Tetrapolydactyly: a rare presentation and review of the literature

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Polydactyly is a common congenital anomaly characterized by the presence of extra digits on the hands and/or feet. It can occur as an isolated condition or as part of a syndrome. Polydactyly is typically classified as preaxial, postaxial, or central. Postaxial polydactyly is most frequently observed in African and African–American populations, whereas preaxial polydactyly is more common in Asian populations [1,2].

Tetrapolydactyly, which involves all four limbs, is a rare presentation that can manifest as non-articulated nubbins of soft tissue or fully articulated extra digits involving bone. The condition may occur sporadically or show a familial pattern, often with autosomal dominant inheritance [1-3].

This report describes a rare case of tetrapolydactyly in a female neonate. This case provides valuable insights into the clinical diversity of tetrapolydactyly and underscores the need for ongoing research and documentation.

Case report

This study was approved by the Institutional Review Board of Sapporo City General Hospital (R06-064-1132). Written consent was obtained from the patient for the publication of this report including all clinical images.
The patient is a female neonate born at 37 weeks and 5 days of gestation via cesarean section with an Apgar score of 9 at birth. Throughout the pregnancy, the mother attended regular prenatal check-ups, and the fetus was consistently observed to be in good health. Upon delivery, the neonate was found to have polysyndactyly in both hands and feet and was promptly referred to our plastic surgery department for evaluation.

All members of the patient’s immediate family (father, mother, and older brother) were phenotypically normal. A comprehensive examination did not reveal any other congenital abnormalities. The patient was discharged in excellent condition 3 days after delivery.

The left hand had an additional digit with a bony component on the ulnar side. This extra digit had a metacarpal bone budding from the middle part of the fifth metacarpal bone and contained two phalanges (Fig. 1). Similarly, the right hand had an additional digit on the ulnar side with a bony component comprising two phalanges but without bony articulation or connection at its base (Fig. 2).

The left foot had both preaxial polysyndactyly and postaxial polydactyly (Fig. 3). The preaxial polysyndactyly bifurcated at the first metatarsophalangeal joint, with the skin component conjoined distally at the interphalangeal joint. The postaxial polydactyly bifurcated at the level of the metatarsal bone. The right foot showed postaxial polydactyly, with the additional digit branching off at the level of the metatarsal bone (Fig. 4).

At 7 months of age, the patient was diagnosed with Kawasaki disease and received intravenous immunoglobulin therapy. Otherwise, her growth and development has proceeded normally. She underwent surgical intervention for the extra digits at 9 months of age.

The additional digit on the left hand was excised at the metacarpal level, and the abductor digitii minimi was reattached to the base of the fifth proximal phalanx. On the right hand, the floating type digit was removed without complications, ensuring no residual bone or cartilage remained. For the left foot, the preaxial extra digit was disarticulated at the metatarsophalangeal joint, with subsequent joint reconstruction using periosteum. The postaxial extra digit was excised at the origin from the foot.

Fig. 1. Clinical photograph and radiograph of the left hand at 9 months of age. The images show polydactyly on the ulnar side with an additional finger containing two phalanges articulating at the metacarpal head and budding from the fifth metacarpal bone. The abductor digitii minimi was inserted into the additional finger, and it was reconstructed as the base of the fifth proximal phalanx.

Fig. 2. Clinical photograph and radiograph of the right hand at 9 months of age. The additional finger on ulnar side was the floating type.

Fig. 3. Clinical photograph and radiograph of the left foot at 9 months of age. The left foot showed both preaxial and postaxial polydactyly. The preaxial toe and the first toe had an incompletely divided metatarsal bone, with the additional toe articulating with this metatarsal bone. The postaxial toe was articulated to a metatarsal bone, with a Y-shaped configuration.
metatarsal bone, followed by reconstruction of the abductor muscle. For the right foot, the postaxial extra digit was removed, and the abductor digiti minimi was reconstructed.

Fig. 4. Clinical photograph and radiograph of the right foot at 9 months of age. The right foot showed postaxial polydactyly, demonstrating an asymmetric pattern of polydactyly.

