

Review of a Case with a Rare Heterotopic Redundant Limb^{*)}

Shoichi WATARI, Touru HIRABAYASHI and Yoshinori FUJIMOTO

*Department of Orthopedic Surgery, Hiroshima University School of Medicine, 1-2-3, Kasumi,
Minami-ku, Hiroshima 734, Japan*

(Received March 23, 1984)

Key words: Redundant Limb, Total limb duplication, Limb malformation

ABSTRACT

Recently a case considered to have an extremely rare heterotopic supernumerary limb was experienced.

The case was a one month old girl who had a supernumerary limb appended on the caudad side of the ninth thoracic vertebra which could not be identified as to whether it was an upper or lower limb. The limb was readily excised, and studies by softex X-ray, arteriography and anatomical procedures were performed. The findings noted are introduced.

INTRODUCTION

Congenital malformations routinely encountered in the realm of orthopedic surgery most frequently involve the extremities, particularly the hands and feet. Of the supernumerary structures, the main congenital malformation involves the digits, as represented by polydactyly of the thumb. However, such redundancies decrease markedly in frequency towards the proximal aspect of the body. For example, mirror hands are one of such abnormalities, and although the clinical picture is widely known, there is hardly any chance to actually observe such a case. Therefore, knowledge on duplication of structures in the proximal portion of the body has been limited to descriptions in textbooks, and thus although they may be understood theoretically, it has been considered difficult to confirm their actual presence. As we recently experienced a case with a heterotopic supernumerary limb, it accorded us the opportunity to study this abnormality. Since the findings revealed it to be an extremely rare case, it will be introduced together with some discussion.

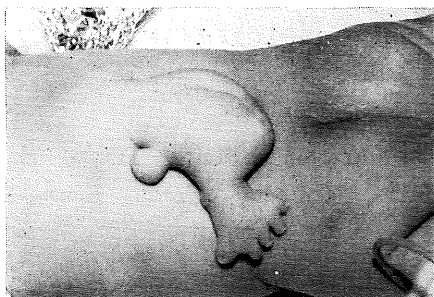
CASE

The father and mother are both 23, and the patient, a girl, was born as their second child. The only sibling, an elder brother, had no abnormalities, and there were no remarkable findings in the parent on their respective families. The mother was diagnosed as being pregnant during the 2nd month of gestation, and as she experienced a few episodes of genital bleeding, she consulted her doctor on each occasion and received injections to prevent miscarriage. She also reports also having taken medication for a cold during the 4th month of gestation. Delivery was at full term and uneventful. Birth weight was 3,820 g. At time of delivery, the presence of an abnormal appearance on the back was pointed out. As this abnormal findings was beyond the generally accepted concept, it caused great psychological unrest to the mother, and she visited our hospital to seek surgical treatment for her baby at age one month. The baby was immediately admitted to our ward.

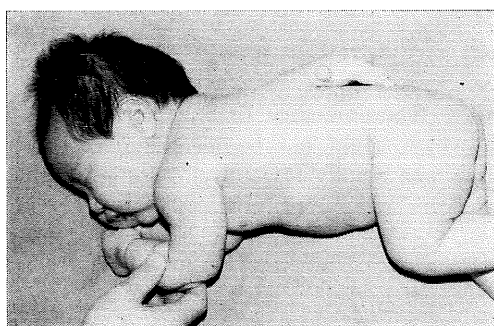
FINDINGS ON ADMISSION

Examination at time of admission showed

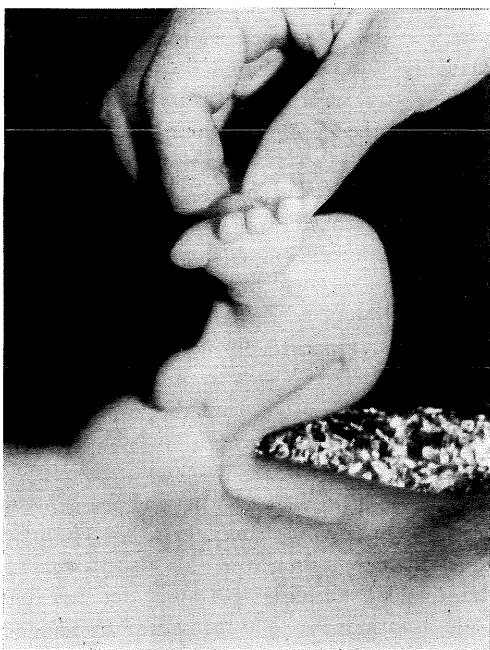
^{*)} 渡 捷一, 平林 徹, 藤本吉範: 異所性過剰肢の一例



A



B



C

Fig. 1. External findings are shown.

A. A frontal view. A supernumerary limb appears from the lower thoracic vertebrae and extends caudadly. It shows a 90 degree curvature where joint-like movement is possible. A round, soft wart-like projection is seen on the concave side of the curvature (inner side). The shape of the tip resembles a foot with five digits, which are short, with those on the inner side being succeedingly larger in size. Morphologically, they resemble toes.

B. A lateral view. The site of appendage is large and the back of this portion appears to be swollen.

C. The supernumerary limb is held up. It is definite that there is a bony structure at the site of appendage. The limb cannot be rotated any further cephaladly.

the startling finding of what may be described as a supernumerary limb appended to the back as shown in Fig. 1. The frontal view is as shown in Fig. 1-A. The appendage was located on the lower portion of the thoracic spine almost on the median line. It curved almost 90 degree to the left at the one third caudad side from its site of attachment. A joint-like movement was possible. Noted on the concave side of the curvature (referred to hereinafter as the inner side) was a round, soft, wart-like projection. The distal end of the limb had five small digits, each having nails, which from the morphologically of the finger pulp, resembled a foot. The pedicle appended to the back was thick and contained bony structure. As shown in Fig. 1-C, it could be lifted, but could not be rotated cephaladly. It was generally soft,

and was fatty tissue-like on palpation. No muscle-like tissue could be discerned and no active mobility could be observed. Simple X-ray views are reproduced in Fig. 2. As can be seen in the frontal view (Fig. 2-A), a long tubular bone extending from the lower edge of the 9th thoracic vertebra caudadly to the upper edge of the 4th lumbar vertebra can be seen to the right of the spinal column, and appears to be attached to a rather flat bone via a joint. However, it cannot be discerned as to whether it is a humerus or a femur, and the bony structures of the distal portion cannot be well visualized.

The surgical treatment of choice in this case was excision, and no difficulty was anticipated. However, the greatest concern was the possibility that there was continuity with visceral organs

