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Solitary giant neurofibroma as a retroperitoneal mass: A rare case

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Abstract
The autosomal dominant condition known as neurofibromatosis type 1 is typified by neurofibromas tumors, which are renowned for being many cutaneous masses with a strong likelihood of becoming malignant. On the other hand, benign solitary retroperitoneal neurofibromas (SRN) are rare and manifest alone, without coexisting medical conditions.
This 41-year-old woman complained of a painless lump in the left side of her lower abdomen that had been there for a month when she arrived at the general surgery outpatient department. A solid, non-tender, palpable oval shaped lesion measuring 10 x 7 cm with restricted movements was noted in the left lumbar region. The ultrasonography showed a well-defined hyperechoic lesion with anechoic areas measuring 9.7 X 7.2 cm in the left iliac fossa with internal vascularity.
According to an MRI, there is a large lobulated retroperitoneal mass lesion on the left side at the level of L3 L4 L5 vertebral body levels in the paravertebral area. USG-guided biopsy showed Spindle cells with a plump, buckling nucleus, minimal pleomorphic, and mitotic activity (S100 - > 80% positive) comprise the tumour tissue of a neurofibroma. This retroperitoneal neurofibromas is excised via a conventional open surgery. This case reports presents a rare case of giant neurofibroma presenting as a retroperitoneal tumour, a rare incidence.

Keywords: Giant neurofibroma, retroperitoneal mass, excision.

Introduction
Soft tissue neoplasms known as Nerve Sheath Tumors (NSTs) include benign and malignant schwannomas as well as neurofibromas. They comprise 4% of retroperitoneally situated malignancies [1]. Neurofibromas, also known as peripheral nerve sheath tumors, are benign tumors that grow slowly and are usually non-encapsulated. They are more common in the third and sixth decades of life and equally common in both men and women [2]. One of the numerous diagnostic signs of neurofibromatosis-type 1 (NF 1), an autosomal dominant neurocutaneous condition, is the existence of multiple cutaneous neurofibromas which have a higher chance of turning into malignant peripheral nerve sheath tumors (MPNST) [3].
Neurofibromas that develop in the retroperitoneal space extra-intestinally and manifesting as a solitary benign retroperitoneal tumour devoid of concurrent abnormalities are extremely rare [4]. After taking into account the previously described factors, it can be said that a giant Solitary Retroperitoneal Neurofibroma (SRN) is a very uncommon diagnosis, as shown by the incredibly tiny number of case reports that have been published.

Case Study
The main complaint of a 41-year-old woman was a lump in the left side of her lower abdomen that she had noticed a month prior. The lower abdominal lump was becoming less noticeable in sitting position. It was gradually increasing in size. Not associated with fever, vomiting, loose stools, blood or mucus in the stools, loss of appetite, weight loss, perspiration, trembling, joint pains, or trauma. On physical examination, a single 15 x 10 cm, vertically oval, non-tender, firm swelling with a smooth surface, well-defined edges and not ballot able, was found to be extending over the left iliac and lumbar regions. During the leg rising test, the swelling becomes less prominent, and there were no expansive or transmitted pulsations detected over the swelling.

In the left iliac fossa, ultrasonography revealed a well-defined heteroechoic lesion with anechoic regions of 15 X 7.2 cm and internal vascularity. According to an MRI, there is a large lobulated retroperitoneal mass lesion on the left side at the level of L3 L4 L5 vertebral body levels in the paravertebral area. CT scan showed a well-defined soft tissue density lesion measuring 9.5 (SI) x 8.1 (T) x 9.1 (AP) cm noted in the left lumbar and iliac regions causing displacement of adjacent vessels and bowel loops with heterogeneous enhancement on post contrast study (Figure-1). The tumor along with capsule were completely removed, when the surrounding nerve sheath was accessed (Figure-2). The removed tumor underwent a histological analysis across several cross-sections, which showed a well-encapsulated tumor with spindle cells with buckling nuclei grouped in fascicles and storiform pattern. Certain specimens have cytoplasmic vacuoles, localized hyaline, and myxoid degeneration, along with small, round, blue cells that are richly vascularized and have low mitotic activity, all of which are indicative of neurofibroma (Figure-3). Consequently, this study has verified the diagnosis of neurofibroma. Given the close correlation between neurofibromas and NF1, a thorough medical history and physical examination were conducted to rule out both NF1 and NF2, and no recurrent symptoms were observed throughout the follow-up period.

