# Pelvic neurofibroma

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## Abstract

A 58-year old male presented with a hard lump in lower abdomen extending into pelvis. Clinical examination and computed tomography scan revealed pelvic retroperitoneal mass. Patient underwent exploratory laparotomy - revealing a well encapsulated retroperitoneal mass extending up to coccyx, posterior to the urinary bladder. It had displaced the recto-sigmoid colon and urinary bladder to the right. Histopathological findings revealed a neurofibroma. Pelvic neurofibromas are known to arise from certain pelvic organ. However, present case ruled out any organ involvement both on imaging and intraoperatively. This shows that these tumors can arise *de novo* also. A brief case report and review of literature is presented.

## Introduction

Neurofibroma is a benign soft tissue tumor. It presents in various anatomic locations. However it is an uncommon pelvic retroperitoneal tumor. Only 60 cases of pelvic neurofibroma have been reported till date.[1,2] We report this case of a pelvic neurofibroma in a 58-year old male, which mimicked a soft tissue tumor on presentation.

## Case Report

A 58-year old male presented with a lump in lower abdomen associated with diffuse dull aching pain in abdomen. Patient had no urinary or bowel complaints. Clinically, a hard lump was palpable in hypogastric region measuring 20×7 cm, extending from the left iliac fossa into the pelvis. Lump was retroperitoneal with limited mobility. Patient had no pallor or edema. Per rectal examination did not reveal any abnormality. Ultrasonography and computed tomography (CT) scan revealed a large heterogenous mass in the abdomino-pelvic region posterior to the urinary bladder, pushing the bladder and recto-sigmoid to the right (Figure 1). An image-guided biopsy was inconclusive. A provisional diagnosis of soft tissue sarcoma was made. Patient underwent an exploratory laparotomy through a lower midline incision.

Intraoperative findings revealed a retroperitoneal mass, displacing the urinary bladder and recto-sigmoid colon to the right side. The peritoneum was opened over the mass. The left ureter was dissected and separated from the mass. The mass was well encapsulated and firm. The plane between the mass and urinary bladder and rectum was well maintained. The mass reached up to the coccyx posteriorly. Lump was devascularized and removed en masse. The total operating time was 120 minutes and there was minimal blood loss. Postoperative period was uneventful.

Histopathology revealed a tumor measuring 26×20×12 cm in size, encapsulated and smooth surfaced. On cut section (Figure 2), it was yellowish with a small area of necrosis. On microscopic examination, there were spindle cells in a whorled pattern with dense collagenized connective tissue at the periphery (Figure 3). Immunohistochemistry showed S 100 positivity and was negative for smooth muscle actin and activin receptor like kinase. This confirmed the diagnosis of neurofibroma.

Follow up after 1 year has shown patient to be symptom and disease free.

## Discussion

Neurofibroma is a benign soft tissue tumor arising from Schwann cells. There are 3 types of neurofibroma: cutaneous, spinal and plexiform. Plexiform type has a tendency to turn malignant. Neurofibromas have been classically associated with neurofibromatosis type 1. They are found in various anatomical locations but seldom in the retroperitoneal location in pelvis. While ultrasonography, CT scan helps in detecting the extension of tumors in the pelvic region, only a definite tissue diagnosis can rule out malignancy in a retroperitoneal mass.

In the present case, the mass was pressing on the bladder and stretching the left ureter. Patients with neurofibroma usually don't have any neural symptoms. As a matter of fact, only histology can diagnose them and, if it is per-

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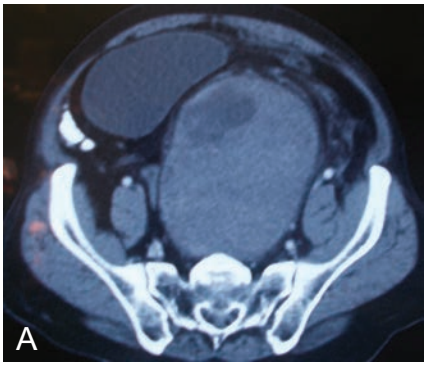


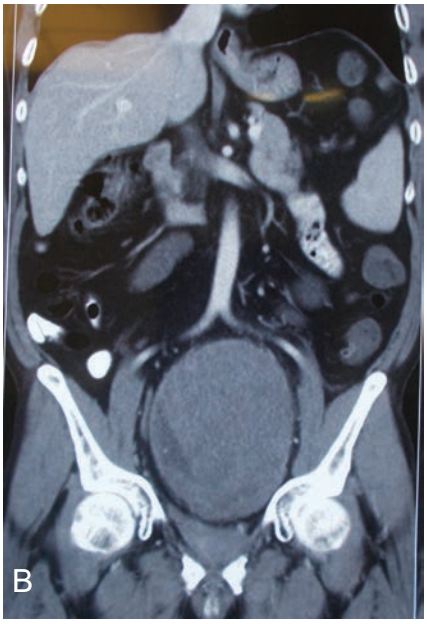


**Figure 1. Computed tomography A) axial and B) sagittal view showing large heterogenous mass in the abdomino-pelvic region posterior to the urinary bladder, pushing the bladder to the right.**

formed preoperatively, it can influence treatment. Most cases reported till date have been associated with a pelvic organ such as urinary bladder, prostate, the bones (sacrum, coccyx), psoas muscle, etc. In present case, there was no particular organ association identifiable.