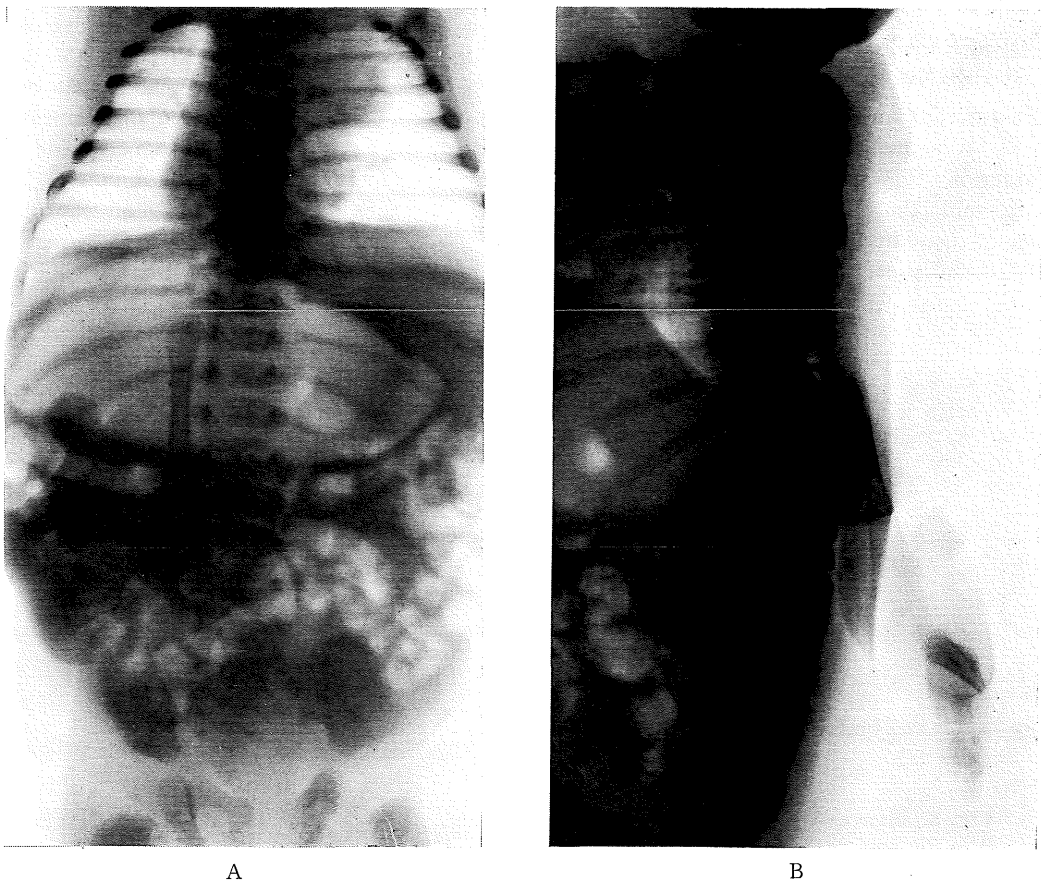


Fig. 2. Simple X-ray images

A shows the frontal view, and B the lateral view. The most proximally located bone of the supernumerary limb seen on the frontal view is a long tubular bone which extends from slightly to the right of T-9 to the caudad side. The other parts are not well visualized.

in the thoracic cavity or with the spinal cord. After careful review from the orthopedic surgery and pediatric viewpoints failed to indicate any such association, excision of the appendage was performed.

OPERATIVE FINDINGS

A curved incision was made on the back in an attempt to detach the supernumerary limb at its base, but in the center was a cartilage-like column. The soft tissue was practically free of structures except for one artery and vein and two peripheral nerves. The nerves were stimulated electrically, but there was no response in the supernumerary limb. Following the cartilage-like column in the center proximally led to a spinous process of the 9th vertebra to which it was attached. Excision of the column together with a portion of the spinous process was carried out.

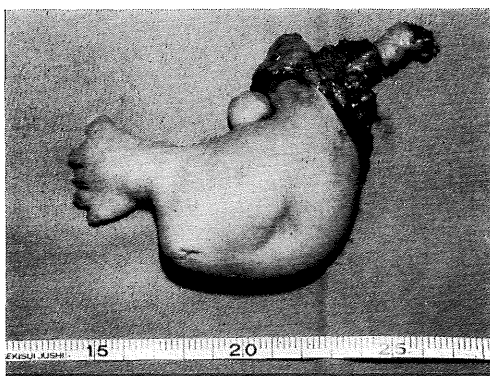
OBSERVATION OF THE EXCISED SUPERNUMERARY LIMB

The external surface is as shown in Fig. 3-A and B. A shows the posterior aspect and B the anterior. The pulp and length of the digital tips resemble those of the foot. However, the nature of the skin on the portion corresponding to the dorsal aspect and that of the sole failed

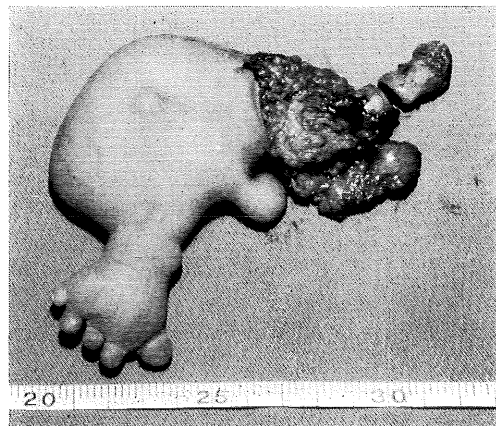
to show any great difference, and as a whole, were almost homogenous. When flexed passively, there was movement in joints which corresponded to the ankle and knee, demonstrating the presence of articulation. On palpation, the limb as a whole felt soft as though it consisted of fatty tissues, and no bony structure could be discerned (Fig. 3-A, B).