Table 1. Summary of cases of tetrapolydactyly reported in the literature

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Country</th>
<th>Sex</th>
<th>Left hand</th>
<th>Right hand</th>
<th>Left foot</th>
<th>Right foot</th>
<th>Family history</th>
<th>Other abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sener [4]</td>
<td>1990</td>
<td>Turkey</td>
<td>Male</td>
<td>Type III</td>
<td>Type III</td>
<td>Postaxial (M2)</td>
<td>Postaxial (M2)</td>
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<td>Yes</td>
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<tr>
<td>Sener [5]</td>
<td>1995</td>
<td>Turkey</td>
<td>Male</td>
<td>Type IV</td>
<td>Type IV</td>
<td>Postaxial (M2)</td>
<td>Postaxial (M2)</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Radulescu et al. [8]</td>
<td>2006</td>
<td>Romania</td>
<td>Female</td>
<td>Type I</td>
<td>Type I</td>
<td>Preaxial (NA) and postaxial (NA)</td>
<td>Preaxial (NA) and postaxial (NA)</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Mangalgiri and Sherke [9]</td>
<td>2009</td>
<td>India</td>
<td>Male</td>
<td>Type II</td>
<td>Type II</td>
<td>Postaxial (M1)</td>
<td>Postaxial (M1)</td>
<td>NA</td>
<td>No</td>
</tr>
<tr>
<td>Wollina and Verma [10]</td>
<td>2010</td>
<td>Germany</td>
<td>Male</td>
<td>NA</td>
<td>NA</td>
<td>Postaxial (NA)</td>
<td>Postaxial (NA)</td>
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<td>No</td>
</tr>
<tr>
<td>Mukherjee et al. [11]</td>
<td>2011</td>
<td>India</td>
<td>Female</td>
<td>Type II</td>
<td>Type II</td>
<td>Postaxial (M2)</td>
<td>Postaxial (M1)</td>
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<td>No</td>
</tr>
<tr>
<td>Atanda et al. [2]</td>
<td>2013</td>
<td>Nigeria</td>
<td>Female</td>
<td>Type I</td>
<td>Type I</td>
<td>Postaxial (NA)</td>
<td>Postaxial (NA)</td>
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<td>No</td>
</tr>
<tr>
<td>Banerjee and Majumdar [12]</td>
<td>2013</td>
<td>Bangladesh</td>
<td>Male</td>
<td>Type IV</td>
<td>Type IV</td>
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<td>Postaxial (M2)</td>
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<td>No</td>
</tr>
<tr>
<td>Mohammed et al. [13]</td>
<td>2017</td>
<td>Italy</td>
<td>Female</td>
<td>Type III</td>
<td>Type IV</td>
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<td>Postaxial (M2)</td>
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<td>No</td>
</tr>
<tr>
<td>Rossi et al. [14]</td>
<td>2017</td>
<td>Nigeria</td>
<td>Female</td>
<td>Type II</td>
<td>Type I</td>
<td>Postaxial (M2)</td>
<td>Postaxial (M1)</td>
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<td>No</td>
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<tr>
<td>Victor and Victor [3]</td>
<td>2023</td>
<td>Nigeria</td>
<td>Male</td>
<td>Type IV</td>
<td>Type IV</td>
<td>Preaxial (type 3) and postaxial (M2)</td>
<td>Postaxial (M2)</td>
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<td>No</td>
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<tr>
<td>Present case</td>
<td>2024</td>
<td>Japan</td>
<td>Female</td>
<td>Type I</td>
<td>Type I</td>
<td>Postaxial (M2)</td>
<td>Postaxial (M2)</td>
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</tr>
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</table>

Key details are summarized, including the patient’s sex, type of polydactyly observed in the hands and feet, family history, and any other combined anomalies. All anomalies involving the hands displayed a postaxial pattern. Rayan and Frey’s classification [15] was used for ulnar polydactyly in the hand. A modification of the Venn-Watson classification [16,17] was used for preaxial polydactyly in the foot. The SAM (syndactylism, axis deviation, and metatarsal extension) classification system [18] was used to evaluate the extent of metatarsal involvement in postaxial polydactyly in the foot. NA, not applicable due to the unavailability of radiographs or lack of description of findings on physical examination.

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17 involved the feet, and six involved both hands and feet.

We identified several relevant case reports by searching PubMed and Google Scholar using key terms such as “tetrapolydactyly,” “tetrapolysyndactyly,” and “polydactyly 24.” Careful review of the reference lists accompanying these reports led to the identification of 15 case reports. Four of these reports were excluded because no full-text version was available. The remaining cases are summarized in Table 1 [2-5,8-14], utilizing previously established classification systems for each type of polydactyly: the Rayan and Frey system [15] for ulnar polydactyly, the Venn-Watson classification [16,17] for preaxial polydactyly, and the SAM (syndactylism, axis deviation, and metatarsal extension) classification system [18] for postaxial polydactyly. Combined anomalies were found in only one case, which presented with sacrosciatic notch narrowing, which is a minor anomaly. A family history was present in four cases, with no familial tendency in the remaining cases. Interestingly, all the cases with hand involvement showed a postaxial pattern and three showed an asymmetric pattern. Heptadactyly was observed in two cases, including ours. The coexistence of an asymmetric number in the feet and an asymmetric pattern in the hands is unique to our case.

During this research, we observed that the morphological features of tetrapolydactyly can be distressing for parents. However, physicians can provide reassurance by explaining that combined anomalies are extremely rare and that hand anomalies can often be managed effectively by surgical intervention. By offering this perspective, physicians can help to alleviate parental anxiety and support a more optimistic outlook regarding their child’s condition.

This case of tetrapolydactyly in a female neonate adds to the limited body of literature on this rare condition. Further research is needed to better understand the genetic basis of tetrapolydactyly and to develop standardized management protocols.

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**References**