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ORIGINAL ARTICLE
Reconstructive

Novel Finding of an Excess Bone in Postaxial Polydactyly of the Foot

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Background: Postaxial polydactyly is a common congenital foot anomaly. However, the severity of the anomaly varies from simple cases with only soft tissue duplication to complex cases with bone and joint disorders. In our clinical practice, we found a new morphological anomaly of postaxial polydactyly. We encountered several cases of postaxial polydactyly with bone fragments located between the fourth and fifth toes. The bone fragments were independent of the joint cavity. The mechanisms underlying its development remain unknown because it is a novel disorder. In the present study, we investigated the characteristics of the excess bone to formulate an embryological hypothesis.

Methods: We examined the frequency and trends in the occurrence of excess bone using data from photographs and radiographs of these cases. An example of a disorder similar to excess bone is mosaic-like alignment, as reported by Iba et al. We also compared the characteristics of the mosaic-like alignment with those of the excess bone. Based on these data and existing embryological knowledge, we hypothesized the origin of the excess bone.

Results: Excess bone and mosaic-like alignments showed different characteristics. Therefore, both were considered completely different disorders. We hypothesized that excess bone was caused by damage to the interdigital ectoderm immediately before interdigital programmed cell death.

Conclusions: We encountered a new form of postaxial polydactyly. This can be a factor influencing the treatment strategy because it can affect alignment and stability.
(Plast Reconstr Surg Glob Open 2024; 12:e5717; doi: 10.1097/GOX.0000000000005717; Published online 5 April 2024.)

INTRODUCTION

Polydactyly is the most common congenital anomaly of the foot, accounting for 45% of all congenital foot anomalies. The estimated incidence is 1.7 per 1000 live births,[1] however, the incidence varies among ethnic groups. In Japan, the incidence is reported to be 4.7 to 5.1 per 10,000 live births.[2,3] Polydactyly is broadly classified into three types: preaxial (radial), axial (central), and postaxial (ulnar) polydactyly.[4] Of these, postaxial polydactyly is the most common, accounting for about 80%[1,5] globally and 86% in Japan.[3]

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Received for publication January 5, 2024; accepted February 15, 2024.

Presented at the 66th Annual Meeting of Japan Society of Plastic and Reconstructive Surgery.

DOI: 10.1097/GOX.0000000000005717

Polydactyly presents with various deformities ranging from a single extra toe connected by thin soft tissue to a complex duplication with bony fusion and deformities of the bones and joints. In our clinical experience with postaxial polydactyly of the foot, we have found several cases of excess bone in the interdigital area. This is fundamentally different from the disturbed alignment of phalanges reported as mosaic-like alignment[3,6] (Fig. 1). No previous studies have reported on this excess bone. Although it has been shown in radiographs and schemas in previous articles,[7] it is considered a metaphyseal or basal bone, and no mention has been made of the excess bone. We investigated how often excess bone occurs in cases of postaxial polydactyly in our hospital and its appearance in relation to the phenotype of postaxial polydactyly. The purpose of this study was to identify trends in the characteristics of excess bone development and discuss the embryological hypotheses that can be derived from these trends.

Disclosure statements are at the end of this article, following the correspondence information.

Related Digital Media are available in the full-text version of the article on www.PRSGlobalOpen.com.

MATERIALS AND METHODS

A total of 54 patients (65 feet) with postaxial polydactyly underwent surgery at Osaka Medical College Hospital (now Osaka Medical and Pharmaceutical University Hospital) between January 2004 and March 2021. In this retrospective study, we reviewed the patients' preoperative and postoperative clinical photographs, radiographs, and medical records. We then extracted data on the age at the time of surgery, preoperative and intraoperative findings, and surgical technique. The information obtained from these data was compared and discussed. First, cases with excess bone and mosaic-like alignment were selected from among our postaxial polydactyly cases and classified based on gross and radiographic findings. Excess bone is a new notion that has not been reported previously. Therefore, we defined this as an excess number of phalanges that deviated from the phalangeal axis. As for mosaic-like alignment, although its existence has been reported, there was no clear definition. We defined mosaic-like alignment as the presence of bones deviating from the phalangeal axis but with a normal number of phalanges. The axis of the phalanx was defined as a line passing through the center of the distal phalanx and the midpoint of the basal phalanx (Fig. 2). By applying these definitions to postaxial polydactyly cases, cases with excess bone and mosaic-like alignment were selected. To categorize the gross and radiographic findings, we used Hirase classification[8] (Fig. 3). We also examined the location of excess bone and surgically resected toes. Based on these data, we compared the trends and

Takeaways

Question: We found a new anomaly of the postaxial polydactyly of the foot. The anomaly is excess bone located in the interdigital area. The etiology of the excess bone is not known. Therefore, further investigation is necessary to determine its frequency and trends.

Findings: Photographs and radiographs were used to gather information and compare data on excess bone and mosaic-like alignment. Based on this data and existing embryological knowledge, we hypothesized the origin of the excess bone.