With the purpose of elucidating the skeletal composition, Softex X-ray was performed. The findings are as shown in Fig. 4-A.

The bony structure of the proximal-most position consisted of a long tubular bone with its epiphysis at the distal end, while the shape of the proximal end resembled more the humerus than the femur. The structure of the distal end simulated a flat bone with its epiphysis located at the proximal end. The site which apparently corresponded to the foot did not have structure resembling the tarsal bones, but there were 4 bones which were consisted with the metatarsal bones. That on the innermost side was longest, and that on the outermost was rudimentary. The digital bones consisted of five proximal phalanges and five corresponding distal phalanges without any middle phalanges. The inner most digit was longest and most powerful, resembling the great toe, while that corresponding to the 5th digit was



A



B

Fig. 3. External view of excised supernumerary limb.

A is the posterior view and B is the anterior view. Both the anterior and posterior views show the shape of the digits to resemble toes, but there is little difference between the skin which corresponds to the sole and the dorsal aspect.

As a whole, it feels soft and fat-like on palpation. To the right of the site of excision is a separate excised section which is the cartilage-like portion that was attached to the spinous process of T-9. A portion of the spinous process can be seen attached to the tip end,

a floating due to underdevelopment of the metatarsal. Also on the 1st metatarsal was located the 1st and 2nd toes which corresponded to two digital rays, thus suggesting fusion of the 1st and 2nd metatarsals.

Next, arteriography using barium was performed and the image visualized is as shown in Fig. 4-B. The main artery bifurcated a little beyond the joint in the center from where each extended on to the digits. This is a finding common in both the upper and lower extremities, and thus cannot be used as a basis to differentiate as to whether the appendage is

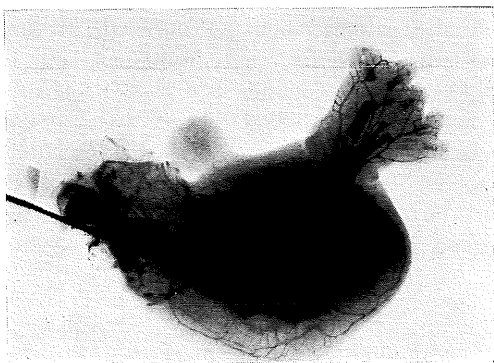
an upper or lower limb.

Lastly, the skin of the limb was stripped to study the internal structure, but it was all fat-like fibrous tissue without any intrinsic structures other than bone, nerve and vessels. Histological study revealed that this soft tissue was merely fibrofatty tissue without any other remarkable findings.

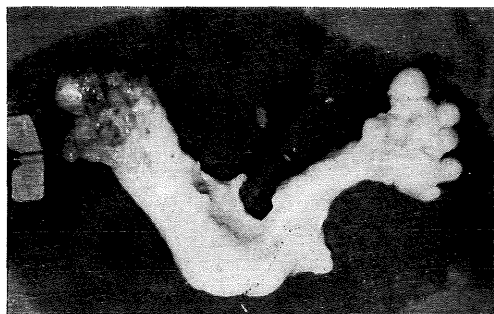
The post operative course was uneventful, and she was discharged after the sutures were removed. At present she has but only a small operation scar, and has no symptoms whatsoever.



A



B



C

Fig. 4. Various reviews made of the supernumerary limb.

A. Findings of Softex X-ray

The most proximal portion of the bone appears to resemble the humerus rather than the femur. The distal portion resembles a flat bone. The site that resembles the foot also appears as a foot on X-ray, but there are other interesting findings such as an image suggesting fusion of the 1st and 2nd metatarsals and floating of the 5th digit.

B. Arteriographic findings

Softex X-ray was taken after injection of barium, but this failed to provide findings to conclude that the limb was a lower extremity.

C. Anatomical finding

After the skin was removed inspection showed it to consist of fibrofatty tissue and no muscular tissue could be seen.

