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Case Report

Extra-Abdominal Desmoid Tumor of the Hand: A Case Report and Review of the Literature

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KASUKAWA, Y., OKADA, K., HASHIMOTO, M. and SAGESHIMA, M. *Extra-Abdominal Desmoid Tumor of the Hand: A Case Report and Review of the Literature.* Tohoku J. Exp. Med., 1999, **189** (2), 163-170 —— Extra-abdominal desmoid tumor of the hand is rare and only 10 cases have been described in the literature. We present a 14-year-old boy with a recurrent extra-abdominal desmoid tumor in the dorsal site of the right hand. MR image demonstrated the tumor in the third dorsal interosseous muscle, and adhered to the radial side of the forth metacarpal bone. The lesion revealed iso-signal intensity on T1-weighted images and high intensity on T2. We performed a marginal excision. Histological examination of the tumor showed proliferation of the fibroblastic cells with abundant collagen bundles. He developed local recurrence for the third time. The size of the third recurrent tumor has not been changed for 2 years and 3 months. Therefore, we have not performed any additional surgery. Since extensive resection markedly diminishes the function of the hand, we consider that a marginal surgical margin is acceptable for the quality of daily life of patients with a desmoid tumor of the hand. ———— extra-abdominal desmoid tumor; desmoid; hand; recurrence

Extra-abdominal desmoid tumors arise principally from the connective tissue of muscle and the overlying fascia or aponeurosis (Enzinger and Weiss 1995). This tumor was established as a distinct clinical entity by Nichols (1923). Enzinger and Weiss (1995) reviewed 367 cases of extra-abdominal desmoid tumor at the Armed Forces Institution of Pathology (AFIP) over a period of 20 years, and classified the tumor within the spectrum of benign fibromatosis, as they do not metastasize; however, they have the potential to enlarge, to recur, and to infiltrate neighboring tissues in the manner of a fibrosarcoma. The frequent anatomical distributions of the extra-abdominal desmoid tumor were the shoulders, thighs,

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arms, posterior thorax, and buttocks. Desmoid tumors of the hand have been rare, accounting for only 1.1% (4 cases) in the Enzinger' and Shiraki's series (1967), and for only 0.5% (one patient of the 194 patients) in the Rock's series (Rock et al. 1984), and only 11 cases (including the present case) of the desmoid tumors of the hand have been reported.

Prominent clinical feature of desmoid tumor is high rate of local recurrence (Enzinger and Weiss 1995), and the literature review showed that the recurrence rate of extra-abdominal desmoid tumors of the hand is higher than that of other areas. It is a purpose of this report to describe a rare case with a desmoid tumor of the hand, and to mention the high recurrent rate of the tumor of this specific location.

Case Report

A 12-year-old boy first noticed a tumor between the fourth and the fifth metacarpals of the right hand in June 1993. Plain radiographs of the right hand, taken at the other hospital before the primary surgery showed a scalloping of the dorsal-ulnar side of the fourth metacarpal bone (Fig. 1). He had undergone surgical resection at another hospital in July 1993. Pathological diagnosis of the tumor was desmoid tumor. He presented at our institution with a recurrent tumor in the dorsal site of the right hand in November 1995. The recurrent tumor was first noticed in November 1994. Physical examination showed a firm and fixed tumor measuring 3×4.5 cm (Fig. 2) with tenderness between the third and the fourth metacarpals in the dorsal site of the right hand. The surface of the tumor was smooth, and its margin was unclear. The scar of the primary surgery was on the ulnar side of the recurrent tumor. The patient also complained of a contracture at the metacarpophalangeal joint of his ring finger; active extension

