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**CASE REPORT**

## Incidental finding of Solitary Colonic Neurofibroma: A case report

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**Abstract:**

**Introduction:** Colonic neurofibroma is an extremely rare condition. Solitary Neurofibroma have also been reported in other parts of the gastrointestinal system such as mesentary, esophagus, soft pallet, gall bladder, bile duct and anal canal. Usually neurofibroma colon is asymptomatic but sometime it can presents with abdominal pain, constipation, anaemia, manena or an abdominal mass. Sometimes it may presents with serious complications like intestianl obstruction, obstructive jaundice, ischemic bowel, perforation intussusception, and rarely megacolon.

Solitary nuerofiborma is extremely rare and usually occur in association with neurofibromatosis type-1 (NF-1), Von Reckling Hausen neurofibromatosis or peripheral neurofibromatosis. neurofibromatosis type-II is also central neurofibromatosis or bilateral acoustic neurofibromatosis. This disease has variable clinical manifestation involving the skin, nervous system, and gastrointestinal tract. Gastrointestinal involvement has been reported in 25% of patients with NF-1 with most cases shows involvement of small bowel and stomach.

With such low incidence and few cases reported and its potential for malignant transformation in few cases. Long term follow-up is required.

In our case the histo-pathology showed bundles of neural tissues arising from muscularis propria, the tumor has myxoiv central portion with thin fibrous capsule. The cellular element composed of spindle wavy cell with elongated nuclei. This tumour has low potential of converting into malignancy.

**Conclusion:**Neurofibromas of colon are extremely uncommon. With such low incidence and very few cases reported, long-term follow-up and screening are still undetermined at this time, but close clinical follow-up of these patients is important to exclude neurofibromatosis and its associated risk of malignant transformation.

**Keywords:** Von Reckling Hausen neurofibromatosis, Neurofibromas, intestinal obstruction, neural tissues arising from muscularis propria

**Case History:**

A 23-year-old male presented in the Emergency Department with history of road traffic accident. Patient sustained severe abdominal trauma causing grade-III splenic injury, hemoperitoneum, with manifestations of hypovolemic shock.

No history of medical problems. Patient was resuscitated by ATLS protocol, and after resuscitation an exploratory laparotomy was performed.

After controlling bleeding by splenectomy, an incidental mass arising from Right 1/3rd the transverse colon at the anti-mesenteric border (6x6 cm) was found.

There were multiple enlarged lymph nodes along the transverse and ascending mesocolon. Right hemicolectomy was done. Post-operatively patient has smooth recovery. When patient was reviewed, post-operatively café-au-lait patches were noted on the trunk.

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Figure 1: Patients showing cafe-au-lait spots



Figure 2: Gross specimen of right hemicolectomy



Figure 3: microscopic picture showing bundles of neural tissue arising from the muscularis propria layer of the large bowel. The bundle of tumor have a myxoid central portion with thin fibrous capsule

Gross picture showed a mass located and attached to the outer surface of the transverse colon, 6x6cm in size. Cut section of the mass showed multiple variable size white nodules. The multinodular appearance is typical of neurofibromatosis.

Microscopic picture showed bundles of neural tissue arising from the muscularis propria layer to involve the paracolic area. The bundles of the tumour have amyloid central portion with thin fibrous capsule at the periphery. The cellular element composed of spindle wavy cells with elongated nuclei.

#### Discussion:

Neurofibromas are benign neoplasms consisting of neural and connective tissue components like Schwann cells, peri-neural cells and myofibroblasts.<sup>1,2</sup> Neurofibromas can be solitary, multiple or plexiform.<sup>3</sup> They are usually multiple at presentation.

There are two clinical forms: neurofibromatosis type-1 (von Recklinghausen neurofibromatosis or peripheral neurofibromatosis; NF1) and neurofibromatosis type-2 central neurofibromatosis or bilateral acoustic neurofibromatosis; NF2.<sup>4,5</sup>

The disease has variable clinical manifestations involving the skin, nervous system, eyes, bones, gastrointestinal tract and other body parts. Gastrointestinal tract involvement has been reported in 25% of patients with NF1,<sup>6</sup> with most cases involving the stomach or small bowel.

Neurofibromas exist rarely in the colon as isolated neoplasms<sup>5</sup>. Outside the classical clinical picture of von Recklinghausen's disease was present in this case as shown in figure-1 (neurofibromatosis type-1, NF1)

The pathological forms of gastrointestinal involvement consist of ganglioneuromatosis characterized by hyperplasia and hypertrophy of the nerve plexuses and ganglionic cells in the mucosa.<sup>5,6</sup> Characterized by hyperplasia of neuronal cells in the submucosa, muscularis propria or even from the serosa.

They are benign neoplasms consisting of neural and connective tissue components like Schwann and perineural cells and myofibroblasts. These disease entities have variable clinical expressions with manifestations involving the skin, nervous system, eyes, bones, gastrointestinal tract, and other body parts.

Intestinal neurofibromas are generally submucosal but may extend to the serosa.<sup>6-8</sup> Dense growths known as plexiform neurofibromatosis of the mesentery or retroperitoneal space may lead to arterial compression or nerve injury.

Although there is no specific symptom associated with gastrointestinal neurofibromatosis, clinical manifestations include abdominal pain, constipation, anemia, melena and an abdominal mass. Serious complications including intestinal obstruction, biliary obstruction, ischemic bowel, perforated bowel, intussusception and megacolon have been reported. Solitary neurofibro-

mas of the colon are extremely rare and usually occur in association with NF1.

Solitary neurofibromas have also been reported in other parts of the gastrointestinal tract, such as the mesentery, the ileum, the anal canal,<sup>8</sup> the esophagus, the soft palate, the gallbladder and the common bile duct.<sup>4-6</sup> The clinical significance of isolated neurofibromas has not been well studied. The natural history of these lesions is typically benign, but malignant transformation has been reported, particularly in association with NF1 and in larger, plexiform lesions.

#### **Conclusion:**

Neurofibromas of colon are extremely uncommon. With such low incidence and very few cases reported, long-term follow-up goals and screening are still undetermined at this time, but close clinical follow-up of these patients is important to exclude neurofibromatosis and its associated risk of malignant transformation.

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#### **Role and contribution of authors**

Dr Essam Muhammad Al-Sayed conceived the idea and did the literature search and wrote the article

Dr Dauda Bawa critically review the article and made final changes

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