Meaning: It should be noted that postaxial polydactyly may occur with excess bone in the interdigital area.

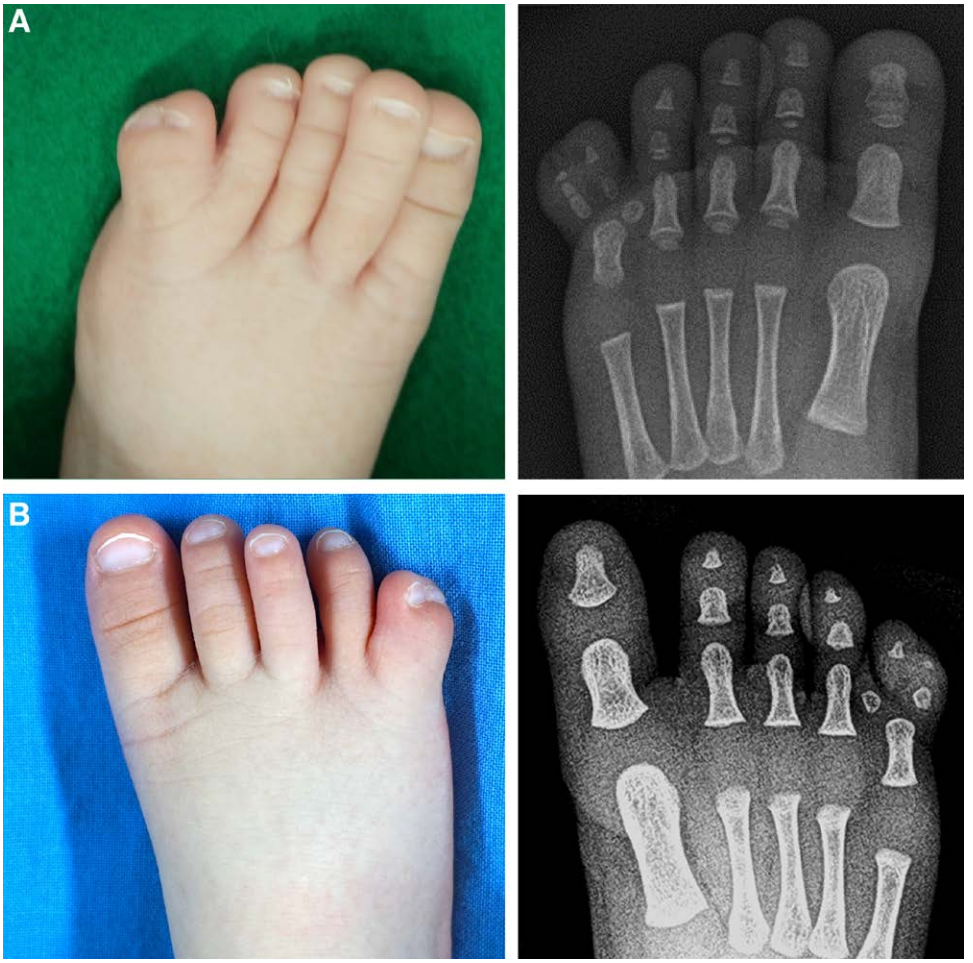

Fig. 1. A, Preoperative photograph and radiograph of excess bone case (Case 3). B, Preoperative photograph and radiograph of mosaic-like alignment case.

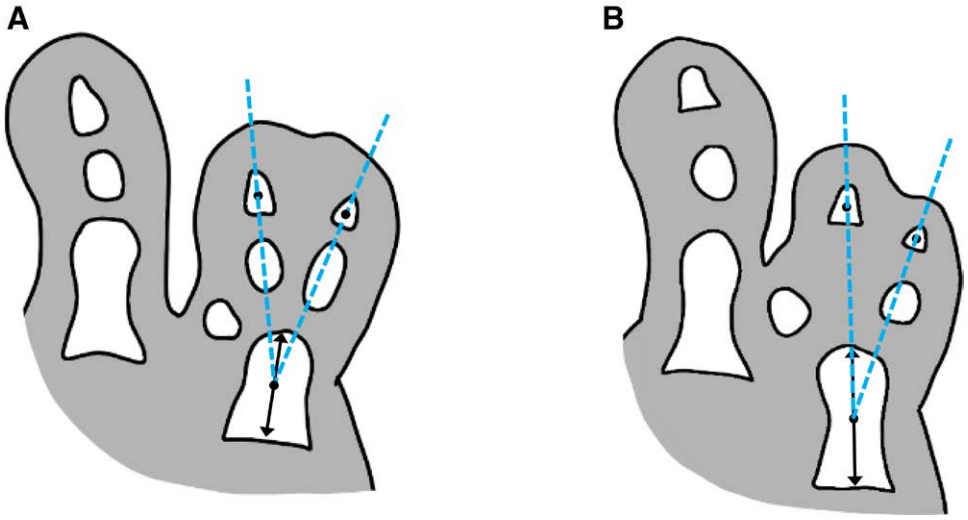

Fig. 2. A, Schema of excess bone definition. B, Schema of mosaic-like alignment definition.