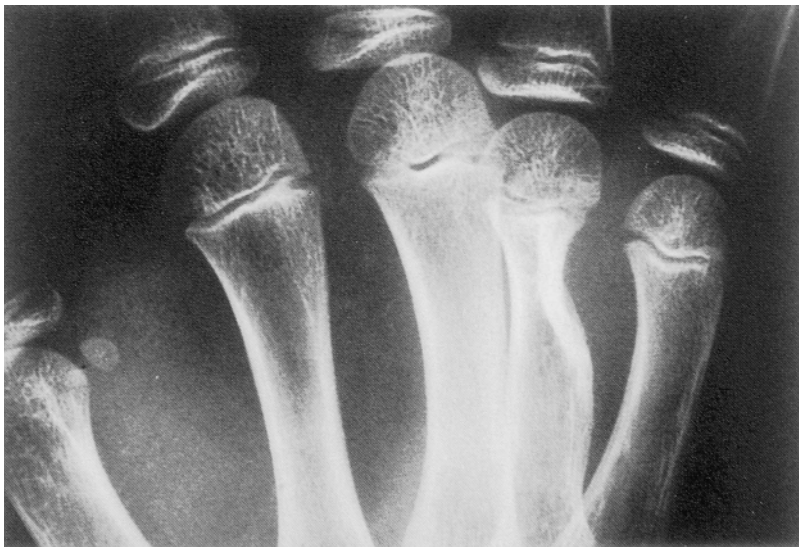

Fig. 1. Radiograph of the right hand taken before primary surgery at the other hospital in 1993 shows a scalloping of the dorsal-ulnar side of the fourth metacarpal bone.

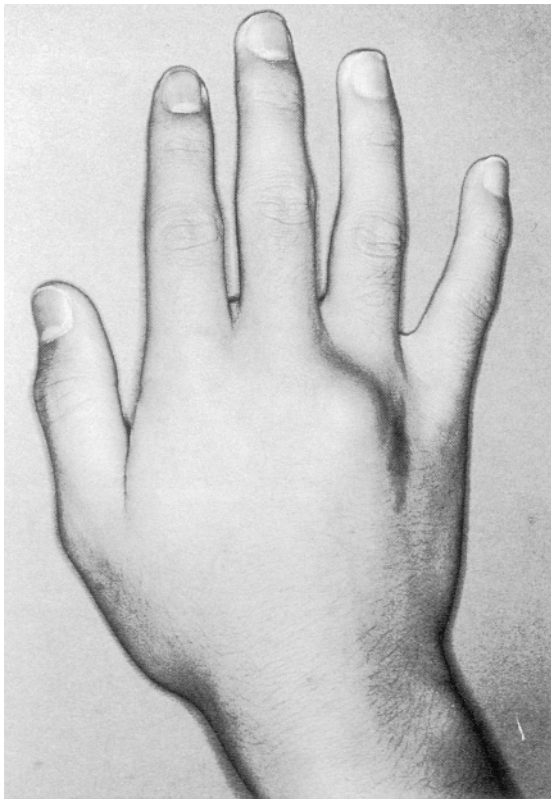

Fig. 2. Photograph of the right hand on admission to our hospital. The tumor was situated between the third and the fourth metacarpal bone, beside the operational scar.

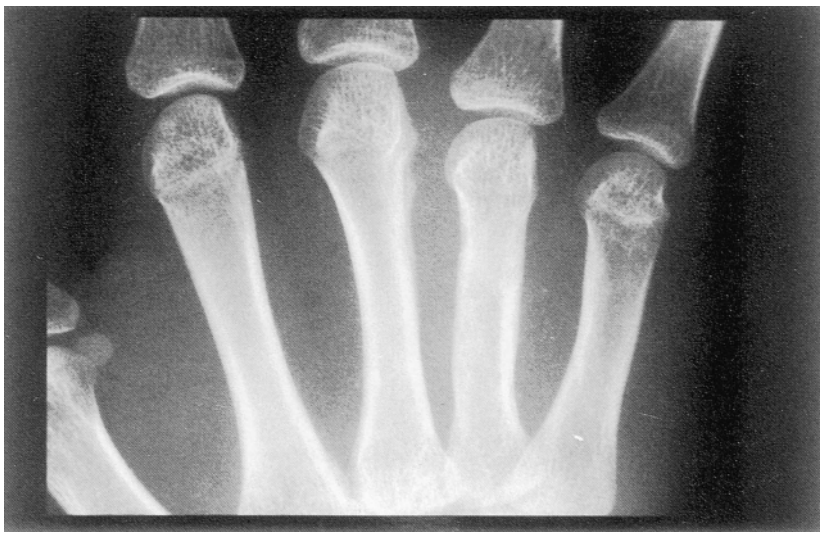

Fig. 3. Radiograph on admission to our hospital in November 1995. Although the scalloping had diminished, mild deformity of the fourth metacarpal bone was observed.

was −45 degrees and passive extension −15 degrees.

