A Huge 5kg-Intra-Abdominal Ancient Schwannoma with A Recurrence-A Case Report.

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***Abstract*: Introduction- This is a rare case of a recurrent huge intrabdominal Ancient Schwannoma, the size of which has not been reported before. This case report aims to enrich the literature on cases of undiagnosed abdominal masses.**

Clinical features-The patient complained of huge abdominal distension, weight loss, and breathlessness of 4 months duration.

The important clinical findings were multiple cutaneous painless swellings, hyperpigmented patches on the trunk, and a mass occupying the whole of the abdomen.

Management-A diagnosis of visceral manifestation of neurofibromatosis was made to rule out an intra-abdominal malignancy. Preoperative Abdominal Computed Tomography (CT) scan revealed a huge intrabdominal mass while a CT-guided biopsy showed a Benign Mesenchymal tumor. He had an exploratory laparotomy, and the mass was totally excised. The recovery was uneventful, and the outcome was excellent. Histopathology showed that the tumor was a Schwannoma with no malignancy. However, 5 months after, the swelling recurred and he was eventually lost to follow up.

Conclusion- The rarity of Intra-abdominal Schwannomas and absent or non-specific signs and symptoms present the surgeon with a diagnostic dilemma. Preoperative Abdominopelvic CT scan, Ultrasound scan, and Magnetic Resonance Imaging are necessary imaging modalities. Complete excision at surgery reduces recurrence. Close follow-up is essential.

***Keywords*- Schwannoma, Neurofibromatosis, Café-au-lait**

I. INTRODUCTION

Ancient Schwannomas are benign masses that rarely occur in the abdomen and even when they occur, they rarely get as big as was seen in this case. They occur more as rare solitary[1] nerve sheath tumors of peri-neural Schwann cells, that affect the nerves of the extremities, trunk, and the head and neck region. They are commoner in females[2], between the third and fifth decades, occur spontaneously but could also present as part of Carney's complex, (comprising conjunctival disorders, myxomas, schwannomas, and other endocrine disorders like Cushing's syndrome)[3], or in association with a group of genetic autosomal dominant disorders called neurofibromatosis 1 and 2 (NF1 & NF2)[4]. They grow slowly[5] in the abdomen without signs and symptoms until they occupy the available space and then begin to impinge on surrounding structures to cause intermittent pain. They could be benign or malignant in the abdomen, or sarcoma in the retroperitoneal space. Only 0.5-5% of all retroperitoneal tumors are Schwannomas[5]. Malignant transformation is very rare.

Ancient Schwannoma refers to the degenerative changes that occur in the tumor with increasing duration. It constitutes a rare and uncommon cause of abdominal masses, which could cause a diagnostic dilemma. Pre-operative diagnosis is difficult even with radiological techniques. However, Fine Needle Aspiration (FNA) or CT-guided biopsy could be used to obtain a preoperative histological diagnosis. However, there is the possibility of a wrong interpretation of malignancy in FNA[6]. There is also the risk of hemorrhage and the inadvertent spread of malignancy.

Treatment is by complete surgical excision. Recurrence could occur especially in malignant cases, which are associated with von Recklinghausen's disease. Recurrence following total excision is reported in 11.7%[6] of cases.

This is a case of a Huge 5kg- Abdominal Ancient Schwannoma, the first of its kind in our center, the Federal Medical Center, Ebute Metta Lagos, Nigeria, a tertiary center and the first of this size as far as we know in the literature.

II. CASE REPORT

This patient is a 35-year-old male who presented with a 4-month history of progressive intermittently painful abdominal distention. This distension began in the pelvic region and extended to the whole abdomen, associated with significant weight loss. Since childhood, the patient had noticed painless papules on the right chest, which increased gradually. There was no other relevant personal history or symptom, and there was no family history of similar multiple cutaneous painless hyperpigmented spots or cancer.

There was no surgical consultation or intervention before he presented at our facility, but he used analgesics periodically to combat abdominal pain. There was a positive surgical history, having had a right herniorrhaphy 18 years, and a left herniorrhaphy 2 years, before presentation.

Examination revealed a young man, in respiratory distress, cachectic, and pale.

The abdomen was markedly distended, with a mildly tender mass extending from the xiphisternum to the pelvis. The liver and spleen were not enlarged, but there was ascites, demonstrated by shifting dullness. Bowel sound was normal. Per rectal examination showed prolapsed hemorrhoids.

