

# AN UNUSUAL CASE OF DUODENAL NEURILEMOMA

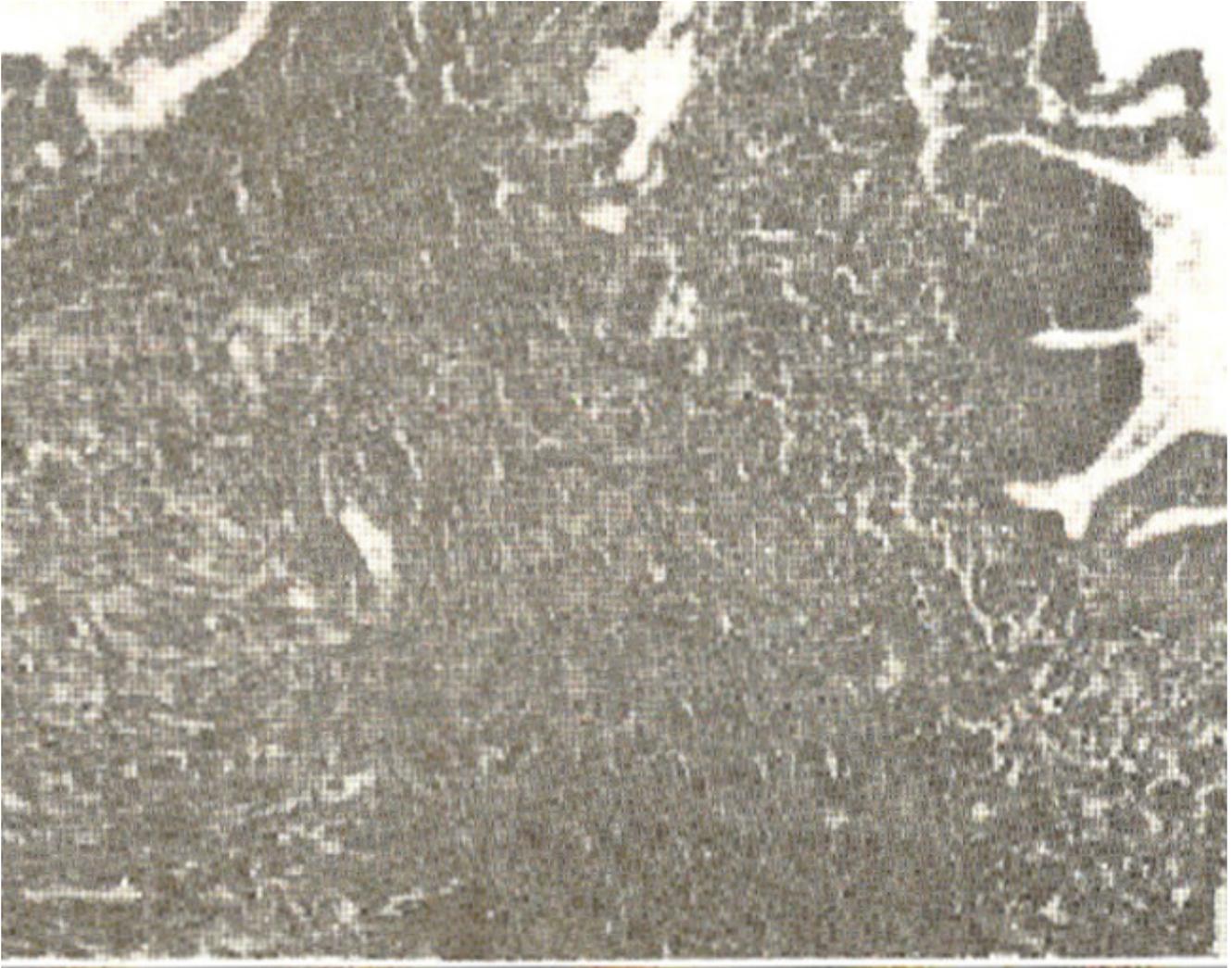
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Neurilemoma is a benign tumor of nerve sheath, commonly found along the course of cranial and peripheral nerves. Less common sites include oronasal cavity, lung, gastrointestinal tract (GIT), breast and thyroid mostly as part of Von Recklinghausens disease (VRD). Isolated neurilemmomas in the GIT are rare, the stomach being the usual site<sup>1</sup>. We report a case of isolated duodenal neurilemoma with several unusual features.

