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Case Report

Schwannoma masquerading as metastatic retrocaval lymph node in a patient with incidental gall bladder cancer: a case report and review of literature

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ABSTRACT

Schwannomas are rare benign neoplasms that arise from neural sheath Schwann cells. Their presence in retroperitoneum can mimic other differential diagnosis especially in the setting of a co-existing malignancy. A 31-year-old lady without comorbidities presented with a diagnosis of incidental Gall bladder cancer diagnosed elsewhere. Upon further evaluation, she was found to have a retrocaval mass resembling a metastatic necrotic Lymph node on Contrast Enhanced Computed Tomography (CECT) scan of the abdomen with no evidence of disease elsewhere. Due to the unusual location, mass was biopsied which was inconclusive. Surgical excision of the same was done during Revision cholecystectomy and Final histopathology report revealed it to be a benign Schwannoma. Retroperitoneal Schwannoma with degenerative changes can be a great mimicker of metastatic necrotic malignant lymph node and create diagnostic confusion. With prompt suspicion in mind and clinicoradiological correlation, it is imperative to make informed surgical decisions to avoid erroneous diagnoses that could lead to over treatment. This case underscores the importance of being aware of the differential diagnoses and the need for proper surgical resection of schwannomas to prevent recurrence and ensure complete treatment. Simultaneously, it emphasizes the importance of not compromising the management of the primary neoplasm, in this case, gallbladder cancer.

Keywords: Retroperitoneal schwannoma, Schwannoma, Incidental gall bladder cancer, Metastatic lymph node, Masquerade, Retroperitoneal mass, Retroperitoneal lymph node

INTRODUCTION

Schwannoma or neurilemmoma is a benign neoplasm that arise from Schwann cells in the peripheral neural sheaths. They are commonly found in the central nervous system, spinal cord or peripheral nerves in the body and are mainly found in head and neck or extremities, rarely found in the retroperitoneum.[1] Majority of them will be clinically occult and may manifest symptoms when they overgrow and start compressing adjacent structures. Retroperitoneal schwannomas are often misdiagnosed in imaging as pancreatic mass, liver tumor, psoas abscesses or as metastatic lymph node.[2-6] Cystic degeneration can occur when the tumor outgrows its blood supply and can mimic necrotic malignant lymph node.[5]

We report a case of a patient with incidental gall bladder (iGBC) cancer who had a retrocaval mass which resembled a metastatic node as shown in the image, which created a suspicion of metastasis. The pathological stage of iGBC was inconclusive due to the inadequate paraffin blocks available hence the patient warranted

revision surgery considering the aggressive nature of Gall bladder cancer (GBC). Revision surgery was performed and the mass turned out to be a benign schwannoma with no other nodal disease involvement. Hence awareness of this differential, though rare is important to avoid erroneous diagnosis and overtreatment.

CASE REPORT

The case has been reported in line with the SCARE criteria.[20] A 31-year-old lady with no medical comorbidities or family history of cancer presented to our clinic with a history of having undergone laparoscopic cholecystectomy elsewhere 4 weeks prior for upper abdominal discomfort.

Outside histopathology report (HPR) revealed poorly differentiated neoplasm with no information on cystic duct margins. Upon further evaluation by CECT scan of thorax, abdomen and pelvis revealed no obvious lesion identified in gallbladder fossa.

Common bile duct was normal and there were no suspicious periportal lymph nodes. However, the scan revealed an enlarged retrocaval node 2.8×2.2 cm size abutting and displacing the Inferior venacava (IVC) at an angle upto 180 degrees and left renal vein (angle upto 270 degrees) with loss of fat planes (Figure 1-2). Posteriorly it seemed to involve Right crura and there was focal involvement of body of Right adrenal gland.

Slide block review at the institute showed high grade dysplasia with foci of invasion as only limited sections were provided. Tumor marker evaluation revealed a Carcinoembryonic antigen (CEA) of 0.959 and Carbohydrate antigen (CA19.9) 3.72. Case was discussed in Multidisciplinary clinic due to the unusual location of metastatic node without any evidence of periportal or Interaortocaval (IAC) node. Node was biopsied which was non-diagnostic with evidence of occasional ganglion cells and on immunohistochemistry (IHC), Pan cytokeratin (PanCK) and CDX2 did not highlight any atypical cells.