Chest showed a soft tissue hyperpigmented mass, 10x8cm over the right hemithorax, covering the right breast and

nipple. There were hyperpigmented skin patches over the chest and abdomen.

Laboratory investigations done, including Liver Function Tests (LFT), Electrolytes, urea, and Creatinine (EUC) were all within normal limits apart from Packed cell volume (PCV) which was 27%.

Imaging modalities used included Abdomino-Pelvic ultrasound scan (USS) which showed a large complex intra-abdominal mass with gross ascites of unknown cause, and Abdomino-pelvic CT scan which showed a huge intra-abdominal mass measuring 21.2 x 11.6 x 11.2 cm, with ascites. The mass showed heterogeneous enhancement with central areas of hypoattenuation suggestive of a necrotic component. No retroperitoneal adenopathy was seen. A Chest CT scan revealed suspicious pulmonary nodules (the film is not available as it was done in another facility). However, a repeat Chest CT scan during one follow-up visit did not reveal any abnormality. A pre-operative CT-guided biopsy of the tumor was done, and a diagnosis of mesenchymal neoplasm most likely benign was made.

The diagnostic challenge was getting funds for immunohistochemistry, which would have enabled us to properly classify the tumor.

A working diagnosis was Plexiform Neurofibromatosis with visceral manifestation, to rule out Gastrointestinal Stroma Tumor (GIST).

The diagnosis was clearly explained to the patient and his guardian, following which written consent for surgery was sought and obtained.

Prognostic characteristics included the size of the tumor and the possibility of recurrence.

The therapeutic intervention was to optimize him for surgery. He was promptly admitted and transfused with a pint of whole blood. He had bowel preparation done and was placed on nil per oral the night before surgery.

At induction, he was administered prophylactic intravenous Ceftriaxone 1g. Access was gained to the peritoneal cavity via a midline longitudinal incision. A mass extending from the epigastrium to the pelvis was seen, attached to the posterior abdominal wall and small and large bowel loops. In the pelvis, it was attached to the posterior surface of the sigmoid and rectum. There were extensive seedlings in the mesentery and anterior abdominal wall. The mass was freed from bowel attachments, commencing with the small bowel, by performing resection and end-to-end – hand-sewn anastomosis, after which it was then freed from the post-abdominal wall avoiding injuries to the major vessels. It was eventually dissected off its attachment to the posterior surface of the sigmoid colon and the rectum. The mass weighed 5kg. (Figure 2).

The patient did well postoperatively and was happy that we were able to remove the mass and that he no longer experienced breathlessness. He tolerated oral intake very well and had a normal bowel habit. Post-operative investigations were normal. There were no adverse or unanticipated events. He was discharged on the 9th day. Postoperative abdominal and Chest CT scans at 3 months were both normal.

The histopathology report was that of a greyish-white mass, weighing 5000g, measuring 26x23x16cm. Cut sections showed a tan-colored tumor with areas of cystic degeneration (Figures 3,4). Microscopic examination showed a biphasic neoplasm composed mostly of hypercellular areas and myxoid hypocellular areas. Tumor cells are narrow, elongated, and wavy (Figure 5). There were no mitotic figures. The features are those of Schwannoma.

At the surgical outpatient clinic for follow-up, he complained of occasional lower abdominal pain which responded well to analgesics. However, about 6 months following surgery, during follow-up, after a period of 6 weeks' missed appointments, he re-presented with huge abdominal swelling, associated with breathlessness and weakness. Examination revealed a huge intra-abdominal mass involving the whole of the abdomen. A diagnosis of recurrent intra-abdominal Schwannoma was made. He was to have pre-operative investigations done, including Abdomino-pelvic CT scan, Chest CT scan, Complete blood count, electrolytes, and urea. However, the patient was lost to follow up and all efforts to reach him failed.

Informed and written consent was taken from the patient to report this case.