## CASE HISTORY

A 45 years old Pakistani cobbler, presented with a history of recurrent episodes of melena associated with mild upper-abdominal discomfort for the preceding five years. In the immediate past, he had also developed excessive fatigability and progressive dyspnoea. There was no history of jaundice or chest pain nor any relevant drug intake. On examination, he looked ill, was pale and orthopneic. There was a marked koilonychia and glossitis, J.V.P. was raised and bilateral ankle edema was present. The pulse was 100/min, regular, bounding with a blood pressure of 100/50 mm Hg. He had a hyperdynamic apex beat in the 5th left ICS 2cms outside the MCL alongwith a presystolic gallop and haemic murmur. There were bibasal crepitations in the lungs. The liver was palpable, 5 cm in MCL. There was no free fluid in the abdomen. Per-rectal examination and proctoscopy were normal. CNS examination was found to be normal. Laboratory examination revealed amicrocytic hypochromic anaemia with a HB of 3 mg/dl and ESR of 60mm/1st hr, low serum iron and high levels of TIBC. The urine showed no abnormality. Stool showed occult blood but ova or cysts were not found on repeated examinations. Liver function tests, bleeding and clotting time, blood sugar, urea, serum calcium, phosphate, creatinine and electrolytes were all within the normal range. Chest X-ray revealed cardiomegaly with pulmonary edema. Electrocardiogram showed left ventricular dominance. Echocardiography showed enlarged left ventricle with normal valves and no pericardial effusion. Isotopic liver scan showed normal uptake of TC 99 and no cold area. Ultrasound examination of the abdomen showed normal viscera and no abnormal mass anywhere. Bone marrow aspiration was found to show reactive hyperplasia. On upper G.I. endoscopy, varices were not seen. Although the gastric and duodenal mucosa were normal, there was a single reddish polyp measuring 2x2 cm, with friable surface in the first part of duodenum. A biopsy sample from this growth showed a neurilemoma on light microscopy with no evidence of malignant change (Figure I).



**Figure 1. Photograph of duodenal mucosa showing  
Neurilemoma (x 20 H & E).**

The patient was treated conservatively and his condition was stabilized. He refused surgery and took his discharge from the hospital.

