A Giant Schwannoma camouflages as a Renal mass – A Case report

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Abstract

Schwannomas rarely occur as neurogenic tumors develop due to nerve Schwann cells. A thirty-five-year-old female reported to our department complaining of Urinary Tract infection (UTI) due to a renal mass. Our radiological investigations indicated a Giant Schwannoma (GS), which emerged from the right L2 nerve root of 7.4x5.5x9cm in size. The tumor was seen extending from L1 to L4 and pressing on the kidney and ureter. The tumor excision was performed using the posterior and retroperitoneal approaches. It was diagnosed as Schwannoma histopathologically. No postoperative complications or recurrence was observed at follow-up of 6 months.

Keywords: Schwannoma, misdiagnosis, Renal mass, neurilemmoma

Introduction

Giant schwannomas (GS) are tumors developing through schwann cells of the peripheral nerve sheaths that affect the head, neck, limb flexor surfaces, etc. In 1908, Verocay Jose was the first to express them, 12 years later, Antoni separated them into two distinct histologic patterns [1]. They affect individuals of all ages, though they are most common in females between the ages of 20 and 60 [2]. Schwannoma in the retroperitoneal cavity is uncommon, accounting for only 1–3% of all schwannomas [3]. Tumors rarely grow larger than 5-6 cm in diameter. Larger schwannomas are infrequent, accounting for only around 0.3–5% of all cases [4-10].

Because of the lack of characteristic symptoms, and physical and radiological examinations, preoperative diagnosis is exceedingly challenging. The most common symptoms seen in retroperitoneal schwannomas at the advanced stage are abdominal pain and a palpable abdominal mass; however, unconventional

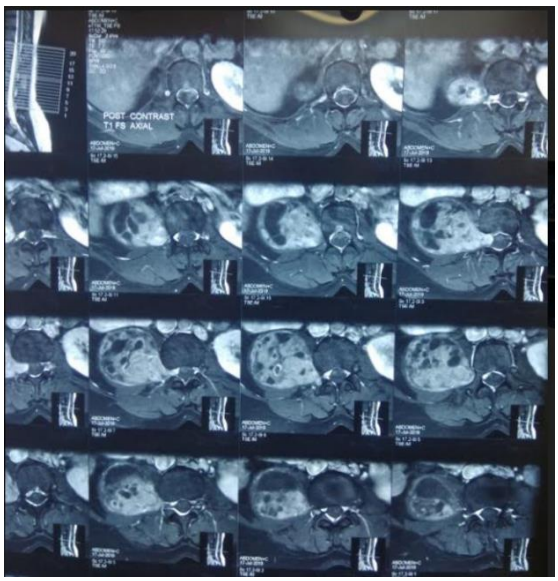
symptoms like renal colic, secondary hypertension, and hematuria have been reported previously [11,12]. Despite advanced imaging facilities like computed tomography, ultrasound, and magnetic resonance imaging, aiding the preoperative diagnosis, an absence of particular imaging signs suggests that the definitive diagnosis of GS must rely on a intraoperative frozen section (IOFS) and a post-op histological examination. To diagnose schwannomas, tumor excision and postoperative pathological examination are required [12]. In the present report, we documented a case of giant schwannoma in a 35-year-old patient who was treated successfully using the retroperitoneal and posterior approach.

