

CASE REPORT

NEUROENDOCRINE TUMOR OF PANCREAS MASQUERADING AS SPLENIC ANGIOSARCOMA: A DIAGNOSTIC DILEMMA

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HOW TO CITE THIS ARTICLE:

Vijaya C, Shilpa G, Lekha M.B, Gurappa V. P, Archana Shetty. "Neuroendocrine tumor of Pancreas Masquerading as Splenic Angiosarcoma: A diagnostic dilemma". Journal of Evolution of Medical and Dental Sciences 2013; Vol. 2, Issue 51, December 23; Page: 10036-10040.

ABSTRACT: Splenic masses are quite rare and spleen is an infrequent site for primary and secondary malignancies. The most common primary tumour is angiosarcoma. We present a case of 48 yr old female who presented with pain and fullness in left hypochondriac region. Both ultrasound(US) and computed tomography(CT) diagnosed highly vascular splenic neoplasm with a possibility of angiosarcoma. Splenectomy was done while doing exploratory laparotomy. On histopathological examination it was diagnosed as neuroendocrine tumour of pancreas with splenic involvement which was confirmed by immunohistochemistry. We are reporting this interesting case of neuroendocrine tumor of pancreas which masqueraded as splenic angiosarcoma on imaging study.

KEY WORDS: Neuroendocrine tumor of pancreas, angiosarcoma spleen, spleen metastasis.

INTRODUCTION: Splenic involvement in neuroendocrine pancreatic tumors is well known but it rarely presents as primary splenic mass. The distal portion of the pancreatic tail extends along the course of the splenic vessels and enters into the hilum of the spleen contained within the splenorenal ligament. As a consequence of these anatomical relationships, the spleen and its vasculature may be involved in disorders of the pancreas. We report a rare case of pancreatic neuroendocrine tumor masquerading as splenic angiosarcoma.

CASE REPORT: A 48 year old female patient presented with pain and fullness in left hypochondriac region. She also gave history of loss of weight and fatigue. The patient was not an alcoholic. Past medical history suggested anemia four years back for which she received treatment. Her vital signs on admission were stable, however on per abdomen examination splenomegaly was detected. The rest of the systemic examination was normal.

Ultrasound of the abdomen showed a heterogenous mass measuring 7.1 x 6.7cms in the spleen, with marked vascularity, possibly neoplastic origin.

Hence a CT scan was advised, which revealed an enlarged spleen with an ill-defined heterogenous enhancing mass lesion in the inferior aspect measuring 8.5 x 8.3 x 6.6cms. Areas of breakdown and calcifications were noted within. No air-fluid level was seen but perilesional fat stranding was noted. Superiorly, it was extending upto the splenic hilum. Inferiorly, it was displacing the left kidney with maintained fat planes. Anterosuperiorly, it was displacing the splenic vessels. Anteroinferiorly, margins were irregular and fuzzy, with displacement of adjacent peritoneal vessels. Superomedially, it was abutting and displacing splenic vessels with infiltration of tail of pancreas. Inferomedially, margins were fuzzy and abutting the collapsed small bowel loops. Posteromedially, the fat plane between the lesion and psoas muscle was maintained. Superolaterally, was limited to the spleen. Inferolaterally, margins were ill defined, fuzzy and abutting the splenic flexure of the colon. There was no evidence of abdominal lymphadenopathy or distant metastasis.

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These features favoured the possibility of malignant solid splenic lesions suggestive of angiosarcoma [Fig-1- CT abdomen showing the splenic mass]. Routine haematological investigations were within normal range, except for low platelet count of 80,000/cumm. Renal and liver function tests were within normal limits.

Surgical intervention was indicated and exploratory laprotomy was performed. Intraoperatively a tumor measuring 8.5 x 8.0 x 6.0cms was seen mainly at the hilum of spleen in close proximity to the tail of the pancreas. A distal pancreatectomy and splenectomy was done. The post-operative course was uneventful.

Gross appearance: Received splenectomy and distal pancreatectomy specimen. Splenectomy specimen showed a yellowish, firm, circumscribed, solid mass measuring 8.5x8.2x6.5cms at the hilum, with infiltrating borders, areas of necrosis and hemorrhage were seen in the tumor [Fig-2 Cut section of the spleen showing a solid mass with infiltrating borders].

Microscopy: Multiple sections from the mass showed tumor composed of uniform cells, arranged in trabecular and glandular pattern with round to oval nucleus, stippled chromatin and moderate amount of cytoplasm [Fig-3 H and E stain showing tumor cells with neuroendocrine features]. The stroma was dense and hyalinized, with area of necrosis and hemorrhage. These tumor cells were seen infiltrating the splenic parenchyma [Fig-4 H and E showing the tumor infiltrating the splenic parenchyma]. Rest of the spleen showed areas of congestion. Immunohistochemistry was done which showed neoplastic cells expressing synaptophysin and chromogranin [Fig 5- IHC of Tumour cells showing chromogranin positivity] and negative for insulin, gastrin and somatostatin. The final report was given as Nonfunctioning Malignant Neuroendocrine carcinoma of the Pancreas with Splenic infiltration, stage T4 (according to TNM classification).

Post-operative period was uneventful and was discharged nine days after the operation. The platelet count returned to normal (1.9 lakh/cumm). Presently the patient is asymptomatic with no evidence of recurrence two years after surgery.

DISCUSSION: Neuroendocrine tumors (NETs) of the gastro-entero-pancreatic (GEP) system are rare and originate from the diffuse endocrine system, located in the gastro-intestinal tract (GIT) and in the pancreas, with extremely varying clinical pictures¹. They comprise 2% of the tumors of the gastrointestinal tract and 1-2% of all pancreatic tumors². The clinical features of gastro-entero-pancreatic neuroendocrine tumors are very heterogeneous. They may be asymptomatic for years, or present with abdominal pain, nausea, vomiting or cholestasis¹. The most recent WHO classification³ categorized all gastro-entero-pancreatic neuroendocrine tumors on the basis of clinical-pathological criteria as follows: (a) well-differentiated endocrine tumours, with benign or uncertain behaviour; (b) well-differentiated endocrine carcