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Histological findings and treatment in floating-type polydactyly of the thumb: A report of two cases

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Abstract：Here, we report two cases of floating-type polydactyly of the thumb. The supernumerary thumb, including the nail and hypoplastic osseous component, had a narrow and long cutaneous pedicle or a wide base. The histological findings of the supernumerary thumbs were notable for the proliferation of nerve bundles, eccrine ducts, and intramembranous ossification. Suture ligation or surgical excision was selected based on the estimation of the functional outcome from the bifurcation level of the supernumerary thumb, and the parent's preference. No major complications were observed in either of the groups.

Key words：floating-type polydactyly of the thumb, rudimentary polydactyly, traumatic neuroma, eccrine duct, intramembranous ossification

浮遊型母指多指症２例の病理像と治療

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概　　要：浮遊型母指多指症の2例を報告する．1例は茎が細く長いタイプ，もう1例は茎が短く太いタイプであった．除去した余剰指の病理学的検査を施行したところ，神経細胞やエクリン汗管の異常増殖や膜性骨化などの特徴的な所見を認めた．治療については，外来での結紮もしくは全身麻酔下での切除を行い，余剰指の分岐位置より機能的予後を予測し，両親の希望とあわせて方法を決定した．術後は大きな合併症なくおおむね良好に経過している．

索引用語：浮遊型母指多指症，痕跡的多指症，断端神経腫，エクリン汗腺，膜性骨化

【Introduction】

As multiple classifications are available for polydactyly of the thumb, floating-type polydactyly is defined as having a supernumerary thumb without an articulated connection to the dominant thumb. Rudimentary polydactyly is another type that differs from floating-type polydactyly in that a supernumerary thumb manifests a small wart-like appearance, unlike the finger[1,2)]. Rudimentary polydactyly resembles traumatic neuroma[2,3)]; however, the histological characteristics of floating-type polydactyly remain unclear. The treatment of floating-type polydactyly of the thumb is not structured, and the literature is still scant. A slightly higher rate of residual tissue in suture ligation than in surgical resection has been reported[4)]. Recent reports

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have revealed that the degree of thenar development depends on the bifurcation level, which enables us to estimate functional prognosis and surgical outcome[5,6]. Histological findings and treatment selection for floating-type polydactyly of the thumb are discussed in two cases, along with a literature review.

【Report of cases】

Case 1: An 8-day-old boy presented with floating-type polydactyly of the thumb in the right hand but was otherwise healthy. The supernumerary thumb, with a finger-like appearance, was connected to the dominant thumb by a narrow and long cutaneous pedicle approximately 2 cm in length at the bifurcation level between the metacarpophalangeal（MP）and interphalangeal（IP）joints（Figure 1. a,b）. Surgical resection was recommended at 1 year of age due to the concerns about the risk of having residual tissue; however, parents expressed concerns that they may not be able to manage the long pedicle for a year, as the pedicle easily gets caught on things. Suture ligation was therefore performed on that day without anesthesia（Figure 1. c）. Although preoperative radiographs were unavailable, the bifurcation level at the epiphysis of the proximal phalanx was estimated based on the postoperative radiograph with marking at the ligation site（Figure 1. d）. No deviation in the IP joint or nail asymmetry of the dominant thumb was observed. Autoamputation was observed at the outpatient clinic after 2 weeks（Figure 1. e）and no complications were observed since（Figure 1. f,g）.

Suture ligation method: With the help of an assistant pulling the supernumerary thumb, the other surgeon performed suture ligation with a 5/0 nylon suture at the base of the pedicle, as close to the normal skin as possible to prevent residual tissue. Bleeding was minimal throughout the procedure.

Histological findings of the supernumerary thumb: Nail, nonarticulated bone, cartilage, and vasculature were observed. The neural proliferation of the Pacinian corpuscles and Merkel cells was observed in the pedicle dermis（Figure 1. h-l）. Proliferation of the Meissner corpuscles was absent. Thin fibrous tissue suggestive of a hypoplastic tendon was observed in the pedicle. Another notable finding included subcutaneous hyperproliferation of the eccrine ducts（Figure 1. i,l,m）.

Case 2: A 1-year-old girl had floating-type polydactyly of the thumb on the left hand but was otherwise healthy. She also demonstrated a supernumerary thumb with a hypoplastic nail and a wide base（Figure 2. a,b）. Radiography revealed bifurcation at the IP joint with a small osseous component near the base（Figure 2. c）. No IP joint deviation or nail asymmetry was observed in the dominant thumb. As planned at her initial visit when she was 4-days-old, surgical resection at 1 year of age was performed under general anesthesia（Figure 2. d-f）. No major complications other than an asymptomatic small bump suggestive of scarring were observed after 3 months（Figure 2. g-i）.

Surgical method: Resection of the supernumerary thumb was performed at deeper level than the base to include not only the nail but also the small osseous component near it, and the volar skin of the thumb was harvested as a flap to cover the defects（Figure 2. d-f）. The residual nail matrix was ablated using bipolar cautery.

Histological findings of the supernumerary thumb: Neurovascular bundles were within the normal range. Tendons were absent（Figure 2. j）. The proliferation of eccrine ducts and hair follicles was observed（Figure 2. k-m）. The osseous component was identified as a mature bone under the nail plate of the supernumerary thumb at the subcutaneous area. Therefore, we assumed it belongs to the supernumerary thumb, which was unclear from the preoperative radiography of the digit. It demonstrated intermembranous ossification（Figure 2. n,o）, which was believed to be atypical for the phalanx.

【Discussion】

For more than half a century, rudimentary polydactyly has been interpreted as either a hypoplastic supernumerary

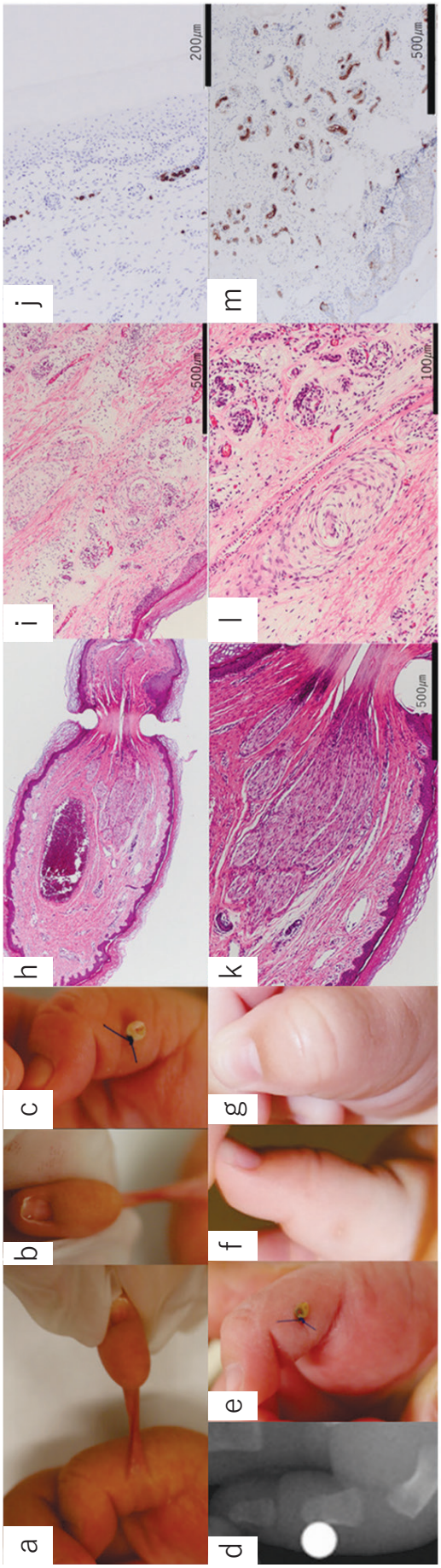

Figure 1. Photographs of floating-type polydactyly of the thumb in case 1. a: Floating-type polydactyly of the thumb with a stalk length of 2 cm. b: The appearance of the radial thumb with the nail. c: Immediately after the procedure. d: Radiography displays bifurcation level at the epiphysis of the proximal phalanx without joint deviation. e–g: Healing course after suture ligation (e: 2 weeks, f: 4 weeks, and g: 2.5 months after the procedure). No nail asymmetry was observed. h–m: Pathological findings, h, k: Neural proliferation at the pedicle in the supernumerary thumb (h: hematoxylin and eosin [HE] × 12.5, k: HE × 100). i, l: proliferation of eccrine duct and Pacinian corpuscle at the pedicle (i, l: HE × 200). j: Merkel cell proliferation at the dermis (j: CK20 × 200), m: Proliferation of eccrine duct (m: epithelial membrane antigen [EMA] × 100).

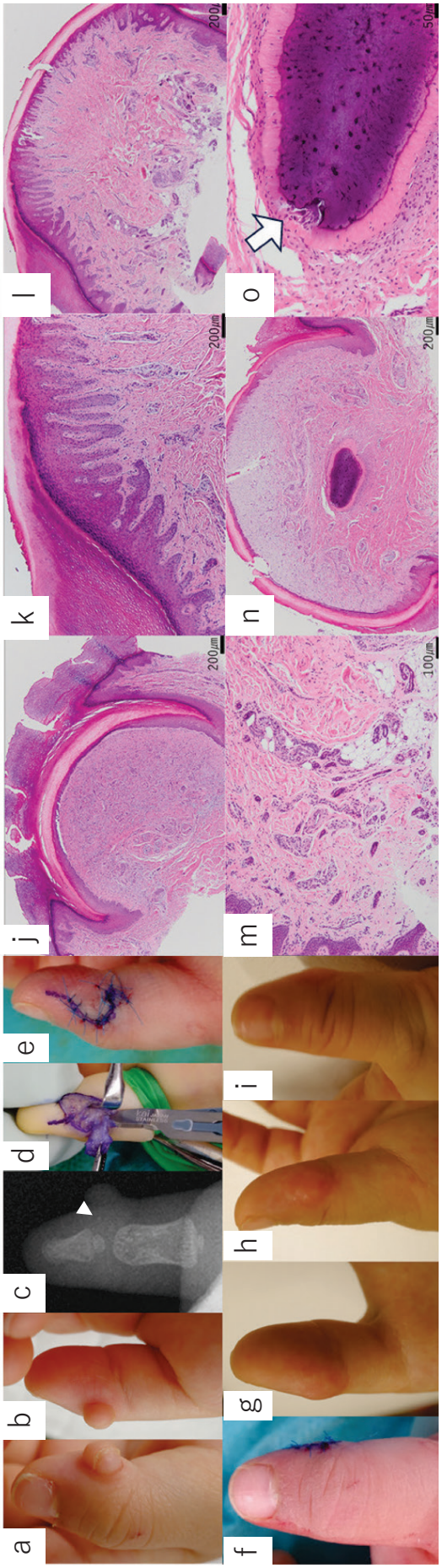

Figure 2. a–i: Photographs of floating-type polydactyly of the thumb in case 2; a, b: Floating-type polydactyly of the thumb. c: Radiography displays bifurcation level at interphalangeal (IP) joint with a small osseous component near the base (arrowhead). d, f: Intraoperative techniques using the skin of the supernumerary thumb as a flap to cover the defect. g–i: Three months after surgical resection, with small scar formation without nail asymmetry. j–o: Histological findings; j: Supernumerary thumb with a nail (HE × 40). k–m: Proliferation of hair follicles and eccrine ducts (k: HE × 100, l: HE × 40, m: HE × 100). n, o: Mature bone of the supernumerary thumb. The white arrow demonstrates osteoblasts, indicating intramembranous ossification (n: HE × 40, o: HE × 200).

digit or a portion of an amputated digit in utero[2]. Although it remains unclear, the latter hypothesis is widely accepted because rudimentary polydactyly histologically resembles a traumatic neuroma, characterized by neural proliferation and an increasing number of Meissner corpuscles, Pacinian corpuscles, and Merkel cells in the dermis[2,3,7-9]. In the literature, these findings were not observed in other types of polydactylies with finger-like supernumerary digits including the floating-type[2]. In contrast, the histological findings in our case 1 were consistent with those of traumatic neuroma, except for the lack of Meissner corpuscles. In another report, wart-like papules on the thumb that included the bone inside to which an extremely narrow skeleton stalk attached were categorized as an intermediate type reflecting the transition between rudimentary and floating-type polydactylies. Moreover, histology at the base of the stalk was suggestive of an amputated neuroma, which further supports the hypothesis that mechanical amputation of the digit occurred in utero[8,9]. Collectively, the histological findings of our case support this hypothesis. Given the fact that Meissner corpuscles start to grow after birth and cytologically mature by 20 to 25 days[10] their absence in our case is explainable. Indeed, according to the literature, numerous Merkel cells without Meissner corpuscles were observed in the rudimentary polydactyly of a 2-days-old baby[11]. The proliferation of eccrine ducts observed in both cases is another notable finding that has not been reported to date. In humans, eccrine sweat glands reach maturity by 2 weeks after birth[12], and the duct length and volume of the coil increase with age[13]. Our cases might have demonstrated the progression of eccrine duct maturity. The overlap of several signaling pathways related to eccrine sweat gland or hair follicle differentiation[14,15] and polydactyly development[16-18] may be another possible explanation. Another rare finding in our case includes intramembranous ossification of the digit. Although the ossification of the distal end of the ungual phalanx has been reported to be intramembranous[19], relationship between the fact and the phenomenon seen in our case is unclear.

Regarding the treatment of floating-type polydactyly of the thumb, more residual tissue is observed with suture ligation than with surgical resection[4], though the literature is scant. As no established structural treatment strategy is available, treatment often depends on the surgeon. As seen in case 1, the long pedicle of the thumb can be an obstacle in daily life. In such cases, suture ligation during the neonatal period is more suitable than elective surgery, even if residual tissue remains and may require additional surgery. However, decision-making during initial treatment should be approached carefully as some cases carry the potential risk of irreversible functional deformities after the procedure. According to a recent study, the degree of thenar development, including tendons that reflect the functional prognosis of the thumb, is near normal when the bifurcation level is more distal to the MP joint, whereas it is confined when proximal, regardless of the morphological maturity of the supernumerary thumb[5]. Indeed, several reports have demonstrated that deformities occur after simple ablation of the supernumerary thumb that is bifurcated proximal to the MP joint[20,21], and are believed to be due to potential immaturity of tendons or possible detachment of the tendons that might have occurred intraoperatively[6]. As estimated before treatment, both cases 1 and 2 revealed that the bifurcation level distal to the MP joint healed well without deformity, which is consistent with the findings of a previous study. Histological findings without aberrant tendon involvement in the supernumerary thumb further supported normal thenar development in both cases. A more detailed study confirmed that nail asymmetry and phalange deformity in the dominant thumb were likely to be observed at the bifurcation level distal to the IP joint or the proximal phalange respectively[6], both of which were not observed in our cases. As neural hyperproliferation at the ligated site was observed in case 1, we should carefully monitor whether postoperative amputation neuromas occur.

【Conclusion】

Our cases demonstrated notable histological findings in floating-type polydactyly of the thumb. Histological examination and estimation of the functional prognosis of

the dominant thumb based on the bifurcation level of the supernumerary thumb can be useful in treatment selection and follow-up observation.

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