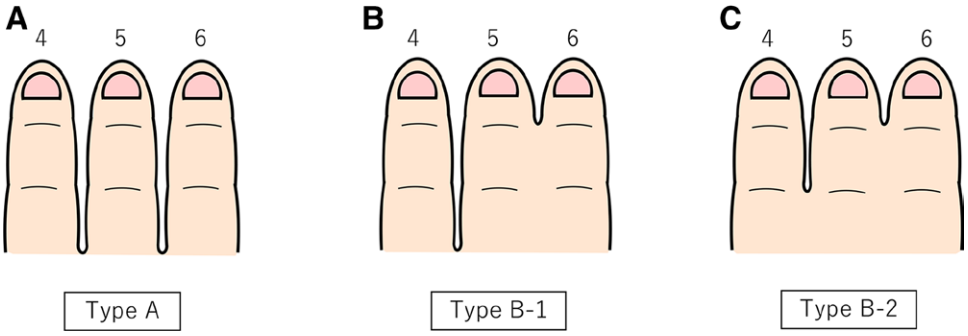

Fig. 3. Hirase classification. A, Type A: each toe is independently separated. B, Type B-1: there is syndactyly between the fifth and sixth toes, but no syndactyly between the fourth and fifth toes. C, Type B-2: there is syndactyly between the fourth and fifth toes and between the fifth and sixth toes.

differences between excess bone and mosaic-like alignment cases. We used three different surgical methods to manage postaxial polydactyly with excess bone: The first was to completely remove the excess bone, the second was to preserve the excess bone, and the third was to split the excess bone in half and preserve half of it. If the medial toe is resected, it is removed entirely along with the resected toe. If there is an axial deviation of the fifth toe of more than 20 degrees, all excess bone is to be removed. Intraoperatively, we check the stability of the toe after removal of excess bone, and if it is unstable, half of the excess bone is returned to its original position (Fig. 4). For mosaic-like alignment cases, we preserve the dominant toe based on gross and radiographic findings similar to that of general postaxial polydactyly. This study was approved by the research ethics committee of Osaka Medical and Pharmaceutical University (No. 2023-036).

RESULTS

Of the total postaxial polydactyly cases, 22 were male infants or boys (28 feet) and 32 were female infants or girls (37 feet). There were 11 bilateral and 43 unilateral cases. The mean age at surgery was 2.3 years (range, 5 months–12 years). Of the patients, five cases (five feet) met the definition of excess bone, with an incidence rate of 7.7% (five of 65 feet). Nineteen patients (22 feet) met the definition of mosaic-like alignment, with an incidence rate of 33.8% (22 of 65 feet). In terms of sex, there were two male and three female cases of excess bone, and 11 male and eight female cases of mosaic-like alignment. All five cases of excess bone were unilateral (Table 1). According to the Hirase classification, the distribution of morphological classification of total postaxial polydactyly cases was that 19 feet (29.2%) were type A, 17 feet (26.2%) were type B-1, and 29 feet (44.6%) were type B-2. The morphological classification of the excess bone showed that the majority of the cases (four of five, 80.0%) were Hirase type B-1 without webbing between the fourth and fifth toes, and only one case (20.0%) was Hirase type B-2 with webbing between the fourth and fifth toes. On the other hand, most of the cases with mosaic-like alignment were classified as Hirase type B-2 (18 of 22, 81.8%), and only a few (four of 22, 18.2%) presented with Hirase type B-1 (Fig. 5). Regarding the distribution of radiographic classification, among all cases of postaxial polydactyly, nine feet (13.8%) were of metatarsal

