

Case reports

Supracondylar femur fracture complicating epileptic insult : a specific and under diagnosed complication ?

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Abstract

Purpose : In epileptic patients fractures are six times more frequent than in the general population. Known predisposing factors are anticonvulsant drugs, malnutrition, lack of physical activity and sunlight exposure.

Methods : In this study we describe two patients, one with a bilateral supracondylar fracture and one with a unilateral supracondylar fracture after an epileptic seizure. The literature concerning femur fracture following an epileptic insult is reviewed.

Results : A review of the literature revealed several cases of femur fractures associated with epilepsy, however no cases were found involving a supracondylar femur.

Conclusions : Our hypothesis is that a tonic seizure with simultaneous contraction of both agonists and antagonists can cause this type of fracture in predisposing patients.

Key words : Seizure ; epilepsy ; spontaneous fracture ; supracondylar femur fracture ; intellectual disability.

Introduction

Children with cerebral palsy, who are not ambulatory, and children with epilepsy have a higher risk of pathological fractures than the general population (Mattson R. H., Gidal B. E., 2004 ; Souverein P. *et al.*, 2005). For patients with epilepsy, the most important predisposing factors include lack of mobility, poor diet and anticonvulsant medication which can cause osteoporosis (Mattson R. H., Gidal B. E., 2004 ; Souverein P. *et al.*, 2005 ; Gross R. A. *et al.*, 2004 ; Jekovec-Vrhovsek M. *et al.*, 2000 ; Sheth R. D. *et al.*, 1995 ; Vestergaard P. *et al.*, 2004 ; Pack A. M. *et al.*, 2004, Pack A. M. *et al.*, 2005 ; Swanton *et al.*, 2007). A large proportion of children with refractory epilepsy have intellectual disabilities and thereby difficulties in communication, as a result the diagnosis of a fracture is often delayed and the risks for complications, such as progression of a nondisplaced fracture to a displaced fracture are increased. The first signs of a fracture include changes in mood and behaviour,

accompanied by localized swelling and abnormal positioning of the leg.

While several types of orthopaedic problems have already been associated with epilepsy, including vertebral compression fractures, shoulder dislocations, proximal humeral fractures, scapular fractures, hip dislocations, femoral neck fractures, acetabular fractures, radius-ulna fractures, ankle fractures and fibula head fractures (Brunner R., Doderlein L., 1996 ; Desai K. *et al.*, 1996), supracondylar fractures of the femur have not yet been reported as a complication of an epileptic insult.

In Pulderbos, an epilepsy centre for approximately 65 children and adolescents, we were in the last 5 years confronted with two patients with supracondylar femur fractures as a complication of epileptic seizures.

Methods and material

The first child is a seven year old boy with a severe cytomegalovirus embryopathy and refractory epilepsy, complicated with a bilateral supracondylar femur fracture.

Clinically the child presented with a severe impairment in motor development (quadriplegic cerebral palsy), visual impairment (bilateral chorioretinitis), microcephaly and a short stature, the patient could not sit or stand independently and equipment such as sitting and standing position orthoses were necessary.

At the time of presentation the child experienced daily, short partial epileptic seizures, occasional severe secondary generalised tonic-clonic seizures and rare status epilepticus.

Previous treatment included several different antiepileptic combinations, when the fracture occurred the child was on a combination of sodium valproate, lamotrigine and vigabatrin.

During an intercurrent infection the child suffered from a series of severe seizures, with tonic contractions of the legs and trunk which was



FIG. 1. — Bilateral supracondylar impaction fracture of the femurs.

stopped with clonazepam treatment intrabuccal. Following this event, despite adequate antibiotic treatment, the patient showed a change in behaviour and irritability, after three days of immobilisation, the leg was still slightly painful. Blood examination revealed a slightly elevated creatinine kinase and phosphates, calcium was normal. X-ray and ultrasound of the hip were normal. The diagnosis of muscular microtrauma was assumed, due to the intensive mobilisation of the knees or due to the tonic contractions during the seizures. Eight days later the boy showed tender swelling on both sides of the leg above the knees. X-rays were performed (Fig. 1) and showed besides the very osteoporotic bone, bilateral supracondylar fractures with impaction of the femur which were at least one week old. Following four weeks of bed rest and analgesic medication the fractures were healed.

