# A Huge Retroperitoneal Neurofibroma mimicking broad ligament fibroid in postmenopausal women: An extremely rare report with review of literature

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**ABSTRACT**

The neurofibroma is a tumour of neural origin. This kind of neoplasm, though, is generally skin located. Rare cases in deep organs or in the peritoneal cavity are also reported in the literature. There are two types of neurofibromas, localized and diffuse; the latter is associated with von Recklinghausen disease and always occurs together with skin neurofibromas. Here we report the case of a 62-year-old woman affected by neurofibroma, but not associated with von Recklinghausen disease.

A computed tomography (CT) scan described a retroperitoneal pararenal lesion with no clear involvement of adjacent viscera. We describe the diagnostic modality, treatment planning and the timing of treatment of this neoplasm, reviewing also the literature.

**Keywords:** benign tumour of kidney, neurofibroma, rare tumour, retroperitoneal tumour

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## INTRODUCTION

Neurofibromas, in general, are rare neoplasm and arise in patients with von Recklinghausen disease, but a solitary variant has been observed in rare cases and its splanchnic location is very uncommon [1].

Paraaortic–Pararenal Neurofibroma is an exceedingly rare tumour location [2]. To our knowledge, only six such cases have been reported worldwide to date . Although their diagnosis and location is similar to that of Neurofibroma, different diagnostic and therapeutic approaches (surgery) have been

used. In all cases, except ours and another one, patients underwent radical nephrectomy. In all likelihood, such an approach is due to the fact that preoperative imaging staging does not often allow one to diagnose these neoplasias.

As a matter of fact, only histology can diagnose them and, if it is performed preoperatively, it can influence treatment. In particular, it is capital to discriminate between malignant and benign lesions thus modifying a surgical approach, conservative versus aggressive ones. In our case, the absence of mitotic activity, lack of necrosis, pleomorphism and infiltrative pattern of growth allow us exclude malignity.

**CASE REPORT**

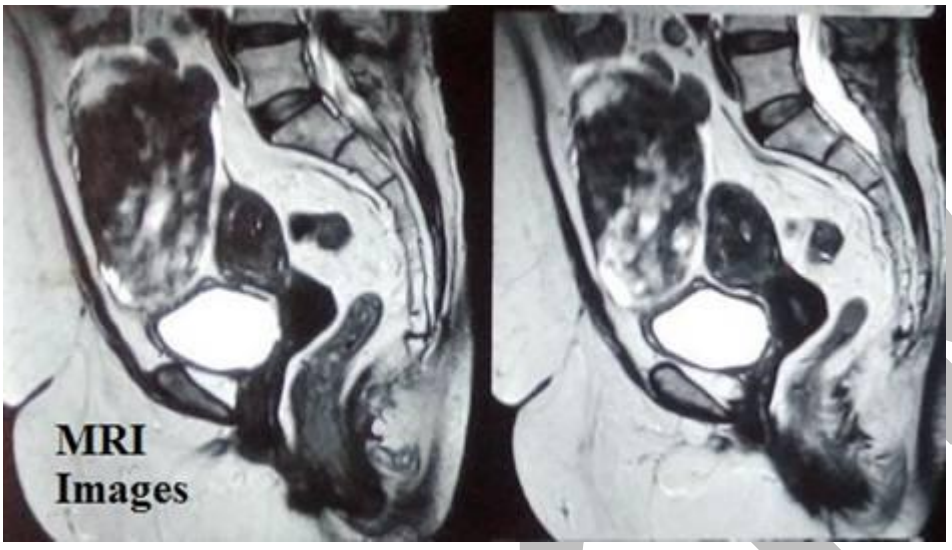
A 62 yr old female with para 3 living 3 tubal ligation done 33 yrs came with c/o abdominal discomfort nausea, anorexia since 1-2 months. Patient is postmenopausal since 15 years and had history of 1 lower segment caeserian section and 2 vaginal birth after caeserian section and last delivery was 33 years back.

