International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Case of Isolated, Giant Pelvic Neurofibroma

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Abstract: ***A 62-year old male presented with a slow-growing, right buttock swelling, with both constipation and incontinence, the latter related to surgery performed by a quack, for anal fistula. It was elastic in feel, and per rectal examination revealed extrinsic compression of anal canal. CT and MRI scans revealed a large, dumb-bell shaped, well-encapsulated pelvic mass with perianal extension; FNAC and Core-needle biopsy were inconclusive. Patient underwent perineal exploration, to yield a large, well-encapsulated mass, with no apparent major pedicles or attachments; patient recovered with no additional morbidity. Histo-pathological examination revealed a neurofibroma. Reports of pelvic neurofibromas are uncommon, though these can arise from any pelvic organ, usually in association with neurofibromatosis. Our case was atypical, in having no apparent origin from any major organ, and bearing no other stigma of neurofibromatosis.***

Keywords: CT- Computed tomography, MRI - Magnetic resonance imaging, FNAC- Fine needle aspiration cytology

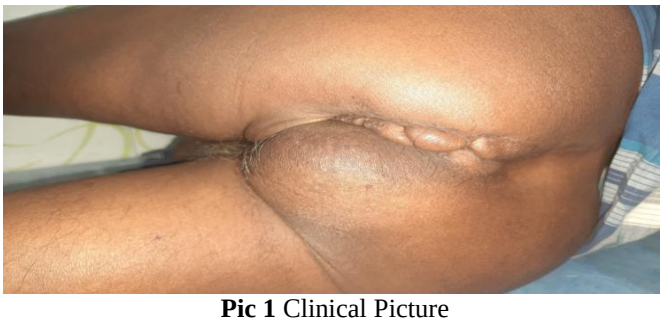
1. Introduction

Neurofibromas are benign(WHO grade 1) peripheral nerve sheath tumour with classic identifiable features including the presence of a neuronal component comprising of transformed Schwann cells and a non-neoplastic fibrous component that includes fibroblasts.

These are usually solitary and sporadic. There is, however, a strong association with neurofibromatosis type 1 (NF1), particularly for the plexiform subtype.

2. Case Presentation

Our patient, a 62–year-old male villager, presented with a right buttock swelling (pic-1) of 2 years duration. Initially small to start with, it was gradually progressive and examination revealed a globular swelling of approximately 8cm x12cm size. There was no pulsation, overlying venous prominence or pigmentation and the surface was smooth. The margins were well-defined and the feel somewhat cystic/elastic. Clear-cut fluctuation could not be demonstrated, but the patient had expansile impulse on cough over the swelling, and yet, it was not significantly reducible on pressure. Inguinal, abdominal and neck nodes were not palpable.

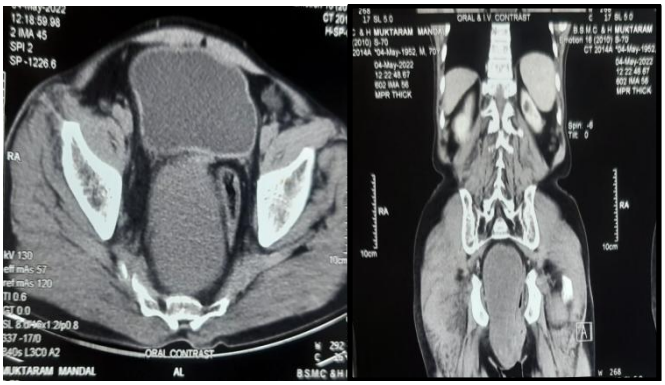


Pic 1 Clinical Picture

Patient also complained of constipation along with faecal incontinence which started after previous, multiple, perianal operations for perianal abscess and fistula (performed by a quack, as per the patient). As a result, the patient also had some other small swellings around the anal verge, but these were more irregularities of the perianal contour due to previous scarring, rather than actual lumps. On per rectal examination, a globular mass was palpable on the right side of the anal canal compressing the lower part of rectum and anal canal. The patient had no other significant local complaint, such as per rectal bleeding or pain, nor systemic symptoms such as, asthenia, anorexia, weight loss, haemoptysis, bone pain, jaundice, suggestive of distant disease.

Ultrasonography revealed a hypoechoic SOL with minimal internal vascularity. CECT (Pic 2a & b) and MRI (Pic 3a & b)displayed a large, elongated, heterogeneously enhancing, well- defined, solid SOL in the pelvis, posterior to the urinary

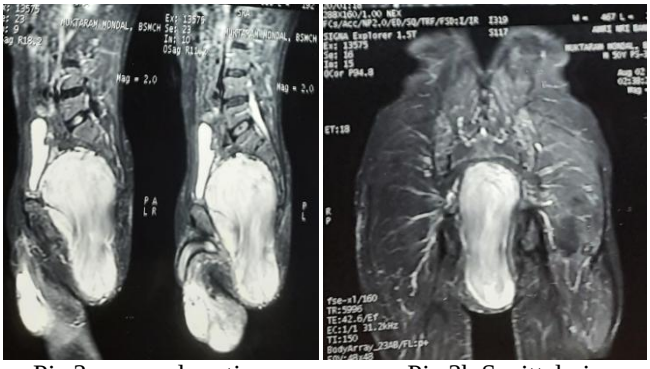
bladder displacing it anteriorly. The swelling was compressing the rectum and extended inferiorly in to the medial aspect of right gluteal region, up to the subcutaneous plane. The MRI report suggested a GIST lesion but neither FNAC, nor core-needle biopsy, yielded any conclusive results.



