Simultaneous presentation of macrodactylia fibrolipomatosis and intestinal lipomatosis

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A 42-year-old woman presented to the emergency department with acute periumbilical pain followed by nausea and vomiting which had continued for 2 days. She had had two prior cesarean deliveries, 7 and 10 years previously, as well as several similar episodes of abdominal pain in the past year, all of which were resolved after conservative treatment. Upon arrival at the emergency room, she had a temperature of 36.4 °C, a pulse of 70/minute, and blood pressure readings of 110/70 mmHg. Her white blood cell count was 10,400/µL with 85.6% segmented neutrophils. Physical examination revealed a palpable soft mass over the right abdominal region without tenderness. The patient also presented congenital macrodactyly, and radiography displayed the bony enlargement of her fingers (Fig. 1). Abdominal Computed Tomography (CT) revealed small bowel lipomatosis with intussusception (Figs. 2 and 3). As a result, the patient underwent laparoscopic surgery immediately. Numerous intramural lipomatous masses of varying sizes were observed intraoperatively between the ligament of Treitz and the distal ileum. There were also some subserosal lipomas on the mesentery. An ileoileal intussusception was reduced extracorporeally and a 35 cm segment of ileum was resected. Surgical pathology revealed multiple submucosal lipomas of 1—7 cm in the intestinal lumen (Fig. 4). The postoperative course was uneventful and the patient recovered completely.

Small bowel lipomatosis is a rare disease even though it can be easily diagnosed by means of CT, given that CT shows diffusion as well as multiple intramural masses with fat densities [1]. Intestinal lipomatosis is a rare condition, and the association of macrodactyly and intestinal lipomatosis is even more unusual. Microscopy confirmed the diagnosis of fibrolipomatosis.

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Fig. 1. Radiograph shows hypertrophy of the third, fourth, and fifth digits of the right hand as well as bony enlargement of these fingers.
obstruction or intussusception often appears in cases of small bowel lipomatosis, which are usually treated by means of surgical intervention. Macrodactyly fibrolipomatosis is a form of macrodactyly in which there is bony enlargement with a diffused overgrowth of adjacent fibrofatty tissue [2]. This mainly affects adjacent digits in a median nerve distribution of the hands. Although both of these conditions are rare, there are three case reports in the literature [3, 4] in which macrodactyly fibrolipomatosis and small bowel lipomatosis are presented together. Unlike the cases previously described in other studies, our patient did not have chronic diarrhea. She had right-handed dactylomegaly, as did the patient described by Kenkare and Ainapurapu [3], which was not limited to a median nerve distribution. The association between these two conditions is unclear, but a complex syndrome with a genetic cause has been considered [4]. Since the reports of the combination of macrodactyly fibrolipomatosis and intestinal lipomatosis have been increasing, this situation seems to not be a casual event and requires further research. Unlike macrodactyly, intestinal lipomatosis would not be diagnosed if there were no related symptoms. A simultaneous presentation of macrodactyly and abdominal pain might be a hint for an early diagnosis of intestinal lipomatosis.

**References**


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**Fig. 2.** Axial view on contrast-enhanced CT reveals small bowel intussusception with a target sign (white arrow).

**Fig. 3.** Coronal view on contrast-enhanced CT reveals multiple dense fatty masses in the small bowel (white arrows) and segmental intussusception in the ileum (black arrow).

**Fig. 4.** Photograph shows the resected specimen, which consists of a 35 cm segment of the terminal ileum. Multiple lipomatous polyps up to 7 x 4 x 3 cm are seen in the intestinal lumen. Congestion and erosion are noted in the obstructed segment (white arrow).