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# Case report: Sigmoid schwannoma as the lead point for intussusception in an adult patient with neurofibromatosis

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# Case Report: Sigmoid Schwannoma as the Lead Point for Intussusception in an Adult Patient with Neurofibromatosis

Adult colonic intussusception is rare and when it occurs over 65% of cases are related to neoplasms.[1] Neurofibromatosis (NF) involves the gastrointestinal tract one third of the time, 5% of which are symptomatic[2]. Lesions are essentially confined to the stomach and the jejunum. The symptoms usually include occult bleeding and obstruction. We report a case of NF leading to intussusception of the sigmoid colon into the rectum.

A 52-year-old man with neurofibromatosis (NF), type II presented with abdominal distention one week status-post resection of an acoustic neuroma. Physical exam revealed a mildly distended, non-tender abdomen, with hypoactive bowel sounds. Abundant mucus output was noted from the rectum which tested positive for occult blood. Digital rectal examination revealed a firm; mobile mass located eight centimeters from the anal verge. Laboratory studies were normal and the patient remained afebrile. Abdominal x-ray suggested pseudo-obstruction. Symptoms failed to improve with a trial of nasogastric suction and supportive care. Computed tomography of the abdomen showed a dilated proximal colon with colonic intussusception. Sigmoidoscopy revealed a bulging, purplish mass (figure 1) in the recto-sigmoid area obliterating the lumen. The mass could easily be displaced using the sigmoidoscope but would quickly return to its position once the scope was withdrawn. The scope was unable to pass beyond this lesion. Laparotomy confirmed the presence of a sigmoido-rectal intussusception, which was resected. (Figure 2)The lead point of the intussusception was a six cm mass which was found to be a plexiform schwannoma, predominantly submucosal and extending into the muscularis propria (figure 3). The abdominal distention resolved following the surgery.

NF is a hereditary disorder associated with café-au-lait lesions, neurofibromas and involvement of multiple organs. The occurrence is 1 in 3500 to 4000 individuals.[3] NF has rarely been reported as the cause of intussusception. Azar et al[1] reported 58 cases of intussusception reviewed over twenty-nine years and found only one to be related to NF. Schwannomas, when found in the gastrointestinal tract, most commonly occur in the stomach. When occurring in the colon, the most common site of schwannomas is the cecum. There are rare reports of NF involvement of the colon presenting as pseudo-obstruction, megacolon and colonic tumor.[4,5]

Schwannomas make up only a small percentage of mesenchymal tumors of the gastrointestinal tract. Miettinen et al[6] reviewed 600 mesenchymal tumors of the colon collected

over twenty-eight years. Only twenty of these tumors were schwannomas. Three occurred in the recto-sigmoid colon and just one in the rectum. An extensive literature search failed to reveal any prior cases of sigmoido-rectal intussusception caused by a schwannoma.

Adult intussusception is most commonly the result of neoplasm. In this patient with neurofibromatosis, we identified a sigmoid schwannoma as the tumor that led to clinical intussusception requiring surgery.

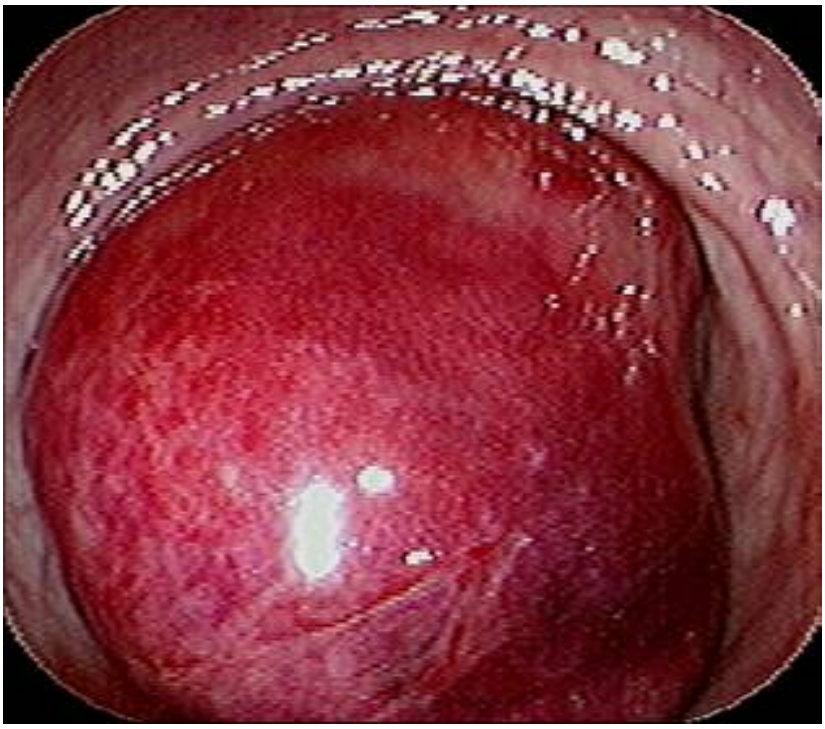

Figure 1: Soft tissue mass protruding in recto-sigmoid area seen during sigmoidoscopy.