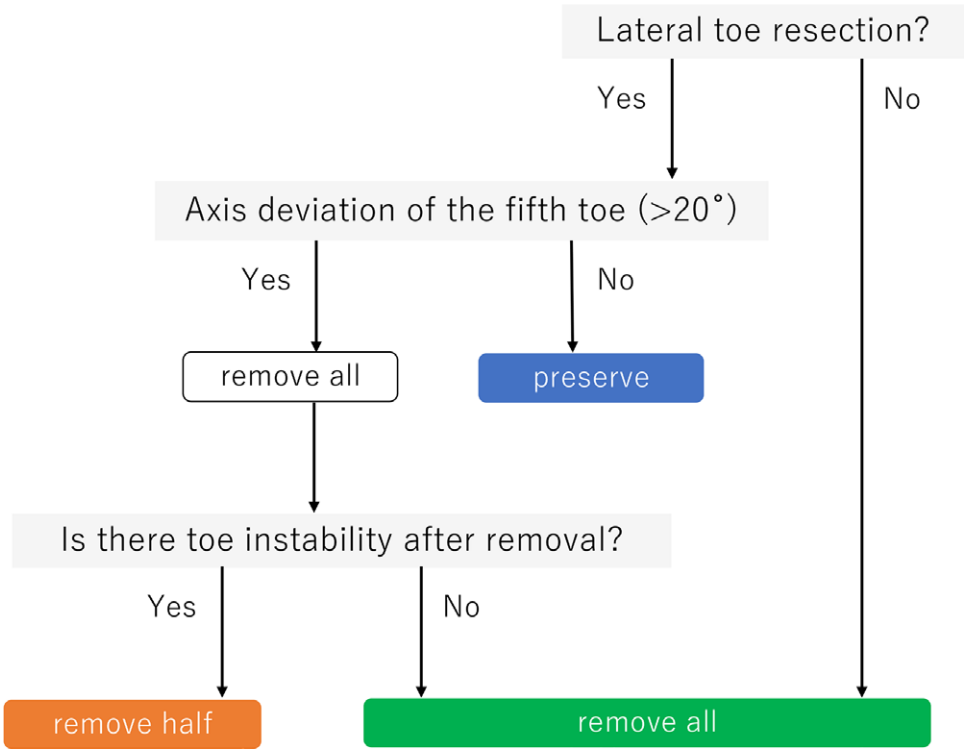

Fig. 4. Excess bone treatment algorithm. The line passing through the center of the fifth distal phalanx and the midpoint of the phalanx just before bifurcation was defined as the ideal axis of the phalanx. Axis deviation was defined as an angle of more than 20 degrees between the ideal axis and the line passing through the center of the fifth distal phalanx and that of the middle phalanx.

Table 1. Incidence of Excess Bone and Mosaic-like Alignment

	Excess Bone	Mosaic-like Alignment
Gross findings	• There is almost no syndactyly between the fourth and fifth toes (Hirase type B-1) • All cases are unilateral polydactyly	• Syndactyly between the fourth and fifth toes (Hirase type B-2) is common • It is seen in both unilateral and bilateral polydactyly
X-ray findings	• Middle phalangeal type or proximal phalangeal type is present • The excess bone is located near the toe bifurcation	• All are middle phalangeal type

type, 18 feet (27.7%) were of proximal phalangeal type, 31 feet (47.7%) were of middle phalangeal type, three feet (4.6%) were of distal phalangeal type, and four feet (6.2%) were unable to be classified. Excess bone cases showed middle phalangeal type (four of five feet, 80.0%) or proximal phalangeal type (one of five feet, 20.0%). All cases of mosaic-like alignment were of the middle phalangeal type (Fig. 6). Excess bone was observed at the phalangeal bifurcation between the fourth and fifth toes.

The operative method for postaxial polydactyly with excess bone included resection of the fifth toe in one foot and resection of the sixth toe in four feet. Of these five cases, the excess bone was removed entirely in two cases, preserved in two cases, and halved in one case (Table 2). Long-term postoperative outcomes in cases of excess bone were satisfactory for all three patterns.

Case 3

The patient was an 11-month-old female infant with postaxial polydactyly of the left foot. The morphological classification was Hirase type B-2, and the radiographic classification was the middle phalangeal type. Preoperative radiographs revealed excess bone on the medial side of the proximal interphalangeal joint of the fifth toe (Fig. 1a). During the first surgery, the lateral toe was removed, and during the second surgery, five months later, interdigital plasty was performed between the fourth and fifth toes. Preoperative radiographs from the second surgery also showed excess bone as independent bone; however, intraoperative findings showed cartilaginous fusion with the proximal phalange of the fifth toe. The excess bone was resected, and cartilaginous fusion with the proximal phalange was observed. In this case, the toe morphology was good, and radiographic images showed good alignment of the toe axis 6 years postoperatively. (**See table, Supplemental Digital Content 1,** which displays the (a) x-ray image of case 3 just before the second surgery, (b) intraoperative photograph of case 3, and (c) radiograph of case 3 at 6 years after the second surgery. **http://links.lww.com/PRSGO/D134**.)

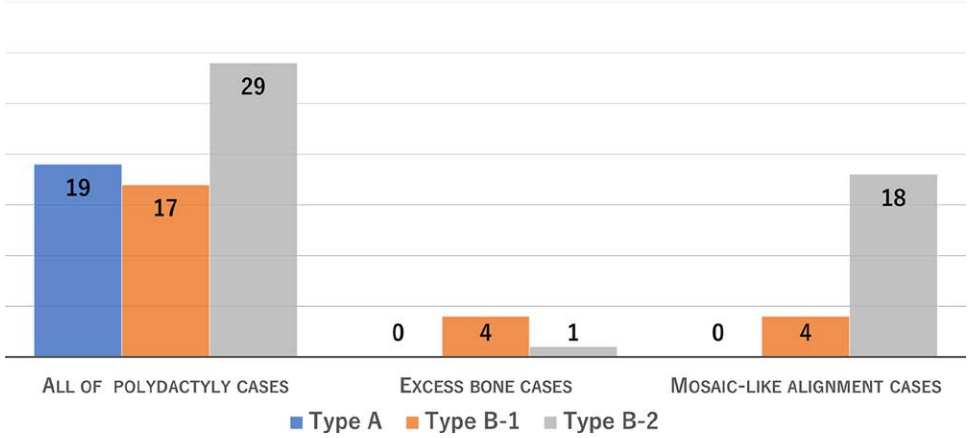

Fig. 5. Distribution of morphological classification.