Case was rediscussed and considering the young age and the primary diagnosis of iGBC, it was decided to proceed with exploratory laparotomy, biopsy the node and proceed further. Intraoperatively, there was a retrocaval well circumscribed mass adherent to posterior wall of IVC abutting the right adrenal gland, and aorta medially, extending till above the level of left renal vein. Mass was mobilized all around by retracting the IVC and left renal vein. It was lifted off from retroperitoneum with precision and was removed enbloc and was sent for final HPR (Figure 3).

Proceeded with revision cholecystectomy by completing the periportal nodal clearance of stations 8,12,13 and sampling of the IAC node. Cystic duct margin was revised and a small wedge of liver was dissected at the GB fossa. Patient recovered uneventfully in the postoperative period and was discharged on post operative day 5. Her final HPR revealed no evidence of malignancy in the liver wedge. Cystic duct margin was free. There was a total of 26 lymph nodes which were negative for metastasis.

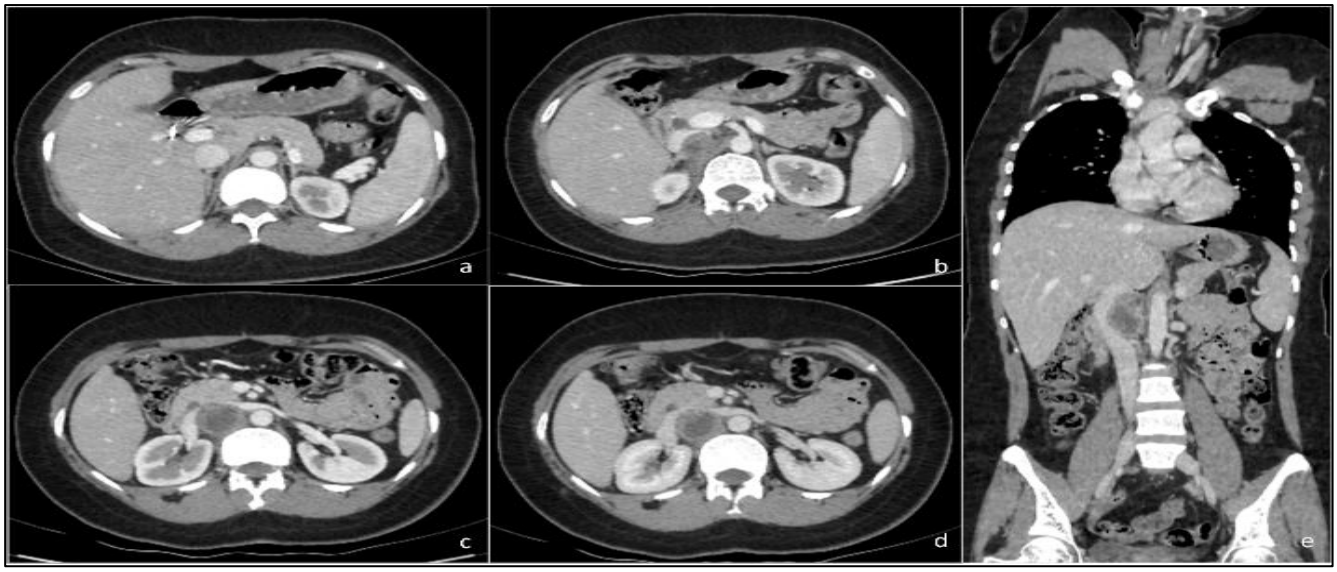

Figure 1: Axial CECT image (a) Gall bladder fossa not showing any significant mass lesion. Surgical clips seen in situ; (b) Origin of the retrocaval mass in close relation to left renal vein. No other significant periportal nodes; (c-d) Relationship of the mass in relation to IVC and aorta; (e) Coronal section showing the mass abutting IVC.

