Single-Stage Surgery Using Calcium Sulfate Pellets in Association with Tumor Resection as Treatment for Intraosseous Hemangioma of the Radius Shaft in a 2-Year-Old Boy

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Abstract

Keywords

- intraosseous hemangioma
- pathologic fracture
- calcium sulfate pellets
- bone substitute

Intraosseous hemangiomas are uncommon benign vascular tumors. Here the authors report a case of intraosseous hemangioma of the radial shaft in a 2-year-old boy, surgically treated by tumor mass resection and application of calcium sulfate pellets with no signs of relapse over a 2-year follow-up. Moreover, the lesion healed with new bone formation, and the last plain radiographs suggested homogenous normal trabecular bone density. The radius grew in length and remodeled to an almost normal shape. Clinically, elbow flexion-extension and forearm pronation and supination were within normal limits at the last follow-up visit.

This report points to calcium sulfate pellets as a valid alternative to autologous bone grafting as void filler in large bone defects. Calcium sulfate pellets enable fast osteoinduction without interfering in follow-up imaging.

Introduction

Intraosseous hemangiomas (IHs) are uncommon benign vascular tumors, accounting for less than 1% of bone tumors.¹ The tumor is most frequently seen in normal bone, usually in the vertebral body and skull (75% of cases) and the scapula, ribs, clavicle, and pelvic bones (in a further 15–20% of all cases).² IHs are extremely rare in the long bones, with a predilection for the lower extremity.^{3,4} Only 3% occur in the long bones of the extremities, with tibia (43%) and femur (26%) the most common sites.⁴

Moreover, IHs are extremely rare in skeletally immature patients younger than 10 years as they are typically seen in adult patients between the third and the sixth decades of life.^{4–6}

As IH can prove difficult to treat, many different methods have been reported, including injection of ethanol, irradiation,

transarterial embolization, and en bloc resection or curettage with or without bone grafting.^{3,7}

Calcium sulfate pellets (CSPs) are a bioabsorbable bone substitute suitable for bone void filler applications in adult and pediatric populations.^{8,9} The osteoinductive and osteoconductive bone graft substitute comes prepackaged in small pellets that can be readily used in a one-stage procedure providing complete bone cavity filling.

The primary aim of this report is to present a case of a 2-year-old boy with solitary IH of the radius shaft with history of recurrent pathologic fractures treated by tumor resection and CSP grafting. The secondary aim is to review the literature on IH located in the appendicular skeleton of skeletally immature patients to assess the treatment outcomes of the different management options reported in the literature.

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Case Report

A 2-year-old boy was referred to the authors' hospital center following a pathologic fracture of the right radius shaft. Plain radiographs revealed the presence of an osteolytic lesion with cystic appearance at the level of the middle third of the radius shaft (► **Fig. 1**). Given the patient's age, clinical presentation, and radiographic characteristics of the lesion, solitary bone cyst was the most suitable presumed diagnosis. The patient underwent long-arm cast immobilization with the elbow in neutral position for 4 weeks followed by a volar splint for a further 3 weeks. One month after splint removal, the patient sustained a new pathologic fracture at the same anatomical location. He was again immobilized in a long-arm cast for 6 weeks followed by a volar splint for further 4 weeks. Despite consolidation of the fracture, the osteolytic area with cystic appearance was still visible on plain radiographs 5 months after completion of treatment. To avoid a new refracture, the lesion was surgically curetted and the bone defect filled with prepackaged CSP (OsteoSet, Wright Medical Technology Inc. Arlington, Tennessee, United States). Before grafting the bone defect with CSP, specimens were collected; then curettage, reaming of the bone internal wall until visible bleeding, and opening of the medullary canal were sequentially performed (► **Fig. 2**). After surgery, the forearm was immobilized in a volar splint for 4 weeks, after which passive and active upper limb mobilization was allowed.

Histologic examination revealed the presence of proliferated capillary vessel and sinuses with cavernous blood spaces without malignant features suggesting a diagnosis of capillary-cavernous IH. CD31- and CD34-positive immunohistochemical stains confirmed the vascular nature of the lesion. There was also positivity for Ki-67 and smooth muscle actin.

Fig. 1 Hemangioma at the mid-shaft of the radius in a 2-year-old boy. Periosteal apposition following healing of the pathological fracture.

Fig. 2 Postoperative X-ray showing the lesion treated with calcium sulfate pellets.