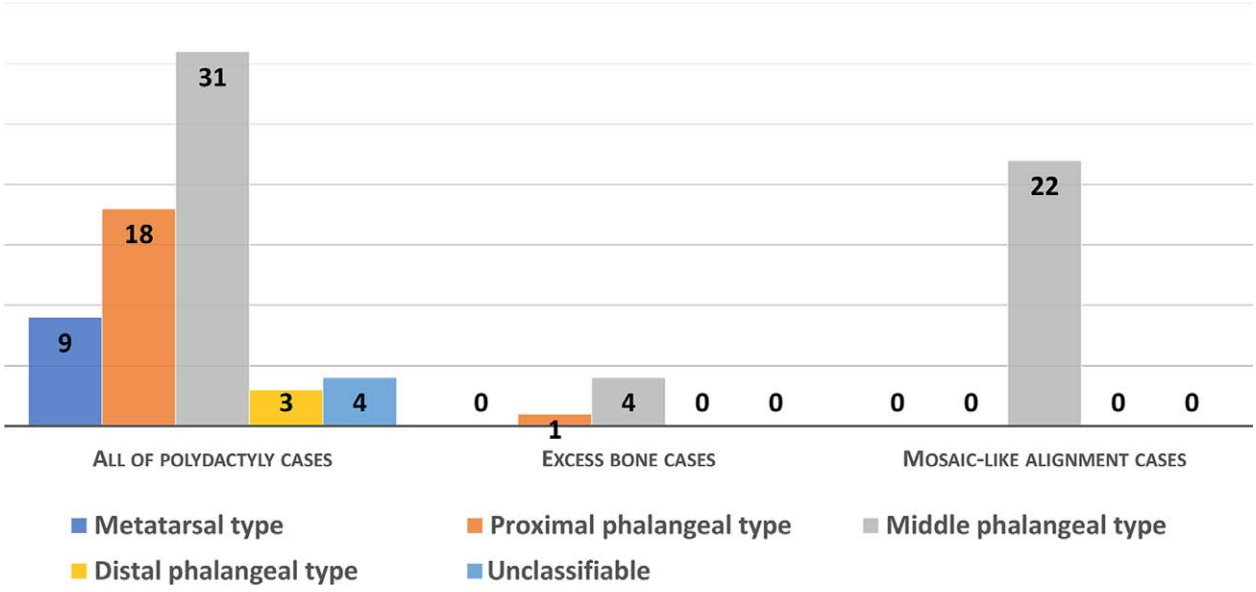

Fig. 6. Distribution of x-ray image classification.

Table 2. Excess Bone Cases

Characteristic	Postaxial Polydactyly (Total)	Excess Bone	Mosaic-like Alignment
No. cases (cases/foot)	54/65	5/5	19/22
Rate of occurrence (%)*	—	7.7	33.8
Unilateral (cases)	43	5	18
Bilateral (cases)	11	0	4
M/F cases	22/32	2/3	11/8
Morphological classification [foot (%)]			
Hirase type A	19 (29.2)	0 (0)	0 (0)
Hirase type B-1	17 (26.2)	4 (80.0)	4 (18.2)
Hirase type B-2	29 (44.6)	1 (20.0)	18 (81.8)
X-ray image classification [foot (%)]			
Metatarsal type	9 (13.8)	0 (0)	0 (0)
Proximal phalangeal type	18 (27.7)	1 (20.0)	0 (0)
Middle phalangeal type	31 (47.7)	4 (80.0)	22 (100.0)
Distal phalangeal type	3 (4.6)	0 (0)	0 (0)
Unable to classify	4 (6.2)	0 (0)	0 (0)

*Incidence of postaxial polydactyly to total.

