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Case Report

Prenatal Torsion of Radial Polydactyly: A Gangrenous Mass at the Base of the Thumb

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A patient was born with a mass at the base of the thumb approximately 1.5 cm in diameter on the radial side of the fingers. The mass had globular swelling filled with hemorrhagic fluid and was dark red. X-rays and histology of the excised specimen suggested the diagnosis of gangrene and torsion of polydactyly. Prenatal torsion of polydactyly is not a common occurrence; moreover, prenatal torsion of polydactyly has only been found in ulnar polydactyly. Our case is a novel case of radial polydactyly that was gangrenous at birth owing to prenatal torsion. Diagnosing such a mass at the base of the thumb is important.

Key words: infant, fingers, thumb, polydactyly, torsion abnormality

Polydactyly is characterized by the presence of extra digits beyond the normal complement of one thumb and four fingers. Polydactyly is the most frequently observed congenital limb abnormality at birth. The reported prevalence of polydactyly ranges from 5-19 per 10,000 live births, with boys twice as likely to be affected as girls [1]. The degree of duplication varies from small soft tissue nicks to complex, fully developed collateral phalanges and digits [2]. Polydactyly can be classified into radial (preaxial), ulnar (postaxial), and central types, with radial being the most common [2]. Wassel's description of the seven types of thumb polydactyly based on the level of skeletal duplication has gained wide acceptance [3]. Prenatal torsion of polydactyly is less common. We report here a patient with prenatal torsion of radial polydactyly.

Case Report

The patient was a 0-day-old Japanese boy who was born as a first child to nonconsanguineous parents at 38 weeks and 5 days of gestation. His birth weight was 3,188 g. Routine ultrasound examinations performed during pregnancy were normal. There was no family history of polydactyly or other congenital anomalies. He was born with a mass at the base of the left thumb approximately 1.5 cm in diameter. The mass had globular swelling filled with hemorrhagic fluid and was dark red (Fig. 1A-C). There were no other external abnormalities. Cardiac and abdominal ultrasound images were normal, and these ruled out heart and kidney abnormalities. An X-ray of the left hand showed a large soft tissue mass arising from the radial aspect of the left thumb base with osseous elements (Fig. 2A). At 2 days of age, the mass was excised from the base. A histopathological examination of the removed mass was performed. Histopathologically, the lesion consisted of

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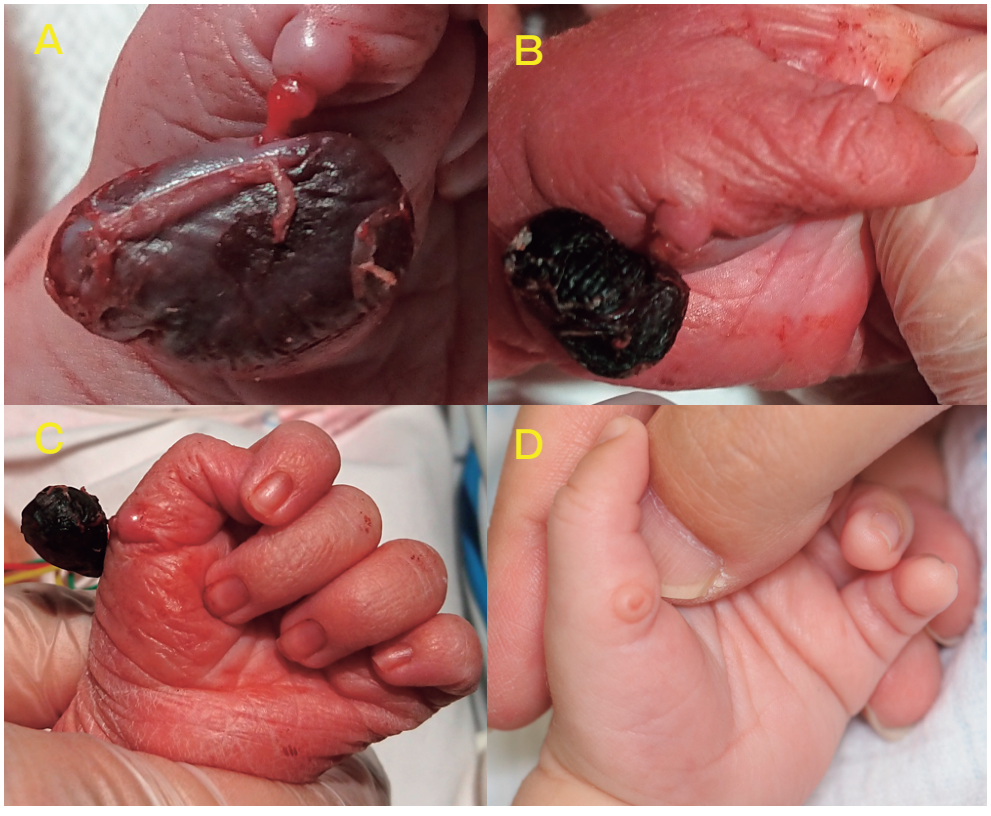

Fig. 1 Images showing torsion of polydactyly. The clinical appearance of an appendage of thumb duplication can be seen. The polydactyl thumb appeared gangrenous at birth. A distal pedunculated, globular, soft, partially compressible, hemorrhagic lesion (approximately 1.5 cm in diameter) was observed. The mass was excised at 2 days of age. (**A**-**C**) Lesion at birth. (**D**) Operated thumb at 1 month of age.