The second patient is a boy of sixteen years of age who developed Lennox-Gastaut syndrome at the age of two years and five months. His epilepsy is refractory despite different medications and vagal nerve stimulation. After his last status epilepticus he was referred to our epilepsy centre where we tried to adjust the anticonvulsant medication. At the time of admission the boy was on a combination of sodium-valproate, lamotrigine, phenobarbital and clobazam although he still had daily tonic seizures mainly during sleep.



FIG. 2. — Old impaction fracture of the tibia and recent supracondylar femur fracture.

During his stay in our centre he developed severe pain in his right knee which became swollen and felt warm, blood examination showed some signs of inflammation. An ultrasound showed an inflammation of the tendon of the patella, doppler investigation was negative, X-ray of the lower limbs showed an old impaction fracture of the right proximal tibia, and a generalised osteoporotic bone. Treatment with ibuprofen and clarithromycine was started. Despite treating the tendonitis the child's condition did not improve and he continued to be irritable. Repeat of the X-ray (Fig. 2) confirmed the old fracture with impaction of the tibia and brought to light a new fracture: a supracondylar femur fracture which was confirmed with a hot spot on bone densitometry. The leg was placed in a cast for four weeks, which was long enough to heal the fracture. In both patients there were no arguments for an underlying disease such as osteogenesis imperfecta or for other traumatic causes such as battering. Additionally, although we did not control for it, there were no arguments for disturbance of parathormone or vitamin D metabolism.

Discussion

In patients with epilepsy fractures are two to six times more likely than in the general population (Mattson R. H., Gidal B. E., 2004 ; Souverein P. C.

et al., 2005 ; Swanton *et al.*, 2007). Finelli and Cardi described the relationship between seizures and fractures, pathological fractures without direct trauma occur in 0.3% of cases of convulsive seizures (Finelli P. F., Cardi J. K., 1989).

PREDISPOSING FACTORS

Various factors of an epileptic insult can cause fractures.

Obviously, atonic or tonic-clonic insults, in which the patient often falls, can cause a fracture. Furthermore, atraumatic fractures can occur as the bone mineral density of patients under anticonvulsant medication can be strongly decreased (Mattson R. H., Gidal B. E. 2004 ; Souverein P. C. *et al.*, 2005 ; Sheth R. D. *et al.*, 1995 ; Vestergaard P. *et al.*, 2004 ; Pack A. M. *et al.*, 2004 ; Pack A. M. *et al.*, 2005). Sheth R. D. *et al.*, 1995 showed that bone density is decreased during valproate treatment, but not with carbamazepine treatment. Vestergaard P. *et al.*, 2004 described that there is a difference between the various antiepileptic drugs, specifically carbamazepine, clonazepam, phenobarbital and valproate were significantly associated with increased risk of fractures, while ethosuximide, lamotrigine, phenytoin, primidone and vigabatrin were not significantly associated with fracture risk. This was confirmed by the study of Gross R. A. *et al.*, 2004 which showed that older women (average age 70 years) treated with carbamazepine have nearly 50% greater rate of bone loss. Sheth R. D. *et al.*, 1995 also described that the reduction in bone mineral density increased with the duration of the valproate therapy.

