
ULTRASONOGRAPHY OF SMALL INTESTINAL INTUSSUSCEPTION IN NEUROFIBROMATOSIS: A CASE REPORT

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ABSTRACT

Ultrasonographic diagnosis of intussusception is now a part of standard clinical practice. There are reports in the literature in which ultrasonography enabled the identification of their underlying causes; however, there has been no report of ultrasonographic features of intussusception caused by a neurofibroma.

We report a case of small intestinal intussusception caused by a neurofibroma. Ultrasound of the abdomen revealed concentric rings with the lead point formed a complex mass in the center of the intussusception. This was proven on operative and pathological findings.

INTRODUCTION

Intussusception is the leading cause of intestinal obstruction in children, however, in adults it is rare and accounting for 5%-16% of all intussusceptions.¹ About 90% of adult intussusceptions occur in the small or large bowel, and over 90% have a demonstrable causes. Of the small-bowel intussusception, 17% were due to malignant tumors and 40% to benign tumors.² Ultrasonography is considered positive for intussusception if a "target sign" or "concentric rings" is seen in transverse section and a "pseudokidney" or "sandwich" sign is seen in longitudinal section. Ultrasound of intussusception with lead points due to various causes were reported,^{3,4} however as far as we know, there is no publication of the ultrasonographic findings of intussusception due to a neurofibroma.

CASE REPORT

A 41-year old woman presented with a history of intermittent abdominal pain for 2 months. On physical examination, there was a 10 cm diameter soft tissue mass at mid abdomen. The mass was movable and there was no tenderness on palpation. Multiple cutaneous neurofibromas occurring predominantly over the trunk and the forearms were noted. Multiple cafe' au lait spots were also present. The vital signs were normal. Ultrasonography was performed and showed a "sandwich" sign in longitudinal section in the left side abdomen. Transverse section through the apex of the intussusception showed a complex mass, about 2.6X4.3 cm in dimensions causing pressure effect on the compressed echogenic mucosa [Fig 1]. Eleven days later barium enema was

done and revealed a soft tissue mass in the right side of pelvic cavity causing pressure effect on the cecum, appendix, and terminal ileum [Fig 2]. There was also a soft tissue mass causing crescent shadow on the distal ileum in the right iliac fossa. Repeated ultrasonography showed a "concentric rings" with a complex mass at the center in the right lower abdomen [Fig 3]. There was no mass in the left side abdomen. Explore laparotomy was performed 6 days later. Intussusception of proximal ileum into

distal jejunum and numerous palpable polypoid masses along jejunum and ileum (about 1.5 feet from ligament of Trietz to 3.5 feet from ileocecal valve) were seen. Small bowel resection at 1.5 feet from ligament of Trietz and at 3.5 feet from ileocecal valve and end to end anastomosis were performed. Pathological findings showed 40 neurofibromas in the submucosa of resected jejunum and ileum. The largest lesion was about 5 cm in diameter and thought to cause intussusception.

DISCUSSION

Neurofibromatosis is a mesodermal and ectodermal dysplasia with a broad spectrum of clinical



Fig 1 Transverse section through the apex of the intussusception shows a "concentric rings" typical for intussusception. A complex mass is seen in the center of the lesion and causes pressure effect on the echogenic mucosa.

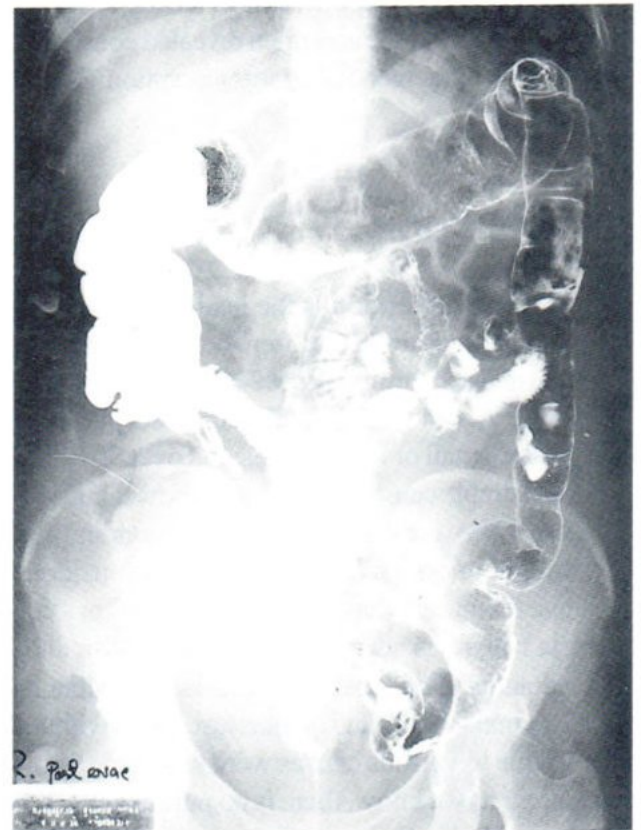


Fig. 2 Barium enema (post-evacuation film) shows extrinsic pressure effect on the cecum, appendix and terminal ileum. A soft tissue mass causing a crescent shadow on the distal ileum is seen in the right iliac fossa.



Fig 3. Repeated ultrasound reveals “concentric rings” with a complex mass at the center of the lesion.

and radiological findings. The disease is characterized by multiple, soft, sessile or pedunculated skin nodules of varying sizes and areas of cutaneous pigmentation (cafe’ au lait spot). The gastrointestinal tract is involved in 25% of patients with neurofibromatosis.⁵ The ileum is most frequently involved. They usually arise from subserosal nerves, Auerbach plexus, or less often from the submucosal plexus. The submucosal lesions are more likely to cause symptoms of recurrent melena or obstruction from intussusception.

The ultrasonographic findings of intussusception is widely recognized. The intussusceptum contained within the intussuscepti has been described as a “target” image with double concentric rings in the transverse section. The echogenic center of the lesion is

formed by the compressed mucosa. The center of the simple intussusception is always a round echogenic area in the transverse section. When a lead point is present, the uniform echogenic center of the intussusception is replaced by a complex mass lesion. The echo pattern of this complex mass depends on the nature of the lead point. When a bowel loop, such as an inverted Meckel’s diverticulum or appendix is present, a complex mass formed by convoluted bowel loops. When a polyp or tumor is present, a polypoid mass arising from the intussusceptum is seen.³ Eustace S, et al.⁴ reported a patient with colonic lipoma-induced intussusception who had increase both in size and echogenicity of the inner core of the leading edge of intussusception.

Although the ultrasonographic findings of a complex mass seen in this patient can be seen in intussusception from other causes, when one encountered a patient with the characteristic skin lesions of neurofibromatosis with a complex mass causing intussusception, the diagnosis of a neurofibroma causing intussusception should be the first consideration.

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