2a. Axial cut 2b. Sagittal view

Pic 2: CECT Scan plate

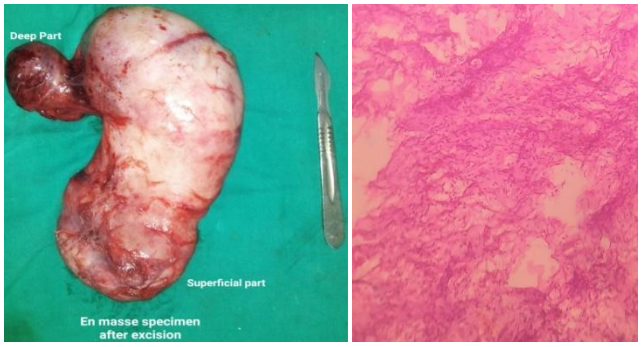
Under general anaesthesia and in lithotomy position, a curvilinear incision of around 7 cm was made 5 Cm from the anal verge. By a combination of blunt & sharp dissection done an approximately 20cm x 8cm large, dumb-bell shaped, well-defined mass (Pic 4), with no apparent major pedicles or attachments was delivered and excised through the perineum. Absorbable gelatinous sponges were placed in the cavity and a suction drain placed. Recovery was uneventful.



Pic 3a coronal section. Pic 3b Sagittal view

Pic 3: MRI plate

On HPE(Pic 5), the un-encapsulated tumour mass was found to be composed of loosely arranged, benign-looking, spindle cells (with increased vascularity) and moderate amounts of intervening strands of collagen, arranged in bundles (shredded-carrot like). The individual spindle-shaped cells had wavy, serpentine nuclei, with pointed ends and indistinct pale eosinophilic cytoplasm. The background had moderate amount of myxoid and collagenous stroma and scattered mast cells. Mitotic figures were not seen. All these suggested a benign Neurofibroma.



(Pic 4: En masse specimen) **(Pic 5:** HPE slide)

3. Discussion

Neurofibromas originate from Schwann cells and are a benign soft tissue tumour which can occur in any region/organ, although they are predominantly found in the skin. Solitary neurofibromas are relatively common. However it is an uncommon pelvic tumor; only 60 cases of pelvic neurofibroma have been reported till date [1,2].

There are 3 types of neurofibroma: cutaneous, intraneural and plexiform. The plexiform type has a tendency to turn malignant (2-3%). Neurofibromas have been classically associated with neurofibromatosis type 1. They are found in various anatomical locations (most commonly in skin) but, as mentioned, seldom in the retroperitoneum, in the pelvis. Murphey et al. reported that localized intraneural neurofibromas, are by far, the most common form of neurofibroma, representing 90% of these lesions. [3] 90% of the solitary neurofibromas are sporadic and 10% are inherited. Incidence of inherited neurofibromas is 1 in 2,600 to 3,000 individuals. Superficial neurofibromas are more common than deep neurofibromas [4].

While ultrasonography and CT scan help in detecting the extension of tumors in the pelvic region, only a definite tissue diagnosis can specify the nature and rule out malignancy in such masses. However, certain characteristic imaging features like the 'target sign' on T2W MRI are highly suggestive of the diagnosis, so MRI may be valuable tool in initial and follow-up evaluation of these patients [2].

Patients with neurofibroma usually don't have any neural symptoms and diagnosis is often clinical. Though preoperative tissue diagnosis can influence treatment, in our case repeated preoperative attempts at tissue diagnosis were inconclusive and we had to depend on excision biopsy. Most cases reported till date, have been of lesions associated with a pelvic organ such as urinary bladder, prostate, the bones (sacrum, coccyx), psoas muscle, etc. In this case, no attachment/origin, to/from any particular organ, could be identified.

Though, in our case, the patient didn't have any specific symptoms, Paul et al, in their study, presented a case of a female with chronic pelvic pain, due to a sacrococcygeal neurofibroma.[5] Manish et al, presented a case of a 12-year-old boy with urinary obstruction due to a prostatic

neurofibroma[1] and Nadkarni et al, presented case of a 4-year old boy with low-back pain that radiated bilaterally into the L-4 and L-5 dermatomes, caused by a plexiform neurofibroma.[6]

4. Conclusion

To conclude, it would be very difficult to diagnose a pelvic mass as a neurofibroma, but it may happen, rarely. These lesions when they occur usually arise from one of the organs locally, but rarely, as in our case, they may arise independently.

References

[1] Jana M, Gamanagatti S, Kumar R, Aggarwala S. Pelvic neurofibroma arising from prostate in a case of neurofibromatosis-1. Indian J Urol 2011;27:415-7
[2] Niku SD, Mattrey RF, Kalota SJ, Schmidt JD. MRI of pelvic neurofibromatosis. Abdom Imaging. 1995 Mar-Apr;20(2):176-8.
[3] Murphey MD, Smith WS, Smith SE et-al. From the archives of the AFIP. Imaging of musculoskeletal neurogenic tumors: radiologic-pathologic correlation. Radiographics. 19 (5): 1253-80. Radiographics.
[4] Abdellatif E, Kamel D. Neurofibroma-general. PathologyOutlines.com website. https://www.pathologyoutlines.com/topic/softtissueneurofibroma.html. Accessed November 8th, 2022
[5] Paul PG, Pravinkumar T, Sheetal B. Sacrococcygeal neurofibroma: rare cause for chronic pelvic pain. J Minim Invasive Gynecol 2012;19:517-20. [PubMed] [Google Scholar]
[6] Nadkarni TD, Rekate HL, Coons SW. Plexiform neurofibroma of the cauda equina Case report. J Neurosurg 1999; 91:112-5. [PubMed] [Google Scholar]