Although the bony change had diminished on admission to our institution (Fig. 3), mild deformity of the fourth metacarpal bone was observed, and the tumor adhered to the radial side of the fourth metacarpal bone. MR images

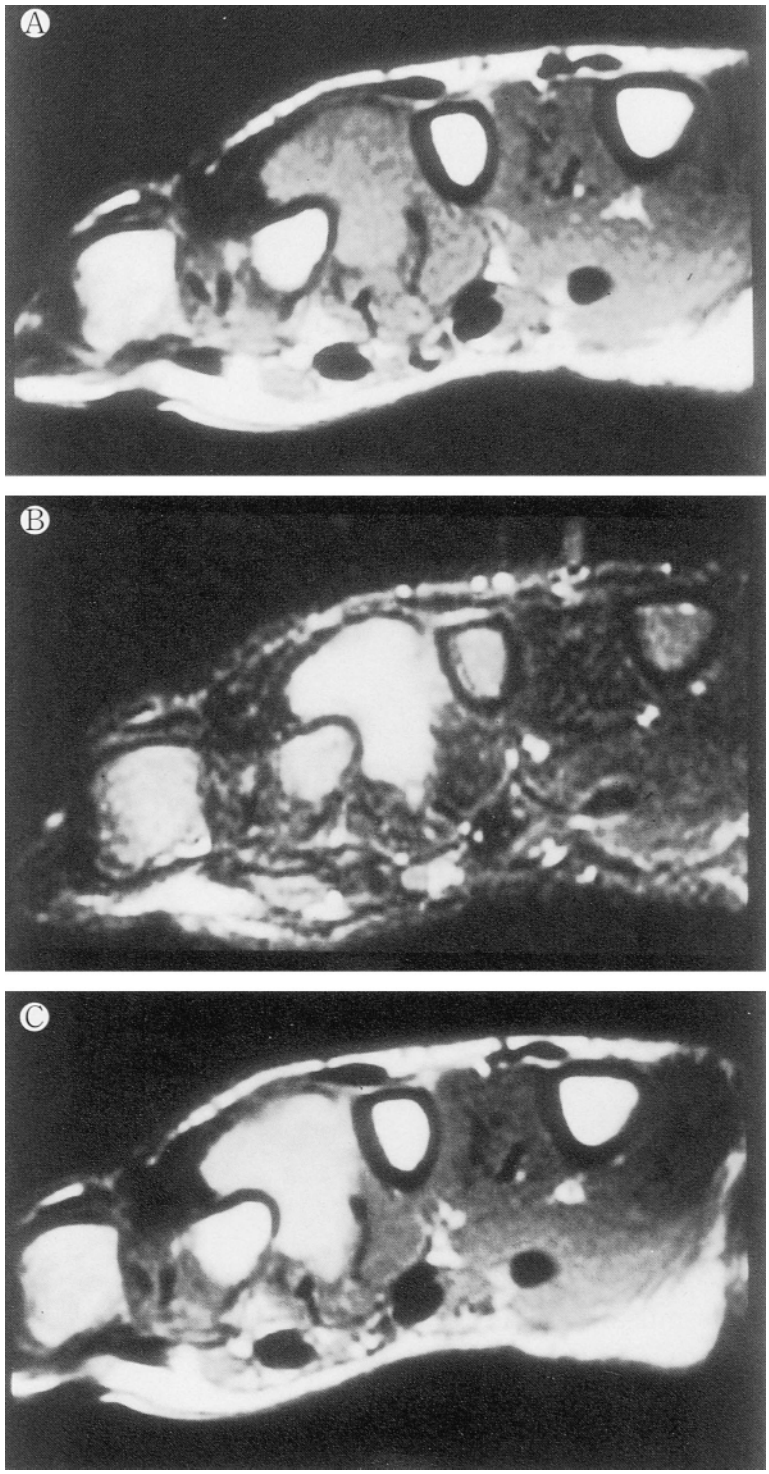

Fig. 4. A: T1-weighted image. B: T2-weighted image. C: post gadolinium MR Images. The lesion revealed a low signal intensity on T1-weighted image and a high intensity on T2, and marked and homogeneous enhancement by gadolinium.