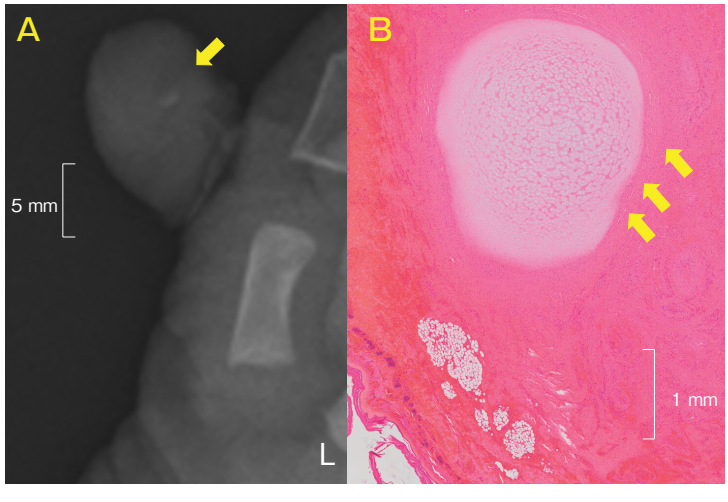

Fig. 2 Results of examinations. (**A**) X-ray of the left hand with a large pedunculated soft tissue mass arising from the radial aspect of the left thumb base with osseous elements. The X-ray shows hypoplastic bone in the phalanx, indicating a duplicated thumb. (**B**) Ulcerated lesion with hyaline cartilage in the center surrounded by fibrofatty tissue (hematoxylin and eosin).

hyaline cartilage tissue in the center surrounded by fibroadipose tissue with marked hemorrhage, which suggested polydactyly. These findings also indicated the diagnosis of gangrene and birth torsion (Fig. 2B). The type of polydactyly was classified as the floating thumb type. He was provided routine care and breastfeeding and was followed closely until 1 month of age. The patient's general condition progressed favorably without any complications of the mass resection surgery (Fig. 1D). His growth and psychomotor development were within the normal range.

Discussion

We present a rare case of prenatal torsion of preaxial polydactyly. Our case is novel because of the finding of radial polydactyly that was gangrenous at birth owing to prenatal torsion.

Postnatal torsion of polydactyly is a well-known complication [4], but prenatal torsion of polydactyly is less commonly reported. We surveyed available English-language studies on "prenatal torsion" or "intranatal torsion" and "polydactyly" using the PubMed database (https://pubmed.ncbi.nlm.nih.gov/) or cited studies. We identified three cases in three reports of prenatal torsion of polydactyly. Kanter *et al.* first described the imminent ischemic necrosis of polydactyly at birth [5]. Saraf *et al.* also described a case of prenatal torsion and gangrene of pedunculated polydactyly [6]. Recently, a case of pedunculated ulnar polydactyly was reported that was gangrenous at birth due to prenatal torsion [4]. All these reports of birth torsion have involved ulnar polydactyly. To our knowledge, there have been no reported cases of radial prenatal torsion of polydactyly, despite the many reports and more frequent occurrence of radial polydactyly. Our case showed radial polydactyly that was gangrenous at birth owing to prenatal torsion. Therefore, our case of prenatal torsion in radial polydactyly can be considered rare. Diagnosing such a mass at the base of the thumb is helpful.

The mechanism of prenatal torsion of polydactyly is thought to be local mechanical trauma; in addition, fetuses have been known to suck their polydactyl digits *in utero*, possibly leading to torsion [5,7]. Prenatal torsion is unlikely to occur in non-pedunculated radial and central polydactyly [4,6,8]. The reason for this lack of torsion is unclear. Our case of prenatal torsion of radial polydactyly is unique. More cases are required to gain a better understanding of the mechanism involved in this process.

Diagnosing patients with a mass at the base of the thumb is important. Polydactyly itself requires detailed clinical, syndromic, and imaging evaluation [6]. A previous case of preaxial polydactyly had a grossly edematous appendage on the radial aspect of the thumb that did not result from prenatal torsion [7]. In such a mass, important differential diagnoses include myofibroma, hemangioma, neurofibroma, leiomyoma, sarcoma, metastatic neuroblastoma, rhabdomyosarcoma, and dermoid cyst [4]. In that case, X-ray examination showed hypoplastic bone in the mass associated with polydactyly [7]. Evaluation of the presence of bone components using X-ray examination may be helpful for diagnosing polydactyly. The torsion of polydactyly invariably leads to pain, infection, and occasionally, considerable bleeding. Torsion of polydactyly should be considered a surgical emergency and should be excised immediately [4,6]. A pathological examination is necessary for a definitive diagnosis.

In conclusion, diagnosing patients with prenatal torsion of polydactyly, such as a mass at the base of the thumb, is important. Our case of prenatal torsion of radial polydactyly is unique in the literature.

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