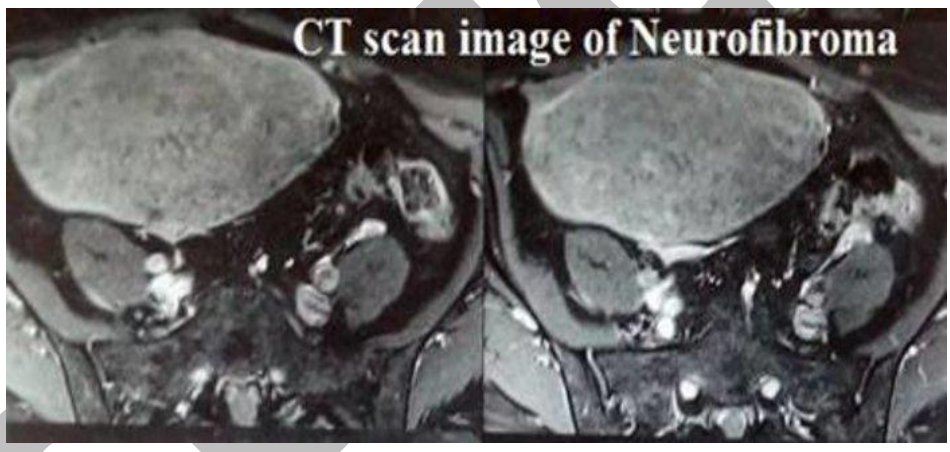
Patient is a known case of hypertension and diabetis mellitus and taking medicines since last 6-7 yrs.her vitals were stable.her systemic examination revealed nothing abnormal. On per Abdominal examination there was a huge mass of approximately of 16cm*9cm*8cm felt with diffuse margins and hard in consistency which was extending from the right sided of iliac region upwards.mobility restricted.

Scar of **LSCS** was healthy. there was no guarding , tenderness or rigidity , on per speculum examination the cervix was flushed with vagina. On per vaginal examination the same size mass was felt on per vaginal examination and was very high up with restricted mobility and hard in consistency.patient's tumour markers were within normal range like AFP - 2.38 iu(0.5-5.5),beta HCG 4.53 (0-5.3),ca 125-12.7 u (0-35).

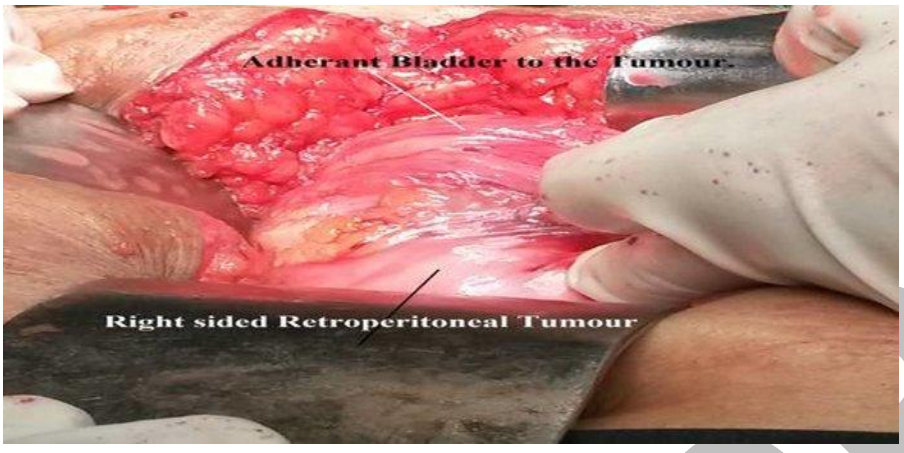
Ultrasound examination suggestive of right adnexal mass of size 6.5 cm x 10.2 cm which appears to be subserosal fundal fibroid or right ovarian mass .magnetic resonance imaging studies showed it as broad ligament fibroid of size 12.5cm x 6.7 cm separate from right ovary well preserved surrounding fat planes.it also showed few small to prominent heterogeneously enhancing lymph nodes at the level of aortic bifurcation which needed further

evaluation.patient undergone (Figure 1), (Figure 2).

