CASE REPORT

Scaphoid non-union in osteogenesis imperfecta

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SUMMARY

We report a case of scaphoid non-union in a child with osteogenesis imperfecta (OI) presenting 7 months after a fall. Following diagnosis, conservative treatment was initiated, but despite 4 months in a plaster cast, the fracture had failed to unite. Open reduction and internal fixation was performed (Acutrak screw) with bone graft harvested from the distal radius. Postoperative immobilisation continued for 10 weeks, and at 4 months the child was pain free and had resumed normal activities. The fracture had fully united radiologically at 9 months. Non-union is reported to occur in 23% of paediatric scaphoid fractures when treatment is delayed, and conservative and surgical treatment have both been described. Since OI increases the risk of non-union in long bone fractures, the scaphoid may also be at risk. We recommend a high level of suspicion for non-union in this patient group and a low threshold for consideration of surgical treatment.

BACKGROUND

Osteogenesis imperfecta (OI) is an inherited disorder characterised by a defect in type I collagen leading to fragile bones that are prone to fracture with minimal force. The Sillence classification divides patients into four types of differing clinical features and severity.¹ Management focuses on reducing the risk of fractures, addressing deformities, treating fractures and preventative treatment using bisphosphonates.²

Non-union may occur and usually affects long bones.³ The risk is higher following an elective osteotomy, but not following a fracture for patients managed with bisphosphonate infusions.⁴ To the best of our knowledge, there are no published reports of scaphoid non-union in OI.

CASE PRESENTATION

A 9-year-old boy first presented to secondary care in 2001, aged 2 years, following a femoral fracture, which was managed conservatively. There were no reported perinatal complications or previous hospital admissions. His father was known to have a mild form of OI, treated with calcium and vitamin D.

Over the subsequent 2 years, the boy sustained multiple minor fractures prompting referral to a geneticist who made a formal diagnosis of mild-moderate OI. Tri-annual bisphosphonate infusions (pamidronate) were started 4 years later following two further fractures. Initially, 20 mg was delivered on each of three consecutive days, but this was increased such that, in August and December 2008, he received doses of 20, 30 and 30 mg. A baseline dual-energy X-ray absorptiometry (DEXA) scan revealed a Z score (anteroposterior (AP) spine

L1-L4) of -2.2 and bone mineral density (BMD) was 0.403 g/cm^2 . A comparative follow-up DEXA scan was scheduled after 3 years to assess treatment response. Despite prophylactic intravenous pamidronate, the boy suffered three further minor fractures, all of which united with conservative treatment.

In 2009, he presented to accident and emergency department, aged 9 years, with persistent left wrist pain and swelling, having fallen onto his outstretched hand 7 months previously (December 2008).

INVESTIGATIONS

Plain radiographs demonstrated a chronic scaphoid fracture, and a plaster cast was applied in the emergency department. Following assessment in fracture clinic, an urgent MRI scan was requested to evaluate the viability of the fracture fragments. Proximal and distal fragments were, indeed, both viable, therefore non-operative treatment with a plaster cast was continued. His progress was monitored with repeated plain radiographs, but after 4 months of treatment, radiographic appearances were consistent with non-union.

TREATMENT

Surgical treatment was recommended and, in December 2009, the boy underwent open reduction and internal fixation of his left scaphoid with bone graft harvested from the distal radius. Fixation was achieved with an Acutrak screw. There were no perioperative complications and plain radiographs confirmed good positioning of the metalwork. Immobilisation in a below-elbow cast was continued for 10 weeks post-operatively. The time from the last preoperative bisphosphonate to surgical fixation was 1 year.

OUTCOME AND FOLLOW-UP

Four months following surgery, the boy was pain free, had resumed normal activities and had no clinical tenderness; plain radiographs demonstrated fracture healing. The fracture had radiologically united at 9 months postsurgery, and he continued to be asymptomatic. Bisphosphonate treatment, which had been withheld following surgery, was restarted in October 2010, and a DEXA scan performed in May 2010 revealed an improvement in Z score (AP spine L1-L4) to -0.7 and BMD 0.573 g/cm^2 .

DISCUSSION

Traditionally, non-union has been considered uncommon in OI, but it appears to be more prevalent than originally thought, with two series

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identifying almost a fifth of their paediatric cohort to be affected.^{3 5} Most of the fractures involved long bones, and non-union often occurred after inadequate or absent immobilisation. Surgical intervention for these non-unions has been met with variable success. Gamble *et al*³ reported that all eight non-unions in their study healed following intramedullary nailing. Another group had disappointing results, with all three humeral non-unions failing to unite.⁵ An alternative and more recently described surgical technique involves the use of sandwich allograft with or without supplementary intramedullary nailing.⁶ A section of long bone fresh frozen allograft is cut longitudinally, and reassembled around the debrided and opposed long bone non-union site. Fixation of the construct is achieved with cerclage wires or screws, and union rates of 92% have been reported. None of the fractures in these studies involved the scaphoid.

Scaphoid fractures are uncommon in children and most frequently involve the waist.⁷ Initial plain radiographs are normal in 30% of cases and useful clinical predictors of fracture are volar scaphoid tenderness, pain with radial deviation and pain

with active wrist range of movement.⁸ While conservative management in a cast is successful in 90% of acute fractures, union rates are much lower (23%) for those presenting late.⁸ Gholson *et al*⁷ identified three independent risk factors of non-union: fracture displacement, chronicity and fracture site. Scaphoid non-union can be effectively treated with surgical fixation, achieving union rates of 97–100%.^{7 9} Prolonged immobilisation has also been described with Weber *et al*,¹⁰ advocating a 3-month trial of conservative treatment in the case of non-unions lacking previous immobilisation due to delayed presentation. All six patients in their series had union at a mean of 5.3 months following cast immobilisation.

Competing interests None declared.

Patient consent Obtained.

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Learning points

- Maintain a high index of suspicion for a scaphoid fracture when a child with consistent clinical features presents despite a normal plain radiograph.
- Treat undisplaced paediatric scaphoid fractures in a plaster cast, as most unite.
- When presentation is delayed, surgical intervention may be considered due to the risk of non-union. Conservative treatment is, nevertheless, an alternative, although patients should be aware that immobilisation may be lengthy.
- For paediatric patients with OI sustaining a scaphoid fracture, it is not clear whether the risk of non-union is higher than in the general paediatric population.

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