Following surgery, the patient underwent regular clinical and radiographic follow-up visits. One month after index procedure, plain radiographs showed partially reabsorbed CSP. Five months postoperatively, the filled cavity appeared dense on plain radiographs with complete reabsorption of CSP (► **Fig. 3**). Two years postoperatively, the area appeared radiologically homogeneous, resembling normal trabecular bone (► **Fig. 4**).

Fig. 3 Postoperative X-ray showing the lesion (A) at 1 and (B) 5 months.

Fig. 4 Postoperative X-ray showing the lesion at 2 years.

The child had resumed full activities with no pain or swelling in his right upper extremity. Elbow and wrist range of motions of the involved arm were normal. Until now, no evidence of recurrence or growth disturbances has been identified.

Literature Review

A search of the Medline database from 1938 to 2016 was performed to identify all cases of pediatric solitary IH of the appendicular skeleton previously reported in the English medical literature. A variety of search terms (“hemangioma,” “skeletal,” “intraosseous,” “appendicular”) were used, including combinations of index and free-text terms, as recommended in the *Cochrane Handbook for Systematic Reviews of Interventions*.

Abstracts were screened and relevant full-text articles were retrieved for further review. The reference sections of the papers retrieved were also screened to identify further literature. All levels of evidence were included. ▶ **Table 1** compiles case reports of pediatric solitary IH of the appendicular skeleton.

Discussion

IHs are uncommon benign vascular tumors characterized by the presence of differently structured (capillary/cavernous) blood vessels. They tend to occur superficially in the skin or deeper at intramuscular level but very rarely within the osseous tissue. IH accounts for less than 1% of all bone tumors, and the appendicular skeleton is the least common bony location.^{1,4}

To date, only 14 cases—including the present one—of IH of the appendicular skeleton have been reported in skeletally immature patients^{3,10–19} (▶ **Table 1**). Previously reported cases showed heterogeneous location. The most common involved bones of the appendicular skeleton are femur (three cases, 23%)

and tibia (three cases, 23%), followed by ulna (two cases, 15%) and radius (two cases, 15%), humerus (one case, 8%), fibula (one case, 8%), and ilium (one case, 8%). Within the affected bone, IHs are more frequently located at the metaphysis, followed by the metadiaphysis and the diaphysis.^{3,10–19}

In contrast to vertebral and cranial hemangiomas that are usually discovered incidentally on imaging examination, IHs of the appendicular skeleton in children and adolescents are symptomatic in more than 90% of cases.^{3,4,10–19} Symptoms include pain, swelling, restriction of function, and pathologic fractures (▶ **Table 1**).

Given its variable appearance on plain radiographs, histologic examination is vital to obtain a certain diagnosis. Malignant and benign processes (including chondrosarcoma, Ewing’s sarcoma, myeloma, fibrous dysplasia, osteochondroma, eosinophilic granuloma and solitary or aneurysmal bone cysts, and/or metastases) must be included in the differential diagnosis.²⁰

Treatment depends on the patient’s symptoms, age, and IH features. Asymptomatic lesions in non-weightbearing areas do not necessarily require treatment and can only be followed up. With growth, the vascular tissue may be replaced by fibrous tissue and achieve self-healing, although growth disturbances have been reported in skeletally immature patients.^{3,5,6,10–19}

However, symptomatic and/or large lesions in weightbearing areas warrant surgical treatment to the increased risk for pathologic fracture. Several surgical options have been reported for treatment of IH in skeletally immature patients, such as trepanation, curettage, or en bloc excision.^{3,10–19} However, the IH can recur in a relatively high number of patients, especially if resection is incomplete.^{10,15,19,20}

After wide bone resection, bone grafting is required, traditionally by autologous bone, to fill the bone defect and facilitate new bone formation. More recently, several clinical studies have been published on the use of bone graft substitutes to fill large bone defects and have reported satisfactory outcome.²¹ In particular, the use of bone substitutes allows to avoid donor-site morbidity and reduce operative time. Furthermore, these materials are osteoconductive and gradually resorb, allowing progressive bone formation.²²

Matsumoto et al reported a case of symptomatic IH affecting the ulna in an 18-year-old girl, surgically treated with curettage and β -tricalcium phosphate grafting. Bone healing occurred, and the patient was free of pain.¹⁶

Here the authors report the case of a solitary hemangioma involving the radius shaft in a 2-year-old boy, causing recurrent pathologic fractures. The patient underwent lesion curettage and grafting with CSP.