Figure 1- Cutaneous masses and spots

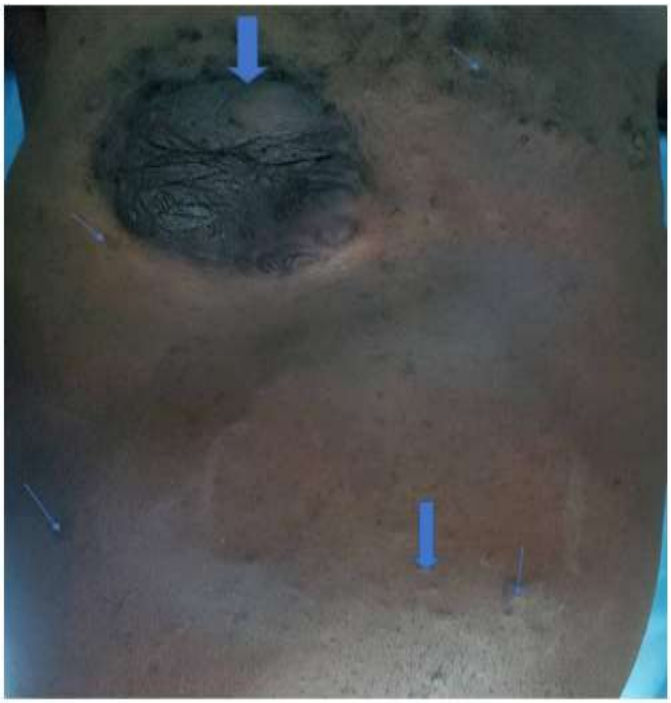


Figure shows Cutaneous Masses (Big Arrows) and Café-au-lait spots (Small Arrows) on the anterior chest and upper abdomen.

Figure 2. Mass at Surgery

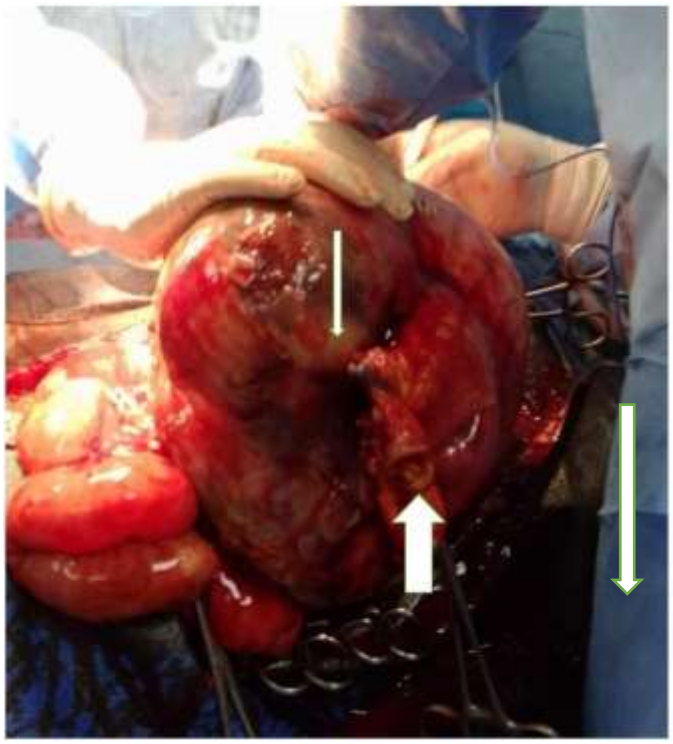
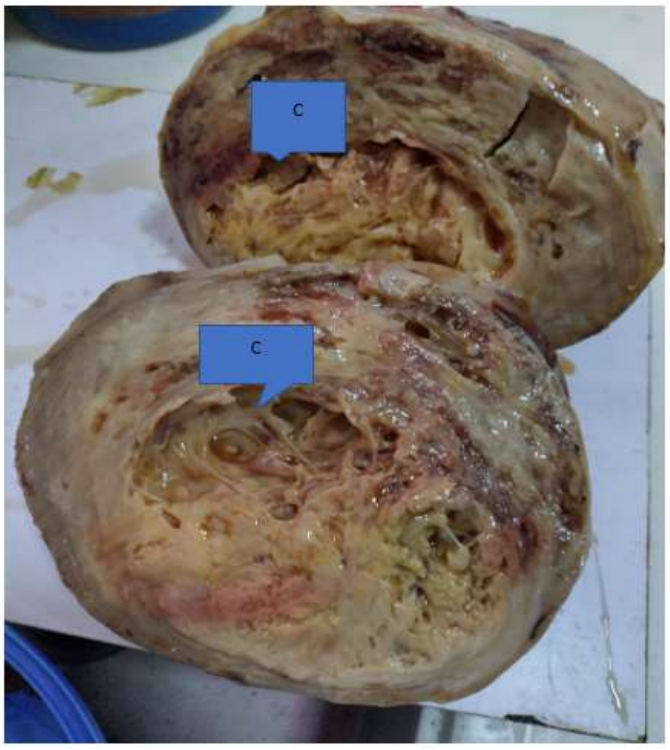