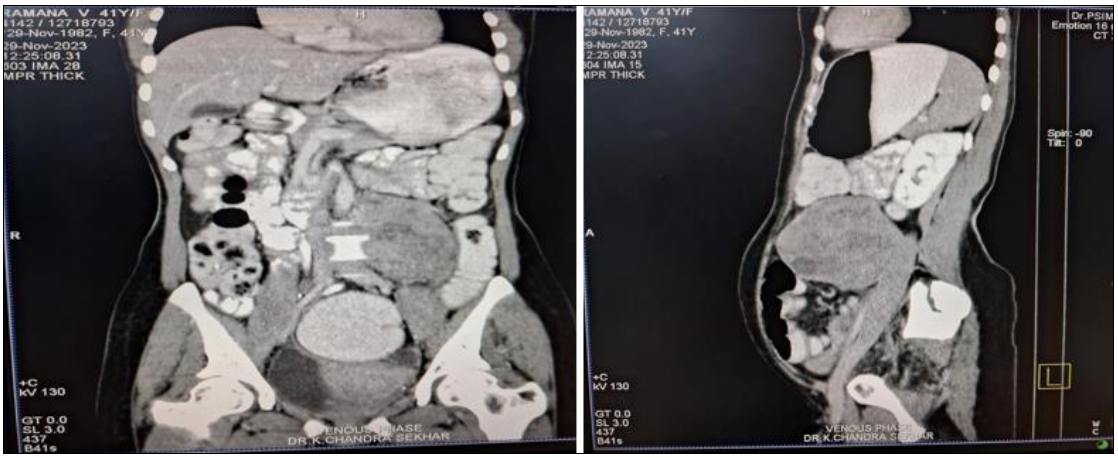

Fig 1: A radiological image of coronal and Sagittal CT shows well defined soft tissue density lesion measuring 9.5 (SI) x 8.1 (T) x 9.1 (AP) cm noted in the left lumbar and iliac regions causing displacement of adjacent vessels and bowel loops with heterogeneous enhancement on post contrast study.

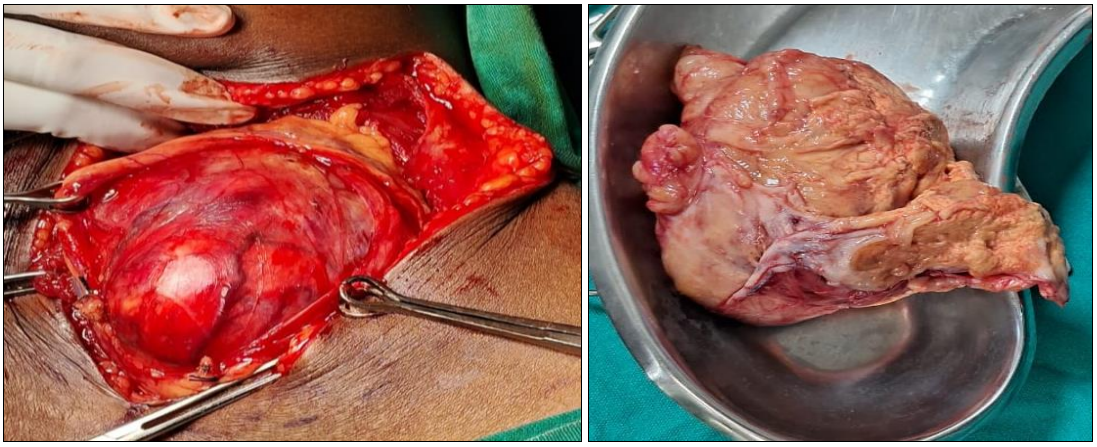

Fig 2: Gross image of well circumscribed tumor of size 8.5cm*6cm*5.5cm with firm consistency. Image was taken after surgical excision.