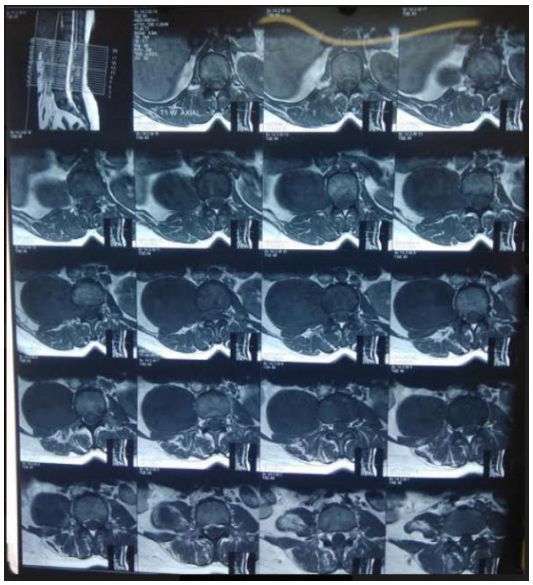
Case report

A 35-year-old female reported to our department with complaint of a progressive right abdominal pain from a UTI for a month, which was relieved by an oral painkiller without vomiting, nausea, urinary frequency, or urgency. Physical examination exhibited a rebound, tender, and large renal mass in the right upper quadrant that was palpable. The MRI revealed a GS emerging from the right L2 nerve root, measuring 7.4x5.5x9cm in size. (Figures 1 and 2) It extended from L1 to L4, creating pressure on the ureter and kidney. There was an absence of neurological deficit. There were no addictions or allergies in the patient. Normal bladder and bowel movements were observed. The general examination was indicated to be uneventful, with normal vital signs. As mass grew to an enormous volume and had ample blood supply, a sufficient volume of blood was prepared. The tumor was excised via the posterior and retroperitoneal approaches.

The posterior approach was initiated by exposing L1, L2, L3, and L4. L2 and L3 laminectomy was performed. The L2 inferior facet and L3 superior facet were removed, exposing the Foraminal and extraforaminal area. The tumor was arising from the right L2 Nerve root. Therefore the right L2 nerve root was ligated and cut. Dura was opened using the 15-number knife to locate the intradural component. No intradural component was found. The tumor on the posterior aspect resected piecemeal as it adhered to the dura and surrounding structures. Gradually posterior part of the tumor was released from surrounding structures and resected. The fusion was performed using the right L2 and L3 pedicle screw, rods, and posterolateral bone graft. Posteriorly, the tumor was isolated from the L2 nerve root, and the intradural and extradural parts were removed. Using the retroperitoneal approach, residual tumor removal was done. An oblique incision was made in the flank region for the retroperitoneal approach. The external and internal oblique muscles were resected. Fascia trasversalis was incised. As the retroperitoneal space was approached, the tumor was identified to be embedded in the substance of the psoas major muscle. (Figure 3) The fibres of psoas major muscle were bluntly dissected from the tumor. It was gradually released from the IVC, kidney, and ureter. Posteriorly, the tumor was released from L2 and L3 vertebra transverse processes. The extraforaminal, retroperitoneal part of the tumor was resected en bloc by blunt dissection. (Figure 4)

The surgery lasted for 7 hours. The blood loss recorded for the anterior and posterior surgery was 300 ml and 1200 ml, respectively. No postoperative complications or neurological deficits were seen, and the patient was mobilized by day 3 and was subsequently discharged. Histopathology done of multiple sections of the renal mass revealed a benign lesion with GS features. The patient was followed for half a year, and there was no recurrence.





Figures 1 and 2: MRI showing Schwannoma

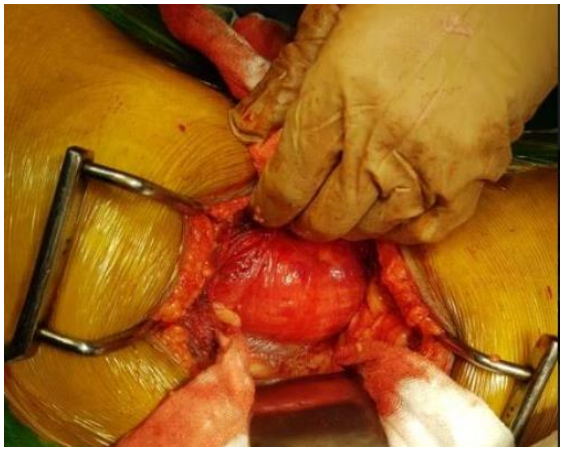


Figure 3: Intraoperative photo of Schwannoma

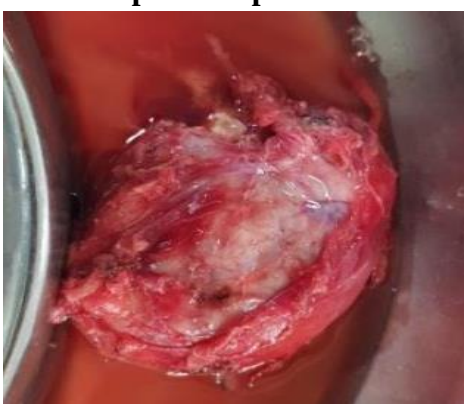


Figure 4: Schwannoma after Excision

Discussion

Giant Schwannomas grow through the nerve Schwann cells, except the first and the second cranial nerves , as they lack Schwann cells, whose visceral areas include the liver, stomach, kidney, pancreas, brain, and heart [13]. They are most prevalently seen in middle-aged females [2], as in our report. GS are typically identified by accident as the retroperitoneal space is flexible, making the diagnosis frequently deferred; as a result, the lesion grows into a considerable size by the time of diagnosis where the patient presents with non-specific symptoms like vague abdominal pain and a dull ache [14,15]. Unconventional symptoms involve hematuria, flank pain, headache, recurring renal colic pain, and secondary hypertension [14].

