Journal of Pediatric Surgery (2007) **42**, E15–E18

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Journal of Pediatric Surgery

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Mesenteric plexiform neurofibroma in an 11-year-old boy with von Recklinghausen disease

Jinyoung Park*

Department of Pediatric Surgery, School of Medicine, Kyungpook National University, Taegu 700-721, Korea

Index words:
Plexiform neurofibroma;
Mesentery

Abstract We experienced the case of an 11-year-old boy diagnosed as having type 1 neurofibromatosis with intraabdominal and pelvic masses. On physical examination, there were multiple café-au-lait spots larger than 15 mm in diameter scattered over the patient's entire body and axillary freckling, but no cutaneous neurofibromas were present. Lisch nodules were detected in the iris by a slit lamp. A large, firm mobile mass was palpated in the lower abdomen. Abdominal computed tomographic scan showed the hypodense masses in the lower abdomen and pelvic cavity. At laparotomy, a 16 × 9-cm, firm nodular mass along the mesentery of the terminal ileum and ascending colon was found. The mesenteric mass was encasing the superior mesenteric vessels and extending into the serosa of the intestine. The mass was incompletely excised together with affected intestine. However, resection of the rectum could not be performed because of the extensive involvement of the entire mesorectum. Histopathologic study revealed a plexiform neurofibroma involving the mesentery and intestine without evidence of malignant transformation. The postoperative course was uneventful. Ten months after the operation, a magnetic resonance imaging showed no interval change of the mesorectal mass.

Type 1 neurofibromatosis (NF-1) or von Recklinghausen disease is an autosomal dominant neuroectodermal tumor characterized by pigment of the skin (café-au-lait spots), cutaneous and visceral tumors (neurofibromas), Lisch nodules, and systemic abnormalities [1]. Gastrointestinal involvement occurs in about 10% to 25% of patients with NF-1 and includes solitary or multiple neurofibromas, leiomyomas, and rarely, plexiform neurofibromas [2]. Although several cases of mesenteric neurofibroma have been reported in the pediatric populations, involvement of the mesentery and mesorectum has not been previously reported [1,3-7].

Presented as a poster at the 39th Annual Meeting of the Pacific Association on Pediatric Surgeons, Taipei, Taiwan, Republic of China, May 14-18, 2006.

* Tel.: +82 53 420 6452.
E-mail address: kpnugs@yahoo.co.kr.

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doi:10.1016/j.jpedsurg.2007.03.055

1. Case report

An 11-year-old boy was admitted at another institution because of left chest wall deformity. He was transferred to our hospital for evaluation of incidentally found intraabdominal mass in the abdominal and pelvic cavity after an abdomino-pelvic computed tomographic (CT) scan was taken. On physical examination, there were multiple café-au-lait spots larger than 15 mm in diameter scattered over the patient's entire body (Fig. 1) and axillary freckling, but no cutaneous neurofibromas were present. Lisch nodules were detected in the iris by a slit lamp. A 10 × 10-cm, firm mobile mass was palpated in the lower abdomen. These findings were consistent with a diagnosis of NF-1. The family history showed that the patient's mother and older brother also had multiple café-au-lait spots over the entire body, but they never sought medical attention. Initial laboratory studies were

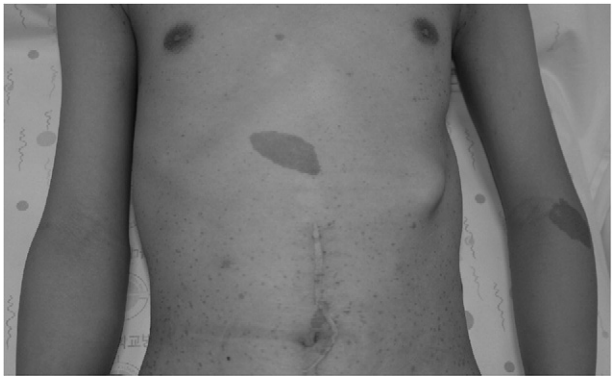

Fig. 1 There were multiple café-au-lait spots larger than 15 mm in diameter scattered over the patient's entire body and axillary freckling.

within normal limits. Abdominopelvic CT scan with contrast medium showed a diffuse, huge hypodense mass with vascular encasement around superior mesenteric vessels (Fig. 2A), as well as extension into the entire mesorectum (Fig. 2B). Ultrasonography-guided needle biopsy was performed and revealed the mesenteric neurofibroma. A laparotomy was performed to confirm the diagnosis and to