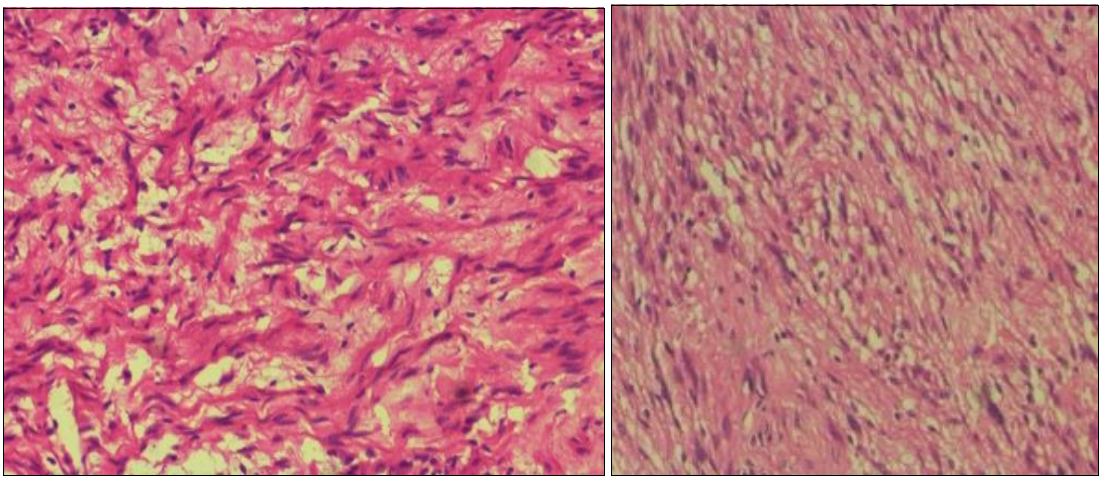

Fig 3: Histopathology across multiple cross-sections revealing a partially

Encapsulated lesion composed of a monotonous population of spindle-shaped Cells with wavy nuclei.

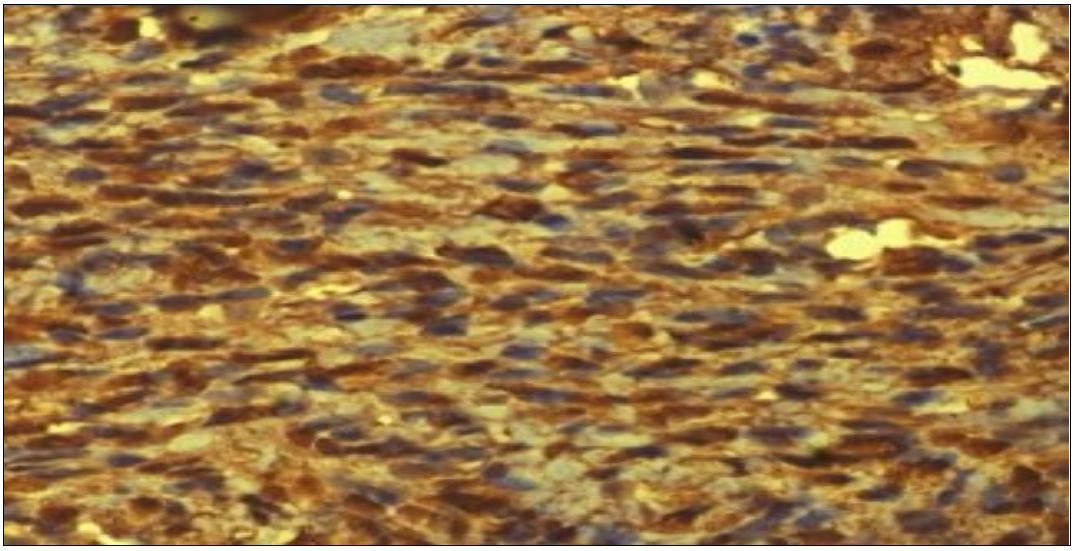

Fig 4: Histopathology with SOX10 Immunostaining showing nuclear staining in tumor cells