In general, benign schwannomas have a diameter of less than 5 cm. The probability of a tumor transforming into a malignant one increases with tumor diameter. Moreover, some giant tumors were accompanied by other pathologies [14]. When describing if a tumor is malignant or benign, biological behavior, tumor size, distinguished histology, radiological findings, and distinct immunohistochemical staining must all be considered. A tumor located retroperitoneally typically develops to a large size. A benign schwannoma is smaller in size, well encapsulated, a lesion that is not adherent within a peripheral nerve, and an axon that does not penetrate it. A large (>3cm) lesion adherent to surrounding tissues, on the other hand, is indicative of a malignant GS. Tumour size is used in some publications to rule out malignancy (>3cm) [9]. An invasion process is replicated when a tumor grows rapidly and as large as it compresses surrounding structures or erodes hard tissues. Malignant schwannomas behave similarly to sarcomas of higher grade, with local recurrence and distant metastasis. No particular radiological features correspond with GS. However, well-confined round mass ,aided by contrast enhancement , which is heterogenous due to its cystic with hemorrhagic changes and/or calcification with cystic changes in large tumors can be suggestive. 40% of schwannomas contain cystic components [6, 8, 16,17].

Since malignancies cannot be ruled out preoperatively, IOFS may provide an inaccurate diagnosis pathologically. As GS could be insensitive to radiation and chemotherapy [10], it is strongly advised that they be treated with complete excision with surgery along with negative soft tissue margins [18,19]. Nevertheless, it is debatable whether or not adjacent tissue and viscera should be removed. Several authors had proposed that simple enucleation or even partial excision excluding the removal of adjoining organs was satisfactory owing to benign nature of the majority of cases and the possibility that loss of adjacent organs could impact the prognosis [20,21]. The choice of surgical approaches, such as robotic laparoscopic resection or laparoscopic surgery, has great potential in treatment. However, they need to be influenced by tumor size and location. The average diameter of GS is 5cm, and involvement of the urinary system is rare, making our case exceptional [21].

A large number of these tumors are seen originating from paravertebral space or the presacral region [21], demonstrate fewer symptoms in the earlier stages due to a large amount of space in the broad expandable retroperitoneum allowing the tumor to achieve large sizes before showcasing any symptoms, resulting in delayed diagnosis. Due to the absence of symptoms, the mass was observed late in our case, delaying treatment which made the surgery more complicated owing to the tumor size. A complete resection resulted in lower recurrence or malignant transformation of about 5 to 10% [22]. Even so, if resection is left incomplete, a local recurrence is seen in 10–54 percent of cases within the first 6 months post-surgery [12]. However, our case exhibited no such recurrence when followed up for 6 months.

Conclusion

Retroperitoneal schwannomas are highly uncommon, with only a few reports documented in the literature. These are rarely symptomatic and are usually discovered accidentally. In cases of retroperitoneal mass, they can be used as a differential diagnosis. Although a majority of the reported patients have had a positive outcome, the biological characteristics of renal schwannoma are still uncertain due to a lack of evidence and the rate of recurrence after surgery. Our current report may provide additional information for further research.

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DECLRATIONS-

Consent- Proper informed written consent taken from the patient and relatives to use the information for publication for academic purposes