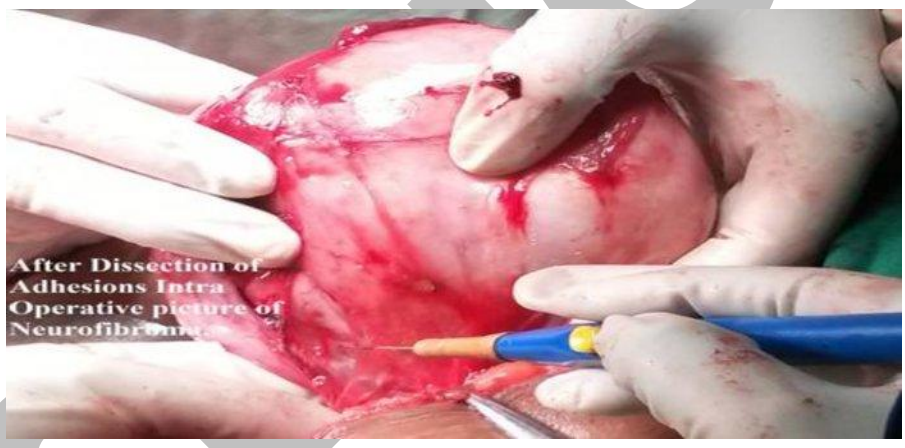


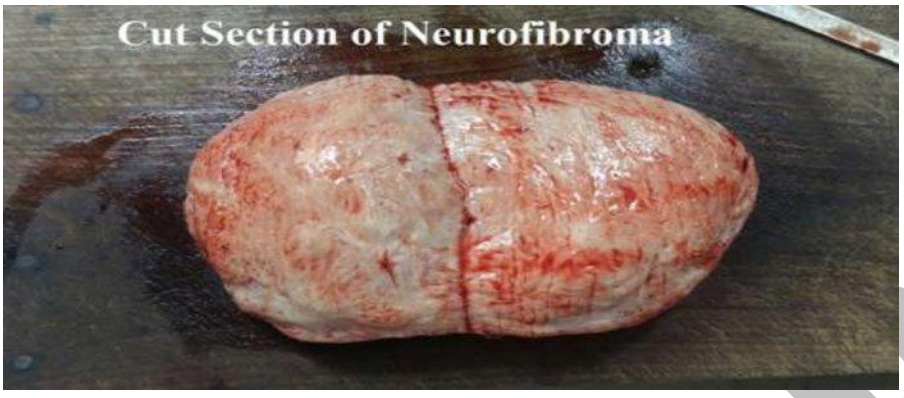

Laparotomy in situ patient had dense multiple adhesions between anterior abdominal wall and parietal peritoneum.whole things were stucked up.bowels was adherent which was separated with fine dissection.the bladder was very much stucked and adherent to the mass arising from retroperitoneum.uterus and adnexae were not seen due to the mass and adhesions.(Figure 3)

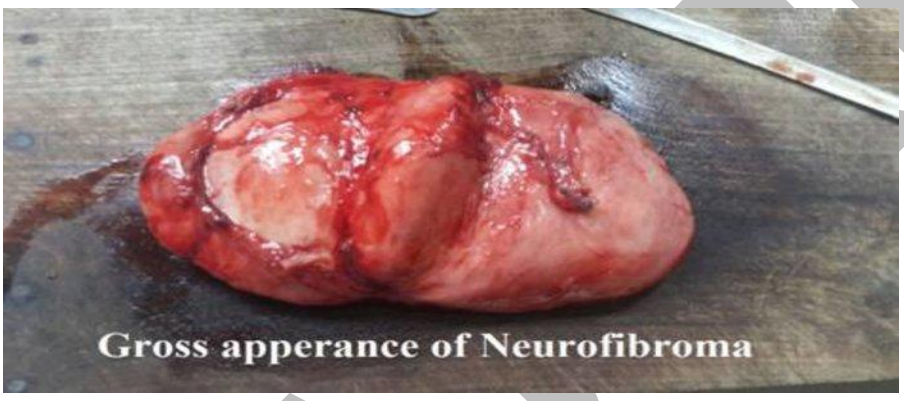



Adhesions were separated by blunt and sharp dissection and adnexal mass isolated .Grossly the mass was a size of 17cm*10cm*8cm roughly.hard in consistency,whitish in colour with smooth and shiny surface. After dissecting and separating the tumour mass sent to frozen section (Figure 4).