demonstrated the tumor in the third dorsal interosseous muscle. The lesion revealed iso-signal intensity on T1-weighted images and high intensity on T2. The lesion was strongly enhanced by intravenous administration of gadolinium. The margin between the tumor and the third palmar interosseous muscle was

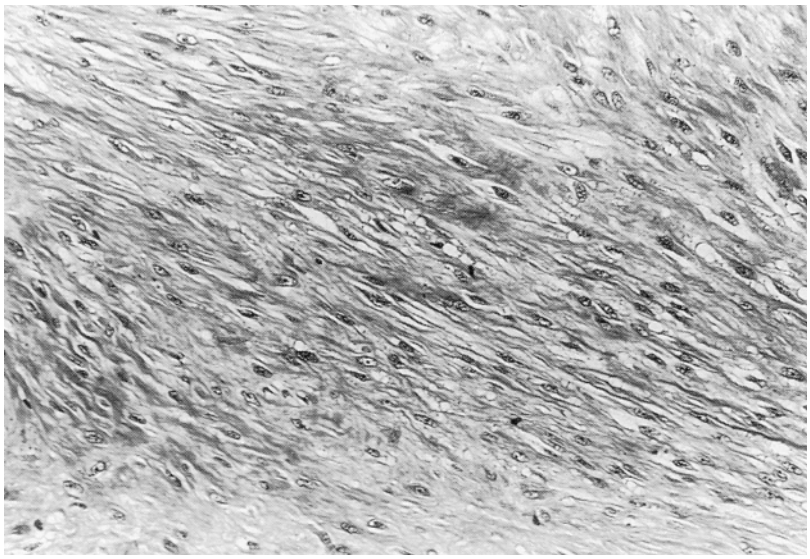

Fig. 5. Pathological features showing the proliferation of fibroblastic cells forming collagenous bundles. Cellularity of these cells was low, and nuclear atypia was mild (H & E, original magnification ×200).

obscure (Figs. 4A, B and C).

In November 1995, we performed a local resection including the entire third dorsal interosseous muscle, the dorsal part of the palmar interosseous muscle, and the periosteum of the radial side of the fourth metacarpal bone. The "wide" surgical margin was achieved except the portion around the fourth metacarpal bone. The portion adjacent to the bone was judged to be "marginal". On the cut surface, the tumor was white and uniform, and there was no hemorrhage or necrosis. Histological examination of the tumor showed proliferation of the fibroblastic cells with abundant collagen bundles. Cellularity of these cells was low, and nuclear atypia was mild (Fig. 5). The histological diagnosis was desmoid tumor. The patient developed a second local recurrence 9 months after the second surgery. The second recurrent tumor was located in the remnant third palmar interosseous muscle. The lesion revealed iso-signal intensity on T1 and high intensity on T2, and there is the erosion of the fourth metacarpal bone. He underwent a further surgical resection in August, 1996. We excised the third palmar interosseous muscle and shaved the fourth metacarpal bone. The surgical margin was "marginal". Seven months after the third surgery, we detected the tiny third local recurrent mass on the palmar side of the fourth metacarpal bone. Since the size of the third recurrent tumor has not been changed for 3 years and 2 months, we have not performed any additional surgery. He has been able to use a pair of chopstiks in the right hand, and enjoy the baseball game without any motion pain.