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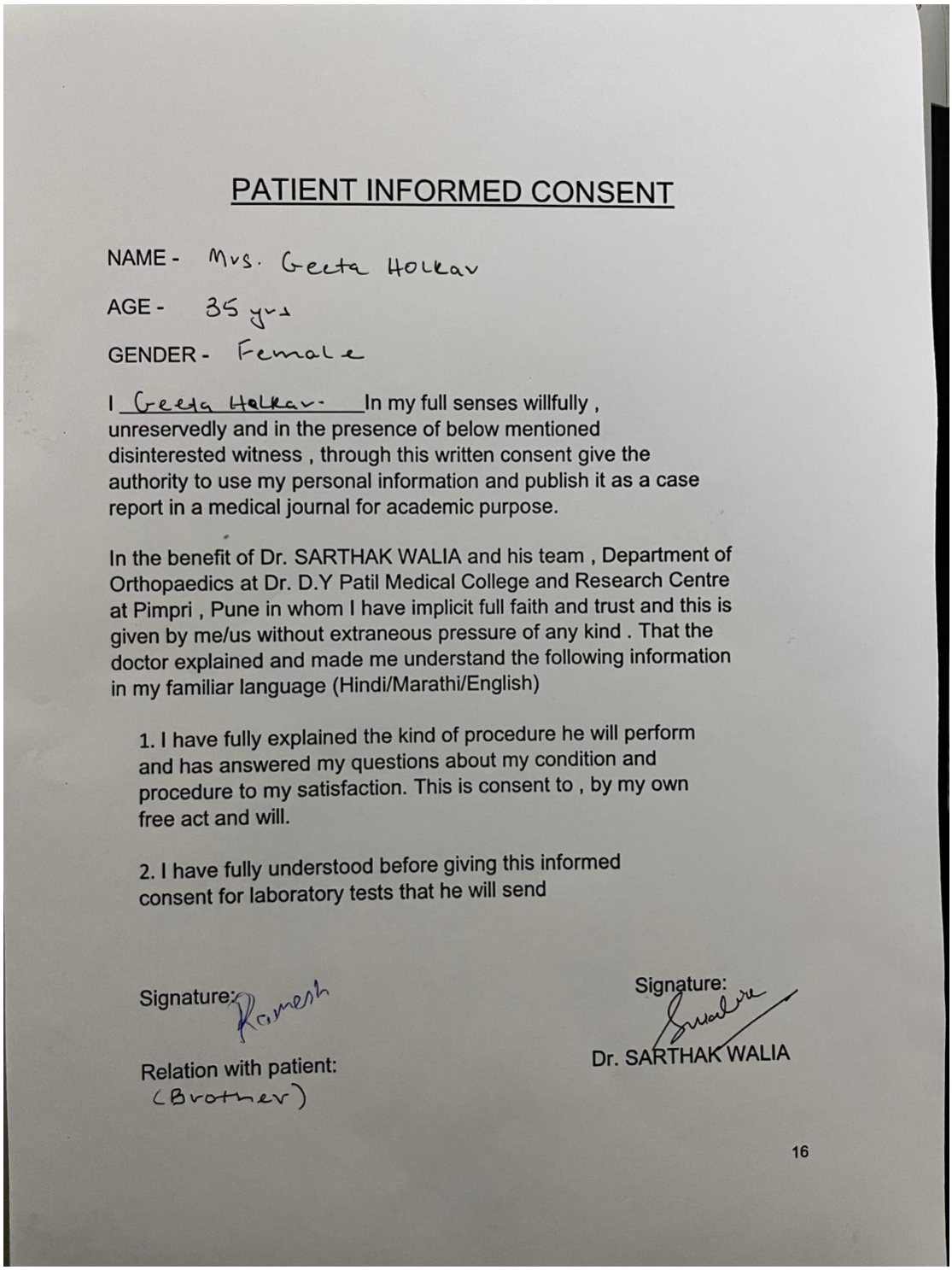
Conflict Of Interest- None



PATIENT INFORMED CONSENT

NAME - Mrs. Geeta Holkar

AGE - 35 yrs

GENDER - Female

I Geeta Holkar. In my full senses willfully , unreservedly and in the presence of below mentioned disinterested witness , through this written consent give the authority to use my personal information and publish it as a case report in a medical journal for academic purpose.

In the benefit of Dr. SARTHAK WALIA and his team , Department of Orthopaedics at Dr. D.Y Patil Medical College and Research Centre at Pimpri , Pune in whom I have implicit full faith and trust and this is given by me/us without extraneous pressure of any kind . That the doctor explained and made me understand the following information in my familiar language (Hindi/Marathi/English)

1. I have fully explained the kind of procedure he will perform and has answered my questions about my condition and procedure to my satisfaction. This is consent to , by my own free act and will.

2. I have fully understood before giving this informed consent for laboratory tests that he will send

Signature: Ramesh

Relation with patient: (Brother)

Signature:

Dr. SARTHAK WALIA

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