Discussion

A broad category encompassing over 60 neoplasms, soft tissue sarcomas (STS) can originate from almost any anatomic location and impact both the old and the very young. Skeletal muscle, adipose cells, blood and lymphatic arteries, connective tissue cells with a shared mesoderm origin and peripheral nerves coming from the neuroectoderm are among the tissue types of STS origin [5]. Multiple tumors can develop in the brain, spinal cord, and peripheral nerves as a result of the hereditary condition neurofibromatosis [6]. There are three varieties: NF1, NF2, and SWN (schwannomatosis). The NF1 gene, which is found at chromosome 17q11.2, codes for a protein termed neurofibromas, which functions as a tumor suppressor of the RAS oncogene signaling pathway. Mutations of this gene can result in neurofibroma, an autosomal dominant disorder [7]. These patients have a 10% chance of developing a malignant peripheral nerve sheath tumor in addition to the common development of numerous cutaneous neurofibromas. The final diagnosis made for our patient was a gigantic solitary neurofibroma in the retroperitoneum because she did not exhibit the usual signs and symptoms of neurofibromatosis. The literature reports that the most common locations for solitary neurofibromas are the limbs, trunk skin, mouth region, and infrequently the retroperitoneum. The traditional signs of isolated neurofibromas are not clearly defined and change depending on where they occur [8]. The majority are found when they impinge on nearby structures, which can cause extra luminal pressure to be exerted, palpable masses, and colicky stomach pain [8]. The typical tumor size at presentation is 15 cm, which is likely explained by the retro peritoneum's isolated location. Due to their great spatial resolution and repeatable axial image capture, CT and MRI are frequently utilized for the examination of soft tissue and retroperitoneal tumors. Rapid picture acquisition, almost ubiquitous availability, and a condensed data set that can be easier for non-radiologists to interpret are some of the benefits of CT scanning. Wider soft tissue distinction is one of MRI's benefits, nevertheless, compared to CT scan, MRI has more implant-related contraindications, is more limited in availability, and is more susceptible to claustrophobia and motion artifact. However, preoperative imaging is usually not enough to provide a definitive diagnosis, and a biopsy is usually required for histopathological confirmation [9]. Surgical management of several retroperitoneal neurofibromas has been described to be challenging due to their large size, extensive infiltration, and abundant vascularity [9]. On rare occasions, certain surgeons decide to operate only when the tumor has very malignant potential, is causing discomfort, or exhibits neurological symptoms. Nonetheless, any undetected mass left in the body for an extended period might have negative implications, so the primary course of treatment for a big retroperitoneal neurofibroma should always be surgical excision. During the follow-up phase, our patient showed no evidence of recurrence and recovered well from the procedure.

Conclusion

There isn't a lot of material since isolated retroperitoneal large neurofibromas are uncommon in the clinical context. They are therefore frequently missed or challenging to spot. A multidisciplinary healthcare team's cooperative efforts are necessary for the proper diagnosis and treatment of neurofibromas. In this uncommon instance, a 41-year-old patient with a single retroperitoneal large neurofibroma has been surgically treated without any additional symptoms or neurological impairment. When dealing with retroperitoneal tumors, particularly giant neurofibromas, a thorough understanding of the surgical anatomy of the area is crucial for dissection in these cases. Meticulous surgical exercise is also required to minimize surgical complications due to the nearby neurovascular and visceral structures. When performing surgery on a retroperitoneal tumor, the surgeon needs to be ready and aware of potential surprises.

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