Discussion

An extra-abdominal desmoid tumor of the hand was initially described by

TABLE 1. *Literature review on extra-abdominal desmoid tumor of the hand*

Author (Year)	Age/Sex	Location	1st Surgery	Rec	Additional Surgery	Bony Adhe	Follow-up
Ritter (1969)	28/F	Hypothenar muscle	Wide	—	None	—	1 y CDF
Hozumi (1971)	40/M	Interosseous muscle	Wide	—	None	—	8 m CDF
Yamauchi (1976)	23/F	Unknown	Excision*	+	2nd: Exhision* 3rd: Wide	+	3 m CDF
Lee (1983)	57/M	Lumbrical muscle	Wide	—	None	—	5 y CDF
Tonai (1984)	52/M	Unknown	Excision*	—	None	—	Unknown
	49/M	Unknown	Excision*	+	2nd: Excision*	+	Unknown
Easter (1984)	14/F	Unknown	Marginal	+	2nd: Excision* 3rd: Lesional	Unknown	6 m AWD
	30/M	Unknown	Marginal	+	2nd: Wide	Unknown	3 m CDF
Segawa (1991)	46/M	Unknown	Excision*	+	2nd: Excision* 3rd: Wide	Unknown	6 y 8 m CDF
Yonezawa (1993)	31/M	Interosseous muscle	Marginal	+	2nd: Marginal 3rd: Marginal	+	5 m CDF
Present case	14/M	Interosseous muscle	Marginal	+	2nd: Marginal 3rd: Marginal	+	2 y 3 m AWD

Excision*, Unknown surgical margin; Wide, Wide excision; Marginal, Marginal excision; Lesional, Intralesional excision, CDF, Continuous disease-free; Adhe, Adhesion; AWD, Alive with disease.

Ritter et al. (1923). Their case was a 28-year-old female, and the tumor was located at the proximal border of the hypothenar muscles of the right hand. After a wide excision of the tumor, there was no local recurrence for one year. To the best of our knowledge, 11 cases (including the present case) of desmoid tumors of the hand have been reported in English and Japanese literature (Ritter et al. 1923; Hozumi 1971; Yamauchi et al. 1976; Tonai et al. 1984; Easter and Halasz 1989; Segawa et al. 1991; Lee 1993; Yonezawa et al. 1993) (Table 1). In these reports, the age of the patients with desmoid tumors of the hand ranged from 14 to 57 years (mean, 34), with male predominance (M : F = 8 : 3). Recurrence rate is extremely high. Nine of these 11 cases had significant follow-up information of recurrence, and in other two cases, informations of local recurrence after the surgery were obscure. In these 9 cases, 7 of the 9 cases (78%) had local recurrence after the first surgery. Since the recurrence rate of extra-abdominal desmoid tumors in the literature was generally 32～68% (Kiel and Suit 1984; Rock et al. 1984; Khorsand and Karakousis 1985; Jones et al. 1986; Markhede et al. 1986; Reitamo et al. 1986; Douglas et al. 1996), it should be stressed that the recurrence rate of extra-abdominal desmoid tumors of the hand is higher than that of other areas. Both information of the surgical margin and follow-up were available in 6 of the 11 cases. Four of the 6 cases had marginal excision in the initial surgery, and all 4 cases developed local recurrence. Two of the 6 cases had wide excision, and remained disease-free for 1 year and 5 years after surgery, respectively. Although it is likely that wide surgical margin is essential to cure the desmoid tumor of the hand, this relationship should be further investigated.

Rock et al. (1984) reported that desmoid tumors of the calf, the foot, the supraclavicular fossa, the popliteal fossa, and the buttock were the most resistant to cure. They stressed that the resistance to cure can be attributed to the technical difficulty of obtaining an adequate margin at operation. We considered that the hand would be one of the most difficult sites of the body to yield complete cure because of technical difficulties obtaining a wide margin resection. In addition, bony adhesion would be responsible for the high rate of local recurrence in the present review of the 11 cases. Six of the 11 cases had information of bony adhesion, and 2 of the 6 cases had neither bony adhesion nor local recurrence, while 4 of the 6 cases had both bony adhesion and local recurrence.

In this review, 4 of the 7 cases with local recurrence were uneventful 3 months to 6 years and 8 months after the second or third surgical treatment, 2 of the 7 cases were alive with disease without any significant disability, and other one was lost for follow up. Since extensive resection markedly diminishes the function of the hand, we consider that a marginal surgical margin is acceptable for the quality of daily life of patients with a desmoid tumor of the hand.

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