

Rare Presentation of Retroperitoneal Schwannoma: a Case Report

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Abstract

The deadly Schwannomas, as uncommon subtype of soft tissue tumors originate from peripheral nerve sheaths. Retro peritoneum is an uncommon site for these tumors.

Here we present the clinical feature and therapeutic implication of a 38-year-old man affected by recurrent retroperitoneal schwannoma who admitted to emergency ward following abdominal pain. Computed Tomographic Scan showed a solid well-differentiated heterogeneous mass, 8.5×6 cm size in portahepatis beneath liver and right periumbilical. On laboratory tests Cancer Antigen 19-9 and Cancer Embryonic Antigen were in normal range. In surgery, a 7×8cm round mass was discovered at juxta renal juxta duodenal retro peritoneum. It was completely adhered to second part of duodenum with no signs of infiltration of the liver and right kidney, unable to simply being enucleated, curative resection was performed by classical whipple procedure. The patient's hospitalization course was uneventful and on the second week he was discharged from the hospital with complete recovery.

Due to the high recurrent rate of retroperitoneal Schwannomas even in benign circumstances, radical resection would be the treatment of choice.

Key words: Schwannoma; Retroperitoneal neoplasm; Whipple procedure

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Introduction

Schwannomas, as uncommon subtype of soft tissue tumors, originate from peripheral nerve sheaths. They are reported most commonly in the head and neck, also flexor aspect of upper and lower limbs, which account for at least 90% of cases. All other sites, including the retroperitoneum, are rare, accounting for less than 10% of cases [1, 2]. 3-3.2% of benign Schwannomas, as in contrast to merely 1.7% of their malignant subtypes occur in retro peritoneum [3-5].

Here we present the clinical feature and therapeutic implication of the patient affected by recurrent retroperitoneal schwannoma who admitted to emergency ward following abdominal pain.

Case Report

A 38-year-old man, presented with chronic abdominal pain that accelerated during last few months and epigastric pain for three months

worsening in last forty-eight hours admitted to emergency ward.

He described constant, moderate pain associated with chronic constipation; other constitutional symptoms were less than 10% weightloss.

His past surgical history was remarkable for similar symptoms in less than five years ago resulted in midline laparotomy and mass resection.

His abdominal physical exam was remarkable for a firm mass of approximately 7×7 cm size, immobile with regular border.

Physical examination was unremarkable for café-au lait spots or other Recklinghausen's disease features.

On laboratory tests, the liver function tests were normal. As following Cancer Antigen 19-9 was 10u/ml (reference range: below 35u/ml.) and Cancer Embryonic Antigen (CEA) 1.5 ng/ml (reference range: below 2.5ng/ml in Non-smokers).



Figure 1. It shows a solid well-differentiated heterogeneous mass, 8.5×6 cm size in portahepatis beneath liver and right periumbilical.

A dynamic sixteen slices Computed Tomographic Scan with non-ionic contrast showed a solid well-differentiated heterogeneous mass, 8.5×6 cm size in portahepatis beneath liver and right periumbilical (Figure 1).

In surgery, a 7×8 cm round mass was discovered at juxta renal juxta duodenal retroperitoneum. It was completely adhered to second part of duodenum with no signs of infiltration of the liver and right kidney, unable to simply being enucleated, curative resection was performed by classical whipple procedure.

The mass neither encase the Superior Mesenteric Artery, nor the porta hepatis confluence.

The patient's hospitalization course was uneventful and on the second week he was discharged from the hospital with complete recovery.

The pathological analysis demonstrated Spindle cell neoplasm composed of alternatively hypo cellular areas with marked nuclear palisading, well-formed verocay body and hyper cellular areas with edema (Figure 2).

Immuo Histo Chemical staining clarified that the neoplastic cells were strongly positive for s100 protein, while CD117(C-Kit), SMA (Smooth Muscle Actin), Desmin, PDGFR (Platelet Derived Growth Factor Receptor) were negative, moreover CD57 (Leu7) was positive.

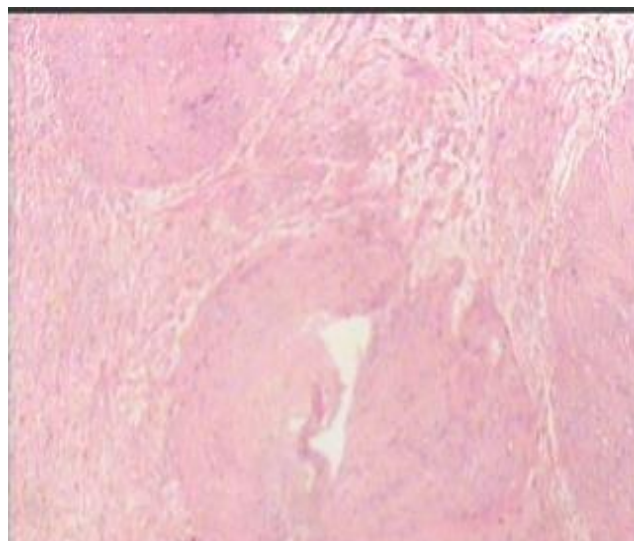


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Discussion

Schwannoma rising from schwann cells are among Schwannoma-neurofibromas entity [6, 7].

A review of literature indicates most Gastro Intestinal involvement among 20-50 year old patients.

However, females outnumbered males, with a ratio of 2:1 [8, 9]. The occurrence of Schwannoma in a retroperitoneal organ such as the pancreas is extremely rare and most likely affects occurrence as a component of Von-Recklinghausen's disease [10]. Schwannomas are encapsulated, single lesions with distinct borders.

Histologically it contains two discrete components Cellular component (Antoni A) arising from spindle cells Hypocellular component (Antoni B) accompanied with degenerative changes [8, 11].

Delayed Diagnosis may be attributed to vast retroperitoneal cavity and Variety of symptoms ranging from vague abdominal pain, distention and secondary hypertension, renal colic may be present [12].

Pre-operative diagnosis modalities; varying from ultrasound and ct to MRI may facilitate the diagnosis [11, 12], but no specific imaging exists [13].

The features suggesting schwannoma in MRI T1, T2 signal imagings include hypo intense, hyper intense mass respectively [13, 14].

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The treatment of choice for retroperitoneal schwannoma is complete surgical resection [15]; particularly in those without expected response to chemo radiotherapy. Some authors believe that complete resection is ideal, an extensive operation sacrificing of vital structures may be warranted in order to achieve negative margins [16, 17].

In patients who underwent partial resection, some investigators have even reported recurrence rates of up to 10-20% even in benign circumstances, thus emphasizing the importance of a complete resection [18].

In our patient, classical pancreaticoduodenectomy (Whipple procedure) was performed due to the complete adherence of tumor to the lateral border of second part of duodenum. After a mean follow up of 12 months no local recurrences or metastases were reported.

Conclusion

Due to the high recurrent rate of retroperitoneal Schwannomas even in benign circumstances, radical resection would be the treatment of choice.

Acknowledgment

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Conflict of Interest

The authors have no conflict of interest in this article.

Authors' Contribution

Mohammad Mozafar and Saran Lotfollahzadeh designed the study; Saran Lotfollahzadeh and Khashayar Atqiaee wrote the paper, Farideh Adhami contributed to the data entry and analyzed the data while Saran Lotfollahzadeh helped in writing the manuscript.

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