CSP is a bioabsorbable bone substitute successfully used in the treatment of bone defect in adult and pediatric populations.^{9,21,23,24} Being osteoinductive and osteoconductive, CSP allows progressive bone formation while they gradually resorb. Furthermore, the structural composition of hemihydrate CSPs allows a pretty complete absorption by an average period of 30 days from grafting into the bone defect. Early resorption allows the examiner to clearly identify bone structure modifications. In this case, complete bone formation was observed 5 months after index surgery with no evidence of recurrence.

Table 1 Reported cases of solitary intraosseous hemangioma of the appendicular skeleton

Author (year)	Age	Sex	Location	Site	Clinical presentation	Treatment	Follow-up	Complications
Geschickter and Maseritz (1938) ¹⁰	8 y	F	Humerus	MD	Fracture (X2)	Radiation therapy, bone graft	–	Recurrent fractures, persistent lesion
	10 y	M	Radius	MD	Trauma, persistent pain, swelling at 2 y	En bloc excision + bone graft	–	None
Sherman (1944) ¹¹	1 m	F	Femur	Me	Swelling for 1 mo, gait problem, pain	Curettage	23 mo	None
Loxley et al (1972) ¹²	15 y	F	Ulna	D (mid)	Painful mass for 3 y	Excision	–	None
Sugiura (1975) ¹³	11 y	F	Tibia	D (mid)	Painful swelling for 5 y	Excision	18 mo	None
Pena et al (1985) ¹⁴	16 y	F	Fibula	D (distal)	Incidental finding	En bloc excision	–	None
Melchior et al (1991) ¹⁵	15 y	F	Tibia	MD	Pain for 3 y	Trepanation; curettage; en bloc excision + bone graft	15 y	2 recurrences
	7 y	F	Tibia	Me	Painful swelling	Curettage + bone grafting; partial curettage; curettage + Gelfoam	10 y	3 recurrences in the talus, 1.5 cm leg shortening
Matsumoto et al (2000) ¹⁶	18 y	F	Ulna	Me	Elbow pain	Curettage + tricalcium phosphate grafting	–	None
Puig et al (2006) ¹⁷	7 y	F	Ilium	–	Hematuria (?)	Surgical biopsy	2 y	None
Song and Shyam (2011) ³	8 y	M	Proximal femur	Me	Limping gait with abductor lurch	Core needle biopsy/decompression	2 y	Shortening of 1.5 cm in right lower limb (coxa vara and coxa valga)
Ferrari et al (2011) ¹⁸	16 y	M	Femur	ME	Pain	En bloc excision	–	None
Shinoara et al (2013) ¹⁹	9 y	F	Radius	MD	Pain	Resection (x2), En bloc excision	18 y	2 recurrences
Present case	2 y	M	Radius	Me	Pathological fractures (X2)	Excision + calcium sulfate pellets	3 y	None

Abbreviations: D, diaphysis; F, female; M, male; MD, metadiaphysis; ME, metapiphysis; Me, metaphysis.

Although successful, the use of bioabsorbable CSP as bone substitute is not free of complications. A low but consistent complication rate, specifically serous drainage from the wound as the calcium sulfate absorbs, has been reported.^{25–27} This complication rate is higher when the material is used in higher volumes (> 20 mL) or in subcutaneous bones (tibia, ulna).⁸ Lee et al did not identify risk factors associated with the development of a sterile inflammatory reaction to CSP, concluding that any adverse reactions (13.8–19.0%) are essentially self-limited and fairly benign.²⁸ In this patient, only 11 mL of CSP was used, yet no serous drainage from wounds was recorded.

Conclusion

IHs of the appendicular skeleton are extremely rare in the pediatric population. Symptomatic and/or large lesions that may lead to pathologic fractures require complete surgical resection and bone grafting. Artificial bone substitutes such as CSP offer a valid alternative to autologous bone grafting. CSPs have properties that enable fast osteoinduction, as the pellet absorption process takes approximately 30 days, and all without interfering in follow-up imaging. At the present time, the majority of relevant reports include basic science investigations, animal studies, or retrospective clinical reviews of varying degrees of quality. CSP remains a cheap, safe, and reliable bone void filler. The cost of the pellets is largely offset by the shorter hospital stays and reduced health care costs, as secondary surgical procedures are not needed.

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None.

Conflict of Interest

None.

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