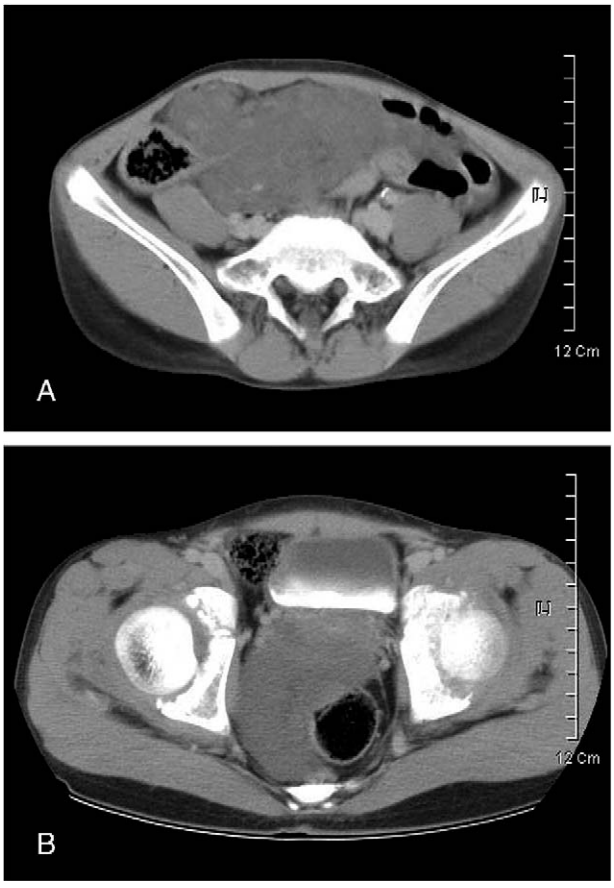

Fig. 2 Abdominopelvic CT scan with contrast medium showed a diffuse, huge hypodense mass with vascular encasement around superior mesenteric vessels (A), as well as extension into the entire mesorectum (B).

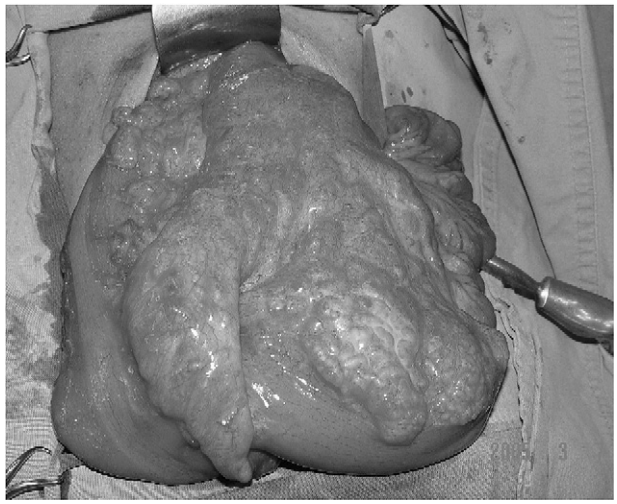

Fig. 3 A firm, nodular mesenteric mass measuring 16 × 9 cm along the mesentery of the terminal ileum and ascending colon was noted. The mass was encasing the superior mesenteric vessels and extending into the serosal layer of the intestine.

exclude a sarcoma. At operative exploration, he was found to have a firm, nodular mesenteric mass measuring 16 × 9 cm along the mesentery of the terminal ileum and ascending colon (Fig. 3). The mass was encasing the superior mesenteric vessels and extending into the serosal layer of the intestine. The mass was incompletely excised together with a 9-cm segment of terminal ileum and a 22-cm segment of ascending colon, and the bowel continuity was restored. However, resection of the rectum was not performed because of the extensive involvement of the entire mesorectum. Pathologic diagnosis revealed a typical plexiform neurofibroma involving the submucosa and muscle layer of the small bowel and colon (Fig. 4). The appendix was also involved with plexiform neurofibroma in the whole layer. However, there was no evidence of malignant transformation. The postoperative course was uneventful. Ten months

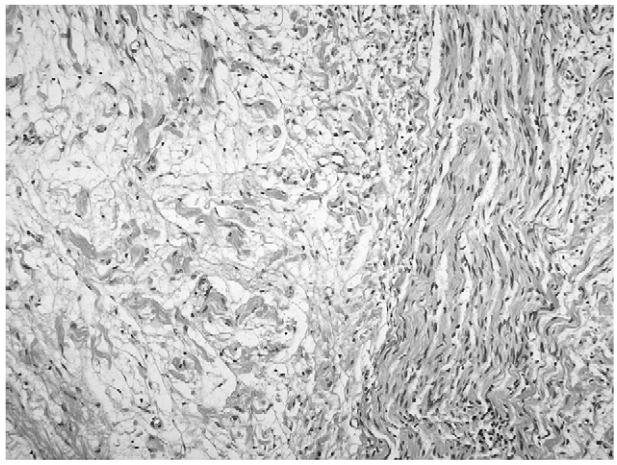

Fig. 4 Microscopic examination shows expansion of the nerve fascicles by pale eosinophilic matrix containing wavy Schwann cells (H&E, ×400 original magnification).