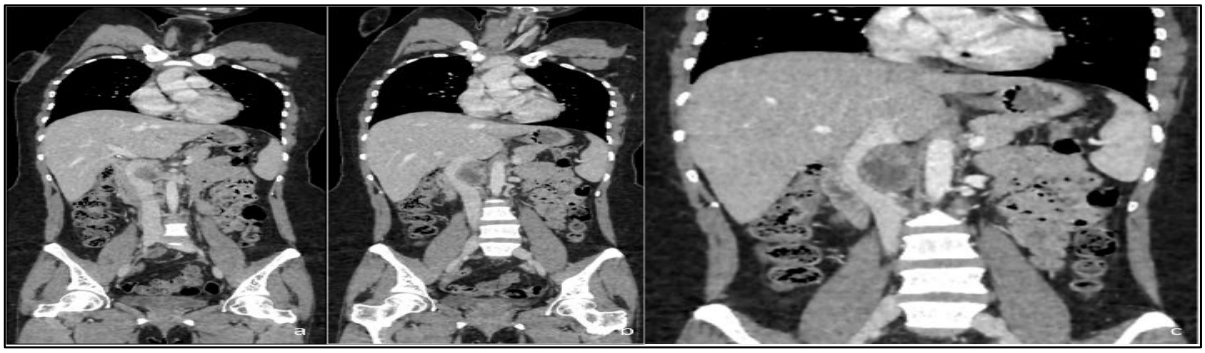

Figure 2: Coronal section CECT images Thorax, abdomen and pelvis; (a) Heterogeneously enhancing retrocaval mass in proximity to drainage of Left renal vein; (b) The relation with Right renal artery at the Interaortocaval region; (c) Enlarged image showing the retrocaval node and its relation with IVC.

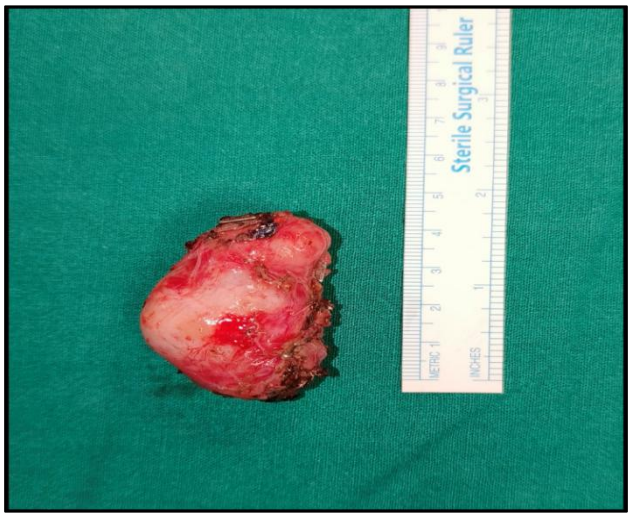

Figure 3: Specimen photograph showing the mass being removed in toto.

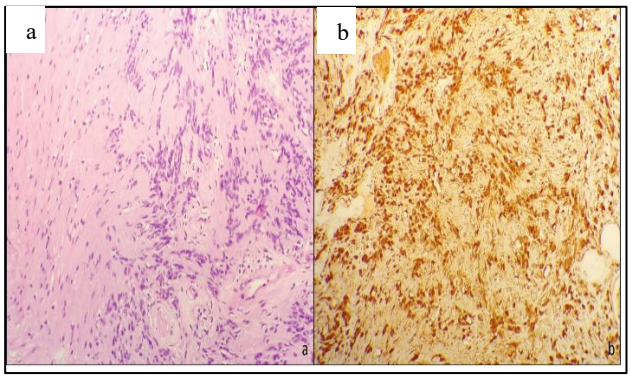

Figure 4: (a) Microscopic image showing Verocay bodies in Antoni A areas (Hematoxylin & Eosin (H&E) stain, 40 X); (b) Immunohistochemistry showing S100 positivity in the tumor cells, (H&E, 40X).

Retrocaval lesion showed an encapsulated lesion composed of spindle cells arranged in loose fascicles with areas of hemorrhage and hyalinization (Figure 4a). No significant nuclear atypia, necrosis or mitosis seen. On IHC, they stained positive for S100 (Figure 4b) while negative for SMA (Smooth muscle antigen) and caldesmon, which favored a primary diagnosis of Benign nerve sheath tumor, favoring Schwannoma. At one month post operative, patient is doing clinically well.