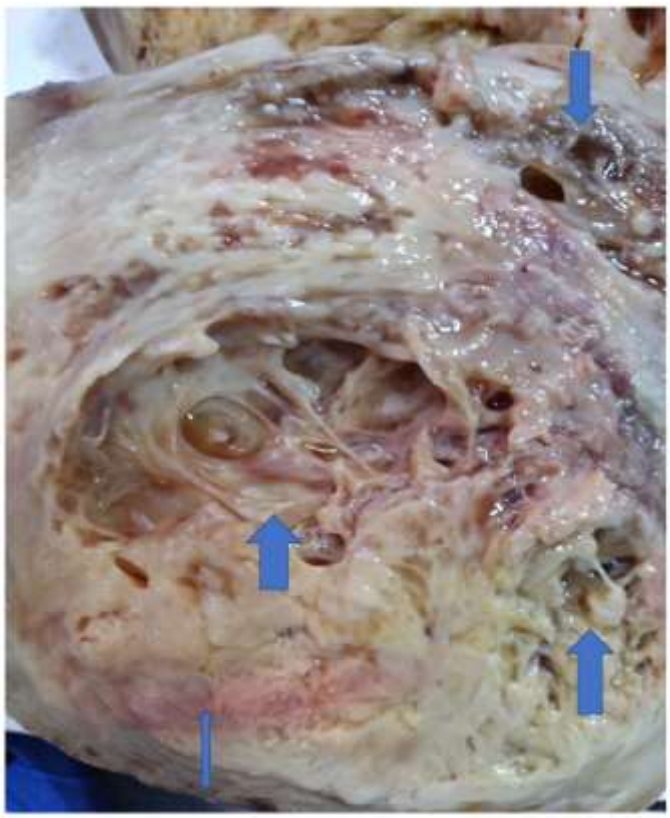
Figure 2 shows the mass at surgery with cystic areas (narrow arrow) and attached bowel (wide arrow)

Figure 3. Cut surface showing cavities and solid tumors.



The cut surface of the tumor shows cavities (C) within the solid mass.

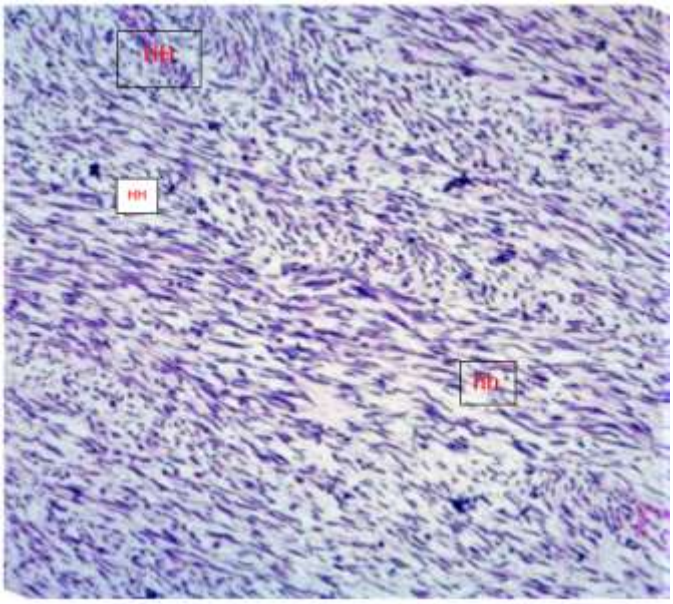
Figure 4. Degenerative changes



This figure shows evidence of degenerative changes (Large arrows) with cystic areas mixed with solid components (small arrow).

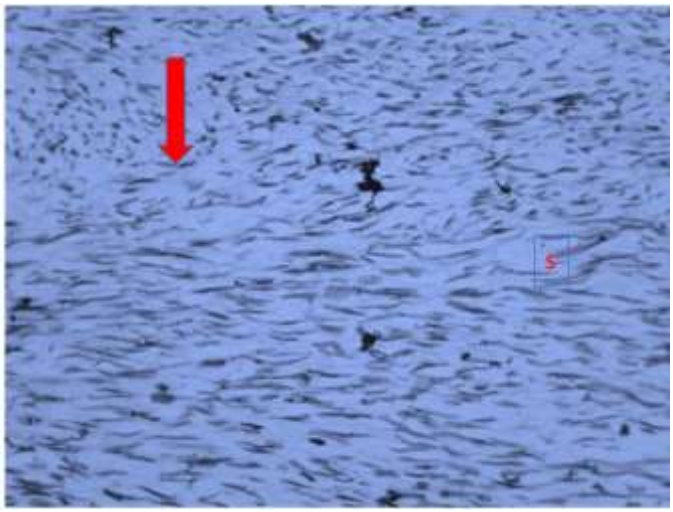
Figure 5.