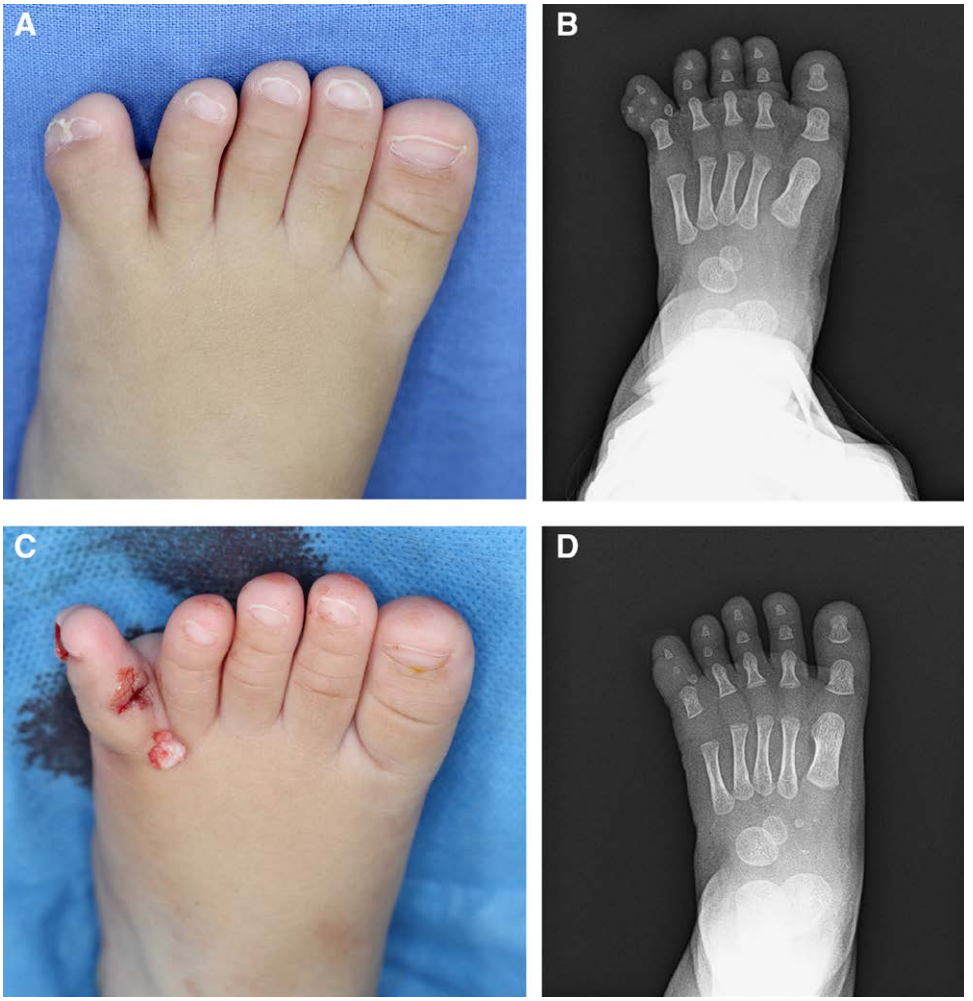

Fig. 7. Excess bone case 4. A, Photograph of the fourth, fifth, and sixth toes. A bulge due to excess bone is seen on the medial side of the fifth toe. B, Radiograph of the fourth, fifth, and sixth toes. Taken at the same time as (A). C, Intraoperative photograph. Removed excess bone was a single bone fragment without joint cavity. The bone was split in half and half put back in place. D, Radiograph on postoperative day 11. The axis of the toe is satisfactory after surgery.

Case 4

The patient was a 1-year-old male infant with postaxial polydactyly of the left foot. The morphological classification was Hirase type B-1, and the radiographic classification was the middle phalangeal type. Preoperative radiographs revealed excess bone on the medial side of the proximal interphalangeal joint of the fifth toe. In the present case, the fifth toe was morphologically and skeletally more dominant than the sixth toe, and valgus was observed in the fifth toe. Therefore, the initial plan was to resect the sixth toe and completely remove the excess bone through another incision. However, toe stability was checked intraoperatively after the removal of excess bone and was found to be unstable. Therefore, the excess bone was cut in half, one of the bone fragments was returned to its original position, and stability was achieved. The postoperative toe shape was good and the postoperative radiograph showed favorable alignment of the toe axis. The long-term outcomes of this case could not be evaluated because the patient stopped visiting the hospital after the one-month postoperative visit (Fig. 7).

DISCUSSION

We compared the gross findings in cases of excess bone and mosaic-like alignment. We found that cases with excess bone rarely showed Hirase type B-2 with syndactyly between the fourth and fifth toes, and one case showed only mild syndactyly. In contrast, most mosaic-like alignment cases showed Hirase type B-2. Radiographic findings showed that the excess bone cases had duplication of the toes at the middle or proximal phalanges, and the excess bone itself was present on the medial side of the fifth toe at the level of the bifurcation of the fifth and sixth phalanges in all cases. In contrast, all patients with mosaic-like alignment showed toe duplication in the middle phalange. The male-to-female ratio in the excess bone cases was 2:3, which was similar to the ratio in all postaxial polydactyly

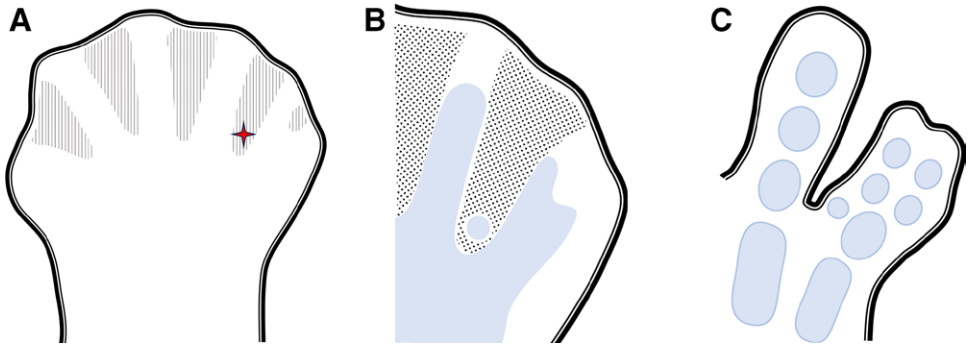

Fig. 8. Schema of the embryological hypothesis of excess bone. A, Just before interdigital PCD occurs, a small wound is made at the interdigital fate of the foot plate. B, Ectopic mesenchymal aggregation occurs at the region of the small wound. The rest of the interdigital area develops normal PCD from the proximal to the distal side. C, Ectopic mesenchymal aggregation becomes cartilage and eventually ossifies to form excess bone.