For many years it was hypothesised that only liver enzyme inducing drugs could elevate the fracture risk (Anderson G. D. 2004 ; Pack A. M. *et al.*, 2005). These drugs influence the metabolism of vitamin D so there is more conversion of active vitamin D into inactive metabolites. Vitamin D deficiency is associated with low serum calcium concentrations and secondary hyperparathyroidism, and thus increased markers of bone resorption and turnover (Anderson G. D., 2004). Valproate is a liver enzyme inhibitor, therefore the decreased bone density cannot be attributed solely to the enzyme inducing effects of the AED's. Other possible mechanisms for AED-associated bone disease include impaired absorption of calcium due to the AED (Anderson G. D., 2004 ; Pack A. M., 2003), direct increases of bone resorption, inhibition of response to PTH (parathyroid hormone), vitamin K deficiency, hyperparathyroidism and calcitonin deficiency (Pack A. M. *et al.*, 2005 ; Pack A. M., 2003).

Swanton *et al.*, 2007 described a significant reduction of bone mineral density in institutionalised adult patients (males more than females) with refractory epilepsy. However, in contrast with other

Table 1

Different mechanism how seizures can lead to a fracture

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| <ol style="list-style-type: none"> 1. Traumatic fracture : falling patient due to seizure 2. Atraumatic fracture : <ol style="list-style-type: none"> a. Reduced bone mineral density <ul style="list-style-type: none"> - Lack of sunlight exposure - Poor diet - Lack of mobilisation - Antiepileptic drugs b. Violent muscular contraction |
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studies, they did not find a correlation with any specific AED.

During a seizure there are extremely forceful contractions of muscles, frequently of agonists and antagonists together as in a tonic seizure, which create a greater risk for fractures and dislocations (Finelli P. F., Cardi J. K., 1989). A fracture in an epileptic patient can therefore be due to the forceful contraction of muscles acting upon bone, which is already weakened by osteoporosis as a result of a combination of nonweightbearing anticonvulsant drugs, lack of exercise, sunlight exposure and poor diet (Gross R. A. *et al.*, 2004 ; Jekovec-Vrhovsek M. *et al.*, 2000) (Table 1).

LOCATION

When the incidence of density ratios was compared between a cohort of patients with epilepsy and a reference cohort the highest relative risk estimate was found for hip and femoral neck fractures (Souveirin P. C. *et al.*, 2005). Our two case reports presented with supracondylar femur fractures, which have not yet been described in literature. It seems logical that this type of fracture can only be caused by a simultaneous severe contraction of flexor and extension muscles as is the case in a tonic seizure.

DELAY IN DIAGNOSIS

In both cases the diagnosis of the fracture was delayed, many factors can explain the delay. First, patients in this population are often severely mentally retarded and do not have normal communication skills. The first sign of a fracture is therefore often rather atypical, like change in behaviour and irritability.

Second, because this is a fracture with impaction there is no displacement of the leg and pain due to mobilisation is rather limited. Most of these patients do not stand alone, so change in functionality will not be recognised. Hence, radiological examination is often required to diagnose these fractures.

PREVENTION

Preventive measures are important. Optimal control of seizures is obvious, but also additional administration of calcium and vitamin D plays an important role (Mattson R. H., Gidal B. E., 2004). Childhood and adolescence are critical periods for skeletal mineralization during which vitamin supplements become even more important. In a recent study Bischof F. *et al.*, 2002 concluded that there was a marked clinical improvement after three months of vitamin D intake in a group of children with epilepsy and decreased bone density. In addition, the mean calcium and phosphate levels increased. Beside vitamin supplements lifestyle interventions like adequate diet, sunlight exposure and exercise are also important.

Conclusion

There are three important factors that can explain the higher frequency of fractures in epileptic patients. First is trauma from a fall caused by an epileptic insult. Second, a more osteoporotic bone due to anticonvulsant therapy, lack of mobilisation, sunlight exposure and poor diet, and third, the strong muscular contractions which can accompany an epileptic insult.

In this report we present two cases of a rather special fracture caused by a tonic epileptic insult in severely retarded children. Rare supracondylar femur fractures, unilateral as well as bilateral, can be caused by violent muscular forces generated during a severe tonic seizure in predisposed epileptic patients.

Since clinical signs may be subtle because of lack of pain symptoms, especially in these severe mentally retarded children, one must always consider a fracture with impaction in a child who stays irritable after a severe convulsion.

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