A. Photomicrograph of the specimen (Low power).



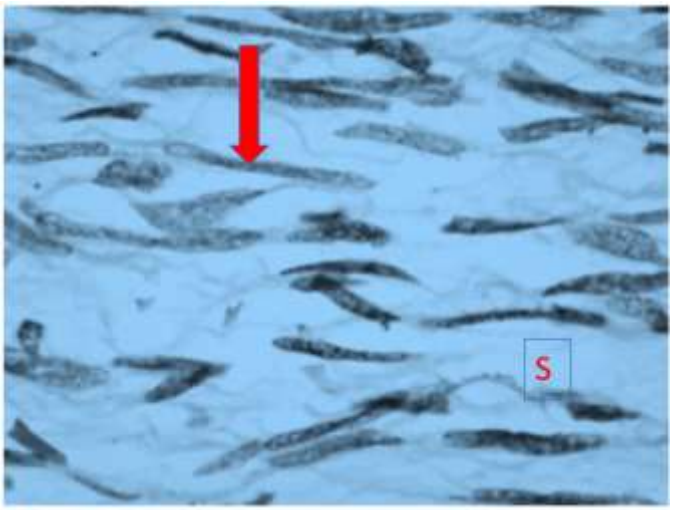
The figure shows areas of hypercellularity (HH) and hypocellularity (hh).

B. Medium Power



The figure shows Schwannoma cells(arrows) and Myxoid Stroma (S).

C. High Power



High Magnification of the Schwannoma Cells (Arrow) and Myxoid Stroma (S).

III. DISCUSSION

Schwannomas are tumors of Schwann cells of nerve sheaths. They occur commonly in the extremities, head, and neck but they are quite rare in the abdomen, with 0.75-2.6%[7] occurring in the retroperitoneum. They are benign well-encapsulated solid tumors with cystic areas due to degenerative changes that occur with tumor longevity, which is due to the loose areolar space that allows the mass to grow slowly and without symptoms until it reaches such a size that it begins to compress on surrounding structures, resulting in pain[8] and numbness. They extend from the paravertebral gutters and pelvis and may eventually occupy the whole abdomen.

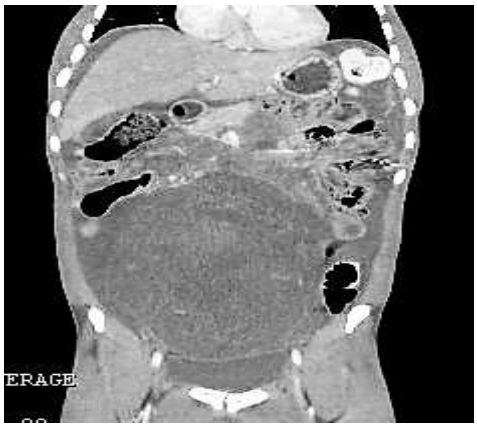
These tumors are associated with neurofibromatosis (NF), a genetic autosomal disorder. There are two types NF1 and NF2. NF1 has mainly cutaneous manifestations with multiple small skin masses, and hyperpigmented skin spots called "café-au-lait" spots spread on the trunk, axillary, or inguinal freckles. There could be a first-degree relative with NF1. It has a prevalence of 1:5000[9]. Their life expectancy is 10-15% lower than the general population[10]. The prevalence of NF2 is 1:60,000[9].

The features the patient presented with satisfied the criteria for diagnosis of NFI[12,13].

Investigations include abdominopelvic CT and ultrasound scans, with complete blood counts and electrolytes, urea, and creatinine. While a pre-operative CT-guided biopsy is good and helpful, it is usually not recommended[7] because of possible complications like hemorrhage and the spread of tumor seedlings.