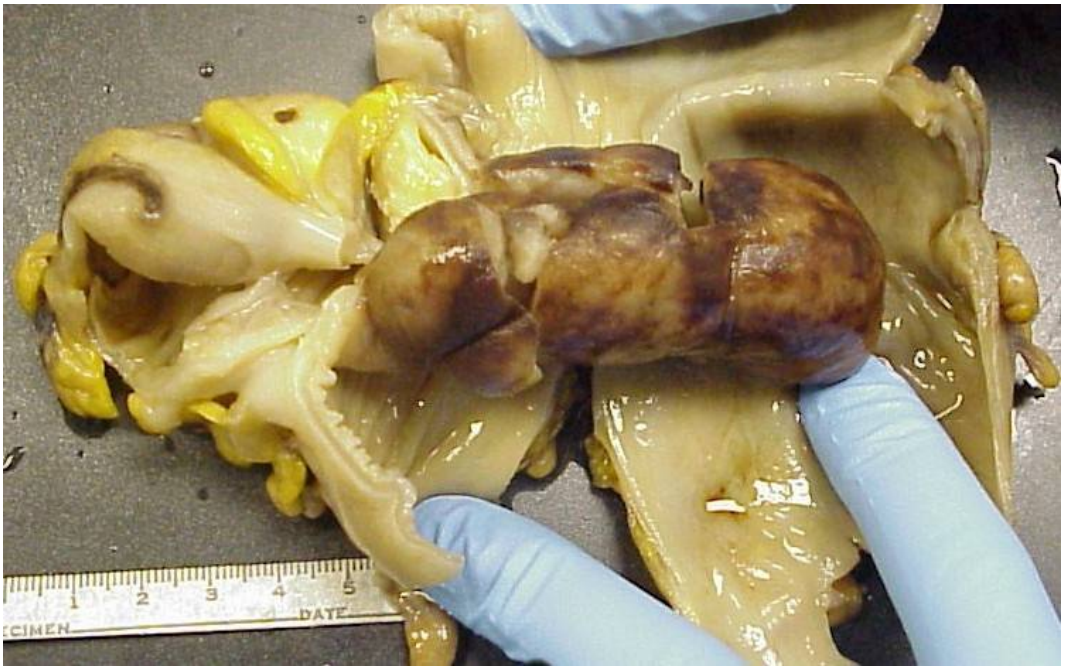

Figure 2: Laparotomy with sigmoid resection showed intussusception of the sigmoid colon. The lead point of the intussusception was a 6 cm mass.

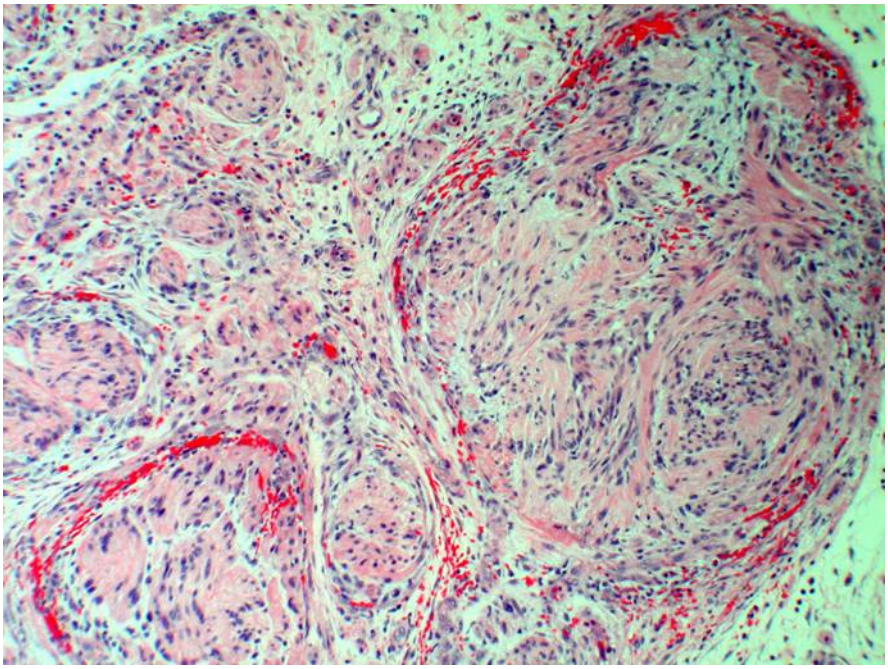

Figure 3: Histology of the mass revealed a plexiform schwannoma, predominantly submucosal and extending into the muscularis propria.

References

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