DISCUSSION

Schwannomas or Neurilemmomas, are benign neurogenic tumors that arise from the nerve sheaths of peripheral nerves.[1] They tend to be encapsulated, with the nerve fibres stretched around the tumor. They have a predilection for the head and neck, and extremities.[7] Deeply situated tumors predominate in the retroperitoneum and posterior mediastinum with reported incidences of 0.5-1.2% which makes accurate preoperative diagnosis very difficult.[8] Cystic transformation can occur when the tumor outgrows its blood supply and can mimic necrotic malignant lymph node.[5,6]

Authors report a case of a young lady diagnosed with iGBC with a schwannoma in the retrocaval location resembling a metastatic malignant lymph node creating a diagnostic dilemma. Correlating clinically, it brought suspicion of metastatic nature of the disease and hence was subjected to biopsy in view of the absent periportal lymph node or IAC node. Biopsy however was inconclusive in this regard.

This highlights the importance of improving radiological standards for diagnosis of this neoplasm and in literature, only 15.9% of schwannoma were diagnosed using preoperative imaging modalities.[9] Had the mass been considered as metastatic node without the suspicion of a differential, the patient would have been deemed palliative and the decision would have been futile. Despite the biopsy results being non-corroborative, a decision was made to proceed with revision surgery, considering the patient's young age and aggressive nature of iGBC and the mass was assessed intraoperatively. It was found to be a well circumscribed mass and was successfully resected en bloc and the patient was

surgically cured by doing a completion revision cholecystectomy surgery.

The locations of primary tumor and retroperitoneal masses are important and are useful indicators to differentiate lymph node metastasis from primary tumors of other origins. There have been various theories proposing the lymphatic drainage pattern of gall bladder cancer. Ito et al have proposed three pathways: the cholecystoretropancreatic pathway, the cholecystoceliac pathway and the cholecystomesenteric pathway and hepatic hilar nodes becomes a common pathway.[10,11]

The presence of retrocaval nodal mass, which would be a metastatic station for GBC, created suspicion in the background of absent nodal disease in primary basins. Imaging modalities like CECT can give more anatomical information and relations of the mass with adjacent structures but cannot be 100% conclusive of the diagnosis.[7] It is difficult to identify the peripheral nerve from which retroperitoneal schwannomas develop.[7] Positron emission tomography (PET-CT) would also have not contributed much which was anyway not done in our case.

Surgical excision and histopathological analysis of the resected mass is important to arrive at a final diagnosis in case of Schwannoma. Histologically, Schwannomas usually consist of two different components, designated by the Swedish neurologist Nils Antoni as A and B. Antoni type A areas are highly cellular, composed of spindle cells with often a palisade or organoid arrangement (Figure 4a). In Antoni type B areas, the tumor cells are separated by abundant oedematous fluid that may form cystic spaces.[7,12] Intense staining of S100 is confirmatory (Figure 4b). They usually occur in young to middle-aged adults, with women twice commonly affected than men.[7]

The recurrence rate is very minimal in benign schwannoma while malignant tumors may be associated with poor prognosis.[13] As authors did a thorough literature study, there have been cases of schwannoma being misdiagnosed as metastatic node in cases of seminoma, bladder cancer and ovarian malignancies. They can also mimic adrenal malignancies and rare case of schwannomas occurring in GIT including colon or gallbladder have been reported.[5,6,14-19] No single case of schwannoma resembling a metastatic node combined with a diagnosis of gall bladder cancer could be found in literature.

This highlights the importance of proper preoperative assessment of the patient as a whole and the need for clinical correlation of radiological findings. The clinicians should hence be aware of the retroperitoneal schwannoma and its distinction from metastatic node to avoid misdiagnosis and to ensure proper treatment.

CONCLUSION

Retroperitoneal schwannomas have often been misdiagnosed considering the low prevalence and lack of specific signs and symptoms. The diagnosis is confirmed only by histopathological examination of surgically resected specimen. The treatment of choice is complete surgical excision. And once completely resected, prognosis is extremely good and recurrence is unusual. In the setting of a coexisting iGBC as in our case, it is important to have a clear index of suspicion to make proper clinical judgement and management of the same.

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