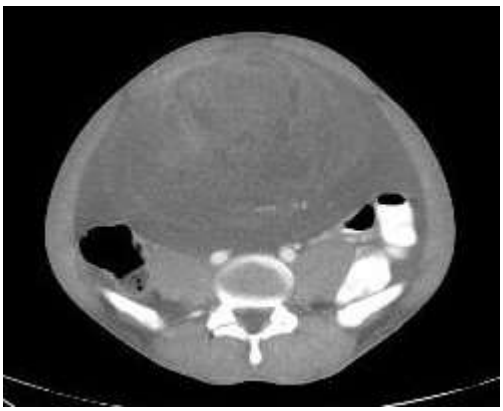
Figure 6- CT image

A.



Coronal view of Abdomino-pelvic CT showing a huge mass in the abdomen down to the pelvis.

B.



CT Image showing a huge central abdominal mass (Arrow).

The presence of ascites and tumor seedlings in the mesentery of the small bowel and abdominal wall raised the suspicion of malignancy. The histologic findings of alternating areas of dense cellularity with dense aggregation of spindle-shaped cells (Antoni A) and reduced cellularity with loose myxoid

matrix (Antoni B)[14] confirm the diagnosis of "Ancient" Schwannoma. It is "ancient" because of the continuous degeneration of the tumor due to the duration ('age') of existence, producing[3,15] cystic, necrotic, and fibrotic changes and nuclear atypia, which can easily be confused for malignancy.

Retroperitoneal Schwannomas are usually larger than at other sites. The abdomen provides a large space for extensive growth of the tumor without causing symptoms. Although signs and symptoms for Abdominal Schwannomas are non-specific[7] or non-existent initially, presentation depends on the location and the duration of the tumor. Those with Schwannomas in the upper gastrointestinal tract like the stomach and duodenum may present with vomiting, nausea, intestinal obstruction, and anorexia, while in the rectum may present with rectal bleeding, change in bowel habit, and intestinal obstruction[15].

Pre-operative diagnosis is difficult and challenging. Those presenting with gastric or intestinal lesions will benefit from upper gastro-intestinal endoscopy[16] and biopsy. Others will benefit from CT and MRI evaluation. Colonoscopy is necessary for patients with colonic masses.

Some authors have reported cases with features of malignancy preoperatively but were histopathologically benign. *Shelat et al* did anterior resection on a patient with a rectal mass with lymph node involvement. However, the tumor was benign and well-circumscribed, and all harvested lymph nodes were negative[15]. Similarly, this index case not only had mesenteric lymph nodes, but also ascites and anterior abdominal wall seedlings which raised the suspicion of malignancy but there was no malignancy in the surgical specimen.

Differentials include Gastrointestinal Stromal Tumors (GIST), leiomyosarcoma, liposarcoma, lymphangioma, and other mesenchymal tumors. It could also be gastrointestinal autonomic nerve tumors (GANT), from the gastrointestinal autonomic nerve plexuses. It is slow-growing, and benign but with an aggressive course and poor prognosis[16]. Immunohistochemistry for staining to protein S100 would have helped but it is not yet available. This was a limitation for us.

Surgery, which could be open or laparoscopic, is the mode of treatment. Complete surgical excision[17] is therapeutic. However, there is no consensus about the practicability of achieving negative margins in all situations due to the proximity and attachment of important structures like blood vessels. Some authors believe such structures must be sacrificed to achieve a negative margin, while others advocate tumor enucleation. This patient had complete excision. Recurrences after complete excision are associated with rare malignant variants and the prognosis is poor[18]. Unfortunately, our patient had a recurrence and we eventually lost him to follow while preparing him for repeat surgery.

Follow-up is essential in case of recurrence. The patient complained of abdominal pain during each follow-up visit. Chronic postsurgical pain occurs in 15-30%[19] of patients who had minor or major abdominal or pelvic surgeries. It could be neuropathic or from the scar. The use of multimodal analgesic techniques[18] could reduce the incidence. Pain could also signal the onset of recurrence.

IV. CONCLUSION

Abdominal and retroperitoneal Schwannomas are scarce, and the paucity of signs and symptoms renders preoperative diagnosis quite challenging in our environment. Investigations largely depend on the mode of presentation and the site of the tumor. Treatment is by complete surgical excision through an exploratory laparotomy for big masses or endoscopic resection in the gastro-intestinal tract where possible. Recurrence is rare since most of them are benign tumors. However, those with recurrences, like in this case, probably have malignant variants, with a very poor prognosis. Therefore, adequate long-term follow-up is essential.

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