cases. In contrast, mosaic-like alignment cases had a male-to-female ratio of 11:8, which differed from the overall ratio. (**See table, Supplemental Digital Content 2**, which displays the characteristic findings of excess bone and mosaic-like alignment. **http://links.lww.com/PRSGO/D135**.) Findings, such as the high-incidence phenotype and the presence or absence of syndactyly, were different in both cases. Therefore, we believe that excess bone and mosaic-like alignment have completely different pathologies. Considering the incidence of excess bone in total postaxial polydactyly that we have experienced (7.7%), it is likely that the occurrence of excess bone is not accidental but present in a certain number of cases.

It is known that limb bones develop through endochondral ossification. In the lower limb, the tip of the lower limb bud becomes the footplate at Carnegie stage 16 (fetal day 38), and mesodermal condensation occurs in the mesodermal tissue of the footplate. Mesodermal condensations in the foot that occur at Carnegie stage 18 (fetal day 44) are called toe rays. Within these toe rays, chondrification of cartilage precursors occurs, followed by ossification of the cartilage to form phalanges.[9–11] It has also been reported that phalanx formation and finger morphogenesis occur simultaneously. The number of finger rays is believed to vary according to the size of the limb buds. Three factors strongly influence the size: the number of progenitors that start up the bud, the rarity of proliferation, and the amount of cell death. When these factors are out of control, it results in a change in the autopodial pattern and number of fingers. Syndactyly is also caused by incomplete cell death in the interdigital areas.[11,12]

We suspect that the development of excess bone in the interdigital area may result from an abnormality in the timing of mesodermal condensation in the area of the future toe. We also noticed that the timing of programmed cell death (PCD) in the interdigital area coincided with the appearance of toe rays (mesodermal condensation in the footplate) and interdigital notches. By observing the morphological localization and sequential changes in PCD in developing fetal mouse limbs, Kimura et al reported that programmed cell death in the interdigital region of the limb begins in the proximal mesenchyme and not in the distal region, progressing from the proximal to the distal region.[13] In addition, Hurle and Gañan observed chick leg buds and found that ectopic chondrogenesis occurs when part of the interdigital ectoderm is removed just before interdigital PCD occurs, whereas interdigital formation occurs as usual. Because part of it later takes the form of a finger, they suspected that the removal of the interdigital ectoderm changes the morphogenetic fate of the interdigital tissues from the interdigital to the finger. They also reported that ectopic cartilage ossification occurred at the base of the interdigital region when the interdigital ectoderm of the dorsal leg bud was excised.[14,15] We believe that this experimental model is the best to explain the etiology of excess bone. We hypothesized that if the same phenomenon occurred in humans, it would seem as excess bone. In our cases, all the excess bone was at the bifurcation site. They rarely experience syndactyly, and even if it does, it is mild. This suggests that a small wound produced by a stimulus or trauma in the interdigital ectoderm just before PCD causes ectopic interdigital mesodermal condensations at the base of the interdigital region even in our cases. Then, it is possible that PCD developed from the interdigital base after ectopic mesodermal condensations, resulting in normal interdigital PCD in the area where the excess bone occurred (Fig. 8). Based on our hypothesis, we determined that it is appropriate to refer to this bone as excess bone.

In addition, we believe that excess bone is different from a mere disorder of the phalanx alignment, such as mosaic-like alignment. Both excess bone and mosaic-like alignment are factors that make the treatment plan difficult; however, if the causes are different, a surgical plan should be considered for each. The goal of polydactyly surgery is to reconstruct a toe with joint stability and appearance equivalent to a normal toe that allows the patient to wear normal footwear and walk without pain. Although polydactyly is one of the most common congenital anomalies, there are no standardized, evidence-based classification or treatment guidelines for it. There is a great deal of discussion as to whether the medial or lateral toes should be removed, but this issue remains unresolved. We chose to resect the less dominant toe in

all cases of postaxial polydactyly, considering the phalanx alignment and contour. However, it is difficult to choose a surgical method for complex polydactyly cases, such as those with excess bone and mosaic-like alignment. For mosaic-like alignment cases, we apply the same strategy as for common postaxial polydactyly. In another approach, Iba et al reported a method in which the excision of the toe and protruding bone was performed without considering the alignment of the middle phalanx, and joint stability was reconstructed using the proximally based ligament and periosteal sleeve from the resected toe.[3,6] To deal with excess bone, we took three approaches based on our original algorithm. Because there are various forms of polydactyly, it is difficult to determine which surgical method is the best. In the future, we need to increase the number of cases and evaluate the postoperative results of each surgical method to determine the best approach for such patients.

