The patient was diagnosed with bilateral pes planus, gastroc-equinus, and a left tibial nonossifying fibroma. The patient's bilateral symptoms that involved muscle pain were attributed to severe pes planus and not the NOF.

Figure 1. Ankle mortise view of the left ankle of an 11-year-old female showing an eccentrically located intramedullary lesion of the distal tibial metaphysis measuring 2.6cm X 4.9cm. Lesion has a well defined sclerotic border surrounding a multi-loculated center. Note also that the long axis of the lesion is parallel with the long axis of the tibia.

Figure 2. Lateral ankle view of left ankle and foot of same patient. In this view, the lesion also measures approximately 2.6cm X 4.9cm.

The patient was sent for magnetic resonance imaging of the area. An eccentric cortical sclerotic lesion was identified in the distal metaphysis of the tibia. The lesion was predominately hypointense on T1 and mixed T2 weighted images. A focus of increased signal intensity was identified within the lesion on T1 weighted imaging, and appeared with a heterogenous signal on T2 weighted images. The findings were consistent with a non-ossifying fibroma containing a small focus of hemorrhage. There was no evidence of a cortical break, surrounding marrow edema, or soft tissue mass.

Treatment of the patient will consist of close observation. It was recommended that plain film radiographs be repeated every 6 months due to the large size of the lesion and the increased weight of the patient. No activity restrictions were placed on the patient because her activity level was already minimal. There have been no complications thus far. The patient was also dispensed custom functional orthotics for bilateral pes planus.

Discussion

Nonossifying fibromas are a common occurrence in children and young adolescents. Moreover, 90% of all NOF occur in the lower extremity.[3] Yet, they are rarely found, due to a lack of symptoms and a high rate of regression. Nonetheless, it is important for physicians specializing

in the lower extremity to readily recognize NOF radiographically and understand the proper treatment course. Large osseous lesions can be very alarming to patients and physicians alike, so the ability to make the diagnosis is helpful in avoiding any unnecessary alarm. Because NOF, and in particular multiple NOF, can be associated with certain diseases, the ability to recognize NOF may aid in the diagnosis of other disorders. Making the diagnosis of NOF solely based on radiographs is possible due to the distinct characteristics, as was seen in the case presented here. The only possible differential diagnosis based on radiograph is chondromyxoid fibroma.[13] However, making a diagnosis on radiographs alone when considering a possible osseous tumor in children carries a slight risk. Thus if the diagnosis is not clear with ancillary imaging, then wide surgical curettage with autogenous bone grafting should be done.[2,13] The curettage acts as a biopsy and the definitive diagnosis can be made.

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