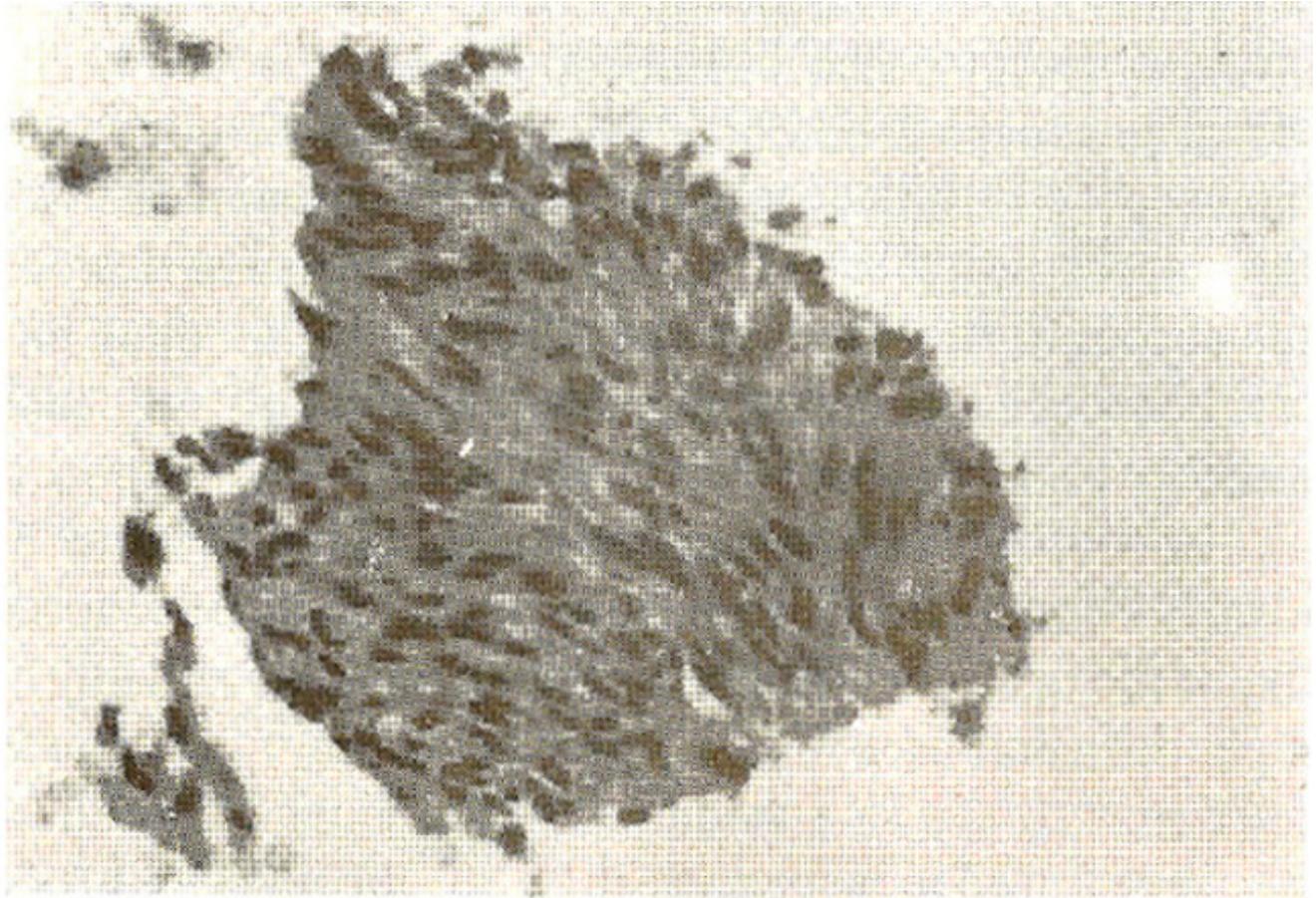
**Second admission**

He was readmitted three months later in a moribund state. He had severe anaemia, was cachectic with features of severe congestive cardiac failure. His liver showed further enlargement but this time it was firm and asymmetrical. Liver scan showed a cold area and ultrasound demonstrated a cystic mass in the liver (Figure 2).



**Figure 2. Photomicrograph of a ultrasound Scan showing a large hypoechoic area alongwith some echogenic shadows in left lobe of liver.**

Endoscopy was repeated which again showed a single duodenal polyp of the same size with no extension. Biopsies were taken from the duodenum as well as the liver. While the histological report from the duodenum was identical with the previous report, the histology of the liver showed the presence of a neurofibrosarcoma (Figure 3).



**Figure 3. Photomicrograph of a Neurofibrosarcoma. Biopsy showing hyperchromatic, elongated nuclei and variable size and shape of cells (x 20 H & E).**

Patient's poor general condition did not allow any aggressive measures. Despite the best medical efforts, his condition gradually deteriorated and he later died of acute pulmonary oedema.

### **COMMENTS**

Duodenum is an uncommon site of either benign or malignant tumors. Though occurrence of neurilemmomas or Schwannomas in the duodenum has been described occasionally but it is always in association with VRD. These are usually benign, slowly growing tumors. Secondary changes like cystic and Xanthomatous degeneration and vascular thrombosis<sup>2</sup> are well known but sarcomatous transformation is very rare<sup>3,4</sup>. Although they may present with epigastric pain or symptoms of obstruction, in most instances there is a pattern of recurrent episodes of melena, hematemesis, and haematochezia followed by relatively asymptomatic periods<sup>5,6</sup> in our patient, though the clinical presentation was quite characteristic but the absence of any external evidence of VRD makes it an unusual case. Another unusual feature was development of a large neurofibrosarcoma in the liver in a short span of three months where the previous ultrasound and isotopic liver scans were normal and the biopsies from the primary site, i.e., duodenal polyp, did not show any malignant change on light microscopy on both occasions. Unfortunately the patient refused surgery during his first hospitalization which not only proved fatal for him but also denied us a better histological specimen. It is possible that

an early sarcomatous transformation of duodenal neurilemoma was missed because of sampling error or because of lack of facilities for electron microscopy and immunocytochemistry.

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