DISCUSSION

According to the textbooks^{6,7)} it is felt two possibilities can be considered responsible for the development of a complete redundant limb. The first is an abnormality which falls under the category of duplication. The definition and classification of duplication provided by Schwalbe⁴⁾ are well known. However, according to his definition it would require at least duplication of part of the body axis, thus this would not apply to our case. One possibility that could be considered is the parasite which comes under asymmetrical duplicitas of his classification, but this was completely ruled out by the operative findings and others. Therefore, the diagnosis for this case could not be sought under a systemic malformation as described above, but would require search in malformations involving the entire upper or lower extremity. That is, the second abnormality which is a double extremity or a polymelia. This entity has been studied in detail by O'Rahilly³⁾ and Weil⁵⁾. O'Rahilly largely classifies the cases into 1) interindividual ambilateral dimelia, 2) intraindividual ambilateral dimelia and 3) intra-individual ipsilateral dimelia. Our case would probably fall under 3) which indicates there is doubling of an ipsilateral limb in one individual, and when this is applied to the upper extremity, it can be classified into dibrachia, diantebrachia, dicheiria and polydactylia. Of these, diantebrachia and dicheiria are well known as so-called mirror hand, and polydactylia is a malformation of the digits of the hand and feet which have the highest frequency of development. However, although it is not clear as to whether the upper or lower extremity is involved in our case, there is no doubt that it is a malformation of the level of the dibrachia, making it rarer in frequency than the mirror hand. Further, classification can be made also on the basis of site, that is, homotopic redundancy and heterotopic redundancy. Our case would come under the latter. The external appearance of the excised supernumerary limb was that of a lower extremity. but there was no evidence on which to draw such conclusion, and thus there was no alternative to diagnose it other than merely a limb. Therefore, our case might properly be diagnosed a heterotopic redundant limb.

Reports on such cases are extremely rare. In Japan, one case reported by Ishii et al.²⁾ in 1982 as a 'third leg'. In their case a duplicate lower extremity was definitely observed appearing from the middle of the right buttock and a complication of various other malformations were present in both legs. Further, Ishii et al. reported that 4 similar cases had been introduced in foreign literature and 1 other in Japan, but we have not yet been able to obtain information on the other case reported in Japan. Of the foreign reports, we found the case of Falton¹⁾ (1904) to be of interest in that he noted slight spontaneous mobility in the hand of the duplicate limb located on the upper extremity and also that there was definite developmental disorder of the entire upper extremity. In our case, there was no muscular tissue in the duplicate limb, and there were no other abnormal findings in the four extremities and body. This is considered to be because it was heterotopically located. Also when the description used by Ishii et al. is applied, ours would have to termed the 'fifth limb'.

Lastly, as to the developmental mechanism of such cases, a fairly well established pathogenesis has been formulated for duplicitas on the basis of animal experiments. However, for heterotopic redundant limbs such as ours, the mechanism responsible is totally unknown, and the only possible explanation is excessive formation of heterotopic limb bud. There are many interesting points with regard to the mechanism involved in the induction of such buds, and thus further research in this area must be performed.

ACKNOWLEDGMENT

The authors are greatly indebted to Professor Kenya Tsuge, chairman of the Department of Orthopedic Surgery, for his kind advices.

REFERENCES

1. **Falton, R.** 1904. Ein Fall von Missbildungen der oberen Extremität durch Ueberzahl. Arch. Anat. Physiol. Anat. Abt: 305-371.
2. **Ishii, M., Izumi, K., Okumura, K., Satomi, T., Kurihara, K. and Maruno, E.** 1982. A case report of a third leg. Teratology 26: 33A.
3. **O'Rahilly, R.** 1951. Morphological Patterns in limb deficiencies and duplications. Amer. J. Anat. 89: 135-193.

4. **Schwarbe, E.** 1907. Die Morphologie der missbildungen des Menschen und der Tiere. II. Die Doppelbildungen. Gustav-Fischer, Jena.
5. **Well, S.** 1922-1923. Diplocheiria und Diplopodie. Z. Orthop. Chir. **43** : 595-607.
6. **Yasuda, M.** 1974. Congenital malformation of the extremities. Current encyclopedia of surgery. 8-B : 167-248. Nakayama Shoten, Tokyo.
7. **Yasuda, M.** 1974. Congenital malformations of the whole body. Current encyclopedia of surgery. 8-B : 249-273. Nakayama Shoten, Tokyo.