Paul *et al*.[3] presented a case of a female with chronic pelvic pain, found to have a sacrococcygeal neurofibroma. Here it shows a presentation with neural pain. Manish *et al.*[1] presented a case of a 12 year old male child with urinary obstruction found to have a prostatic neurofibroma. Nadkarni *et al.*[4] presented case of a 4-year old boy with low-back pain that radiated bilaterally into the L-4 and L-5 dermatomes; intraoperatively, a solid mass, which engulfed the entire cauda equina, could not be dissected from the roots. It was a case of a plexiform neurofibroma. In present case, patient had no specific symptoms.

Wei *et al*.[5] presented a case of plexiform neurofibroma affecting the uterine cervix in a patient with chronic pelvic pain and menorrhagia who had multiple cutaneous neurofibromas and one large paraspinal neurofibroma; patient had von Recklinghausen's disease. Mimata *et al.*[6] presented a case of malignant neurofibroma in urinary bladder without associated von Recklinghausen's disease.

Most patients found to have retroperitoneal neurofibroma have associated neurofibromatosis/von Recklinghausen's disease. In present case, we could not point to a certain organ of origin in the pelvis and the patient did not have any hereditary disorder. It is important to discriminate between malignant and benign lesions thus modifying a surgical approach, conservative *versus* aggressive ones. Present case had no cutaneous neurofibromatomas, hence diagnosis of pelvic neurofibroma was established only on histopathology of the excised specimen.

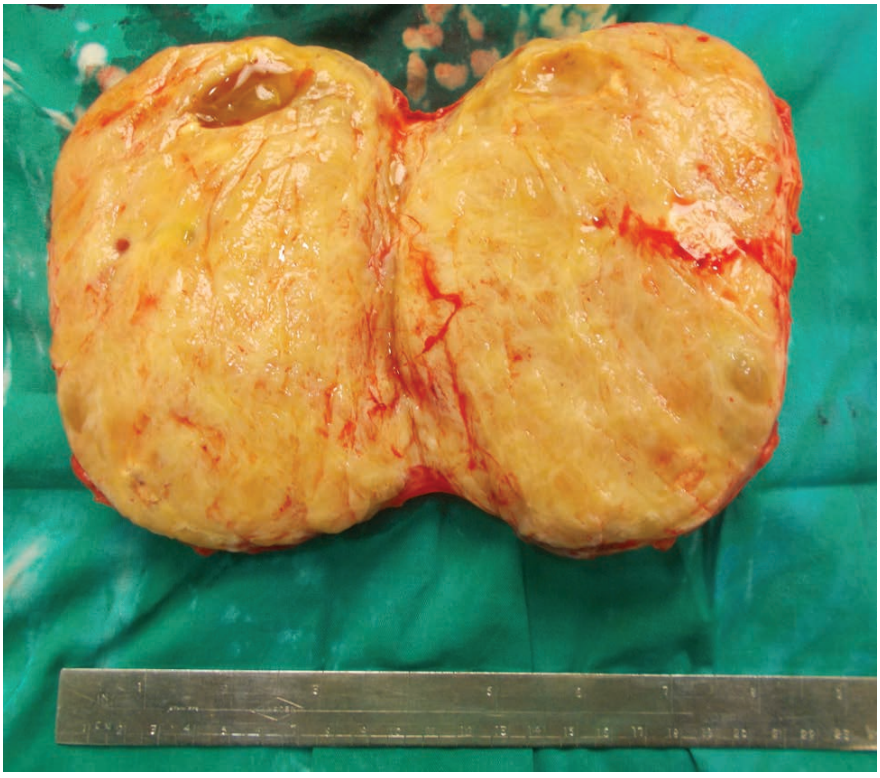

**Figure 2. Cut section of excised tumor.**

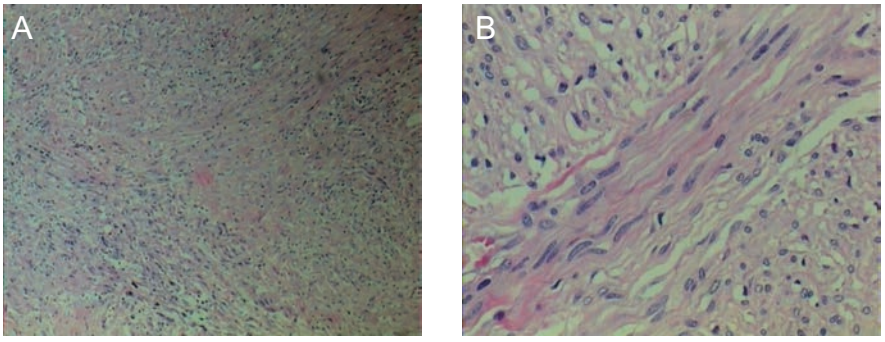


**Figure 3. Histopathology pictures (A: 10x and B: 40x) of neurofibroma showing spindle cells in a whorled pattern with dense collagenized connective tissue at the periphery (hematoxylin and eosin stain).**

## Conclusions

Pelvic neurofibromas are very rare and are usually found arising from one of the pelvic organs. Patients with retroperitoneal schwannoma or neurofibroma usually have neurofibromatosis/von Recklinghausen's disease. Present case of pelvic retroperitoneal neurofibroma shows that these tumors can arise *de novo* with neither any genetic disorder nor any particular organ involvement. Hence, more studies are required to study the *de novo* appearance of such tumor.

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