The frozen section report was suggestive of spindle cell tumour most likely neurofibroma. after the removal of mass then the atrophic uterus and ovaries seen clearly.Total abdominal hysterectomy with bilateral salpingoophorectmy done.postoperatively patient was managed on antibiotics and analgesics with injection insulin for diabetic management .Patient recoverd well with suture removal done on day 10 .Final histopathology report was suggestive of neurofibroma.(Figure 5) ( Figure 6)




**DISCUSSION**

First described by Garre et al [2]. in 1892, glandular differentiation is the rarest form of divergent differentiation seen in peripheral nerve sheath tumors (PNST) which also includes cartilage, bone, chondrosarcoma, osteosarcoma and rhabdomyosarcoma. Around 40 cases have been reported so far in world literature.

Diagnostic criteria includes evidence supporting a finding of nerve sheath tumor, presence of true glandular epithelium, not entrapped glands or pseudoepithelium. The vast majority of PNST harbouring these glands have been malignant peripheral nerve sheath tumor (MPNST) [3]. The glandular epithelium is usually benign though there are sporadic case reports of PNST with malignant glandular component [4].

Glandular neurofibromas are rare and in an extensive review by Woodruff and Christenstein, only 2 out of 25 analyzable cases of glandular peripheral nerve sheath tumors were neurofibromas. Both of these cases occurred in young females, were associated with NF Type 1 and were finally

diagnosed as plexiform neurofibromas [3]. No history of neurofibromatosis was present in the other reported cases of glandular neurofibromas .

Most important differential diagnostic consideration in a case of glandular neurofibroma is schwannoma with entrapped adnexal structures. Schwannomas usually display typical Antoni A and B areas along with presence of Verocay bodies. Trapped adnexal glands are generally seen in clusters and are connected to each other.

In the absence of typical morphological features, differentiation between these entities may be difficult and immunohistochemistry (IHC) may be employed. Glands in true glandular PNST are devoid of myoepithelial cell lining and are hence non reactive for muscle common actin (HHF-35). This glandular epithelium is not only reactive for cytokeratins but also for neuroendocrine cell markers (chromogranin, serotonin and somatostatin). Entrapped adnexal glands in schwannoma show the presence of myoepithelial cell layer and are non reactive for neural markers. Stromal cells show S-100 positivity in both the cases [3].

Though IHC was not employed in our case (case number 1), the tumor was diagnosed as neurofibroma as characteristic features of neurofibroma (spindle cells with short, curved nuclei embedded in a collagenous stroma) were seen on histology sections. Moreover, history of NF-type 1 in the patient corroborated the diagnosis of neurofibroma. The glands seen in case number 1 resembled mucus secreting glands rather than eccrine glands of skin adnexa.

It is interesting to note that adnexal glands have also been reported in neurofibroma and it has been proposed that the tumor appeared in the nerves around the eccrine glands and grown to the subcutaneous tissue, and the glands might have been left behind rather than entrapped by the growing tumor.

The histogenesis of glandular PNST is still not clear and it may be attributed to the metaplastic potential of Schwann cells or the presence of primitive neural crest cells that migrate with Schwann cells along the peripheral nerves [4].

Neurofibroma with rosette like structures is exceedingly rare. Enzinger reviewed a unique case of neurofibroma showing presence of mucus secreting glands and focal rosettes . A recently described

variant of neurofibroma with rosettes is dendritic cell neurofibroma with pseudorosettes (DCNP) . This tumor is seen in adults and has been described mostly in patients without a history of NF-1,. though cases arising in association with NF-1 have also been described. The lesion is well circumscribed and occurs in superficial dermis of head, trunk and extremities. It is comprised of two types of cells. Type 1 cells are small lymphocyte like cells with slightly cleaved nuclei which are concentrically arranged around larger type 2 cells having vesicular nuclei and copious cytoplasm. These type 2 cells have dendritic extensions which form the core of these pseudorosettes. On IHC, type 2 cells and most type 1 cells stain for CD57 and S-100 .

Another important differential diagnostic consideration is Schwannoma with neuroblastoma like rosettes. In this tumor, small round to oval cells are layered around a central eosinophilic fibrillary material. These cells may also show presence of intranuclear cytoplasmic inclusions.

## CONCLUSION

Even though neurofibroma are rare in postmenopausal age group it should be considered as one of the differential diagnosis in case of solid ovarian mass. Preoperative suspicion of retroperitoneal neurofibroma help in preventing intraoparative complications. Our case report also emphasizes that preoperative thorough investigations like CT scan, MRI are necessary in case of mass of uncertain origin.

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