Our hypothesis regarding excess bone has some limitations that should be considered in the future. We performed surgery for postaxial polydactyly at the age of 1 year. Radiographs of the toes at this time show that a part of the phalanx has just appeared, and it is sometimes difficult to predict the entire phalanx form from radiographs in all cases. Nakamura et al compared the skeletal form directly observed during surgery with that obtained immediately before surgery. Subsequently, they reported that the bone that exists close to the medial or lateral side of the proximal phalange is often a branch of the proximal phalange rather than the middle phalange.[16] The cases we identified under direct observation were different. However, we cannot rule out the possibility that other cases identified as having excess bone or a mosaic-like alignment based on preoperative radiographic findings alone may have been included. Additionally, we did not investigate the occurrence of excess bone in the normal toes in this study. Accessory bone, a common skeletal variant, is observed in the foot and around the ankle joint. It is usually asymptomatic but can be painful due to a variety of causes, including irritation of adjacent soft tissues and bone fractures.[17] If a certain number of bone fragments are identified in the interdigital spaces, not only in cases of polydactyly but also in cases with normal toes, it is possible that there is a parallel development of accessory bone, apart from the development of excess toes.

CONCLUSIONS

We have encountered several cases of postaxial polydactyly with excess bone tissue. Excess bone can cause alignment abnormalities and instability and can be a factor influencing the treatment strategy. In this study, we used three methods to treat the excess bone. We plan to continue our study with additional cases in the future.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

REFERENCES

1. Kelly DM, Mahmoud K, Mauck BM. Polydactyly of the foot: a review. *J Am Acad Orthop Surg.* 2021;29:361–369.
2. Hirahara F. Birth defects monitoring systems—Worldwide and Japan. *Acta Obstet Gynaecol Jpn.* 2007;59:N246–N250.
3. Iba K, Wada T, Kanaya K, et al. An individualized approach to surgical reconstruction for lateral polydactyly of the foot with an emphasis on collateral ligament reconstruction. *Plast Reconstr Surg.* 2012;130:673e–680e.
4. Umair M, Ahmad F, Bilal M, et al. Clinical genetics of polydactyly: an updated review. *Front Genet.* 2018;9:447.
5. Boyle MJ, Hogue GD, Kasser JR. Surgical reconstruction of metatarsal type preaxial polydactyly using an amalgamating osteotomy. *J Pediatr Orthop.* 2016;36:e66–e70.
6. Iba K, Ozasa Y, Hanaka M, et al. Midterm and long-term follow-up after surgical reconstruction for lateral polydactyly of the foot with mosaic-like alignment of the phalanx bones. *J Pediatr Orthop.* 2020;40:e30–e36.
7. Nakamura J, Kubo E, Maezawa N. Our repair procedure for polydactyly of the fifth toe ray. *Jpn J Plast Surg.* 1991;34:1071–1079.
8. Konno M, Hirase Y. A clinical review of polydactyly in the foot and postoperative results of lateral ray polydactyly. *J Jpn Soc Plast Reconstr Surg.* 1997;17:211–225.
9. O'Rahilly R, Gardner E. The timing and sequence of events in the development of the limbs in the human embryo. *Anat Embryol (Berl).* 1975;148:1–23.
10. O'Rahilly R. Early human development and the chief sources of information on staged human embryos. *Eur J Obstet Gynecol Reprod Biol.* 1979;9:273–280.
11. Schoenwolf GC, Bleyl SB, Brauer PR, et al. *Larsen's Human Embryology.* 5th ed. London: Churchill Livingstone; 2015.
12. Talamillo A, Bastida MF, Fernandez-Teran M, et al. The developing limb and the control of the number of digits. *Clin Genet.* 2005;67:143–153.
13. Kimura S, Shiota K. Sequential changes of programmed cell death in developing fetal mouse limbs and its possible roles in limb morphogenesis. *J Morphol.* 1996;229:337–346.
14. Hurle JM, Gañan Y. Interdigital tissue chondrogenesis induced by surgical removal of the ectoderm in the embryonic chick leg bud. *J Embryol Exp Morphol.* 1986;94:231–244.
15. Hurle JM, Gañan Y. Formation of extra-digits induced by surgical removal of the apical ectodermal ridge of the chick embryo leg bud in the stages previous to the onset of interdigital cell death. *Anat Embryol (Berl).* 1987;176:393–399.
16. Nakamura J, Kodachi K, Namikawa H. Polydactyly of the Vth toe ray: a comparison of X-rays taken during infancy and the operative findings. *Jpn J Plast Surg.* 1993;36:133–139.
17. Keles-Celik N, Kose O, Sekerci R, et al. Accessory ossicles of the foot and ankle: disorders and a review of the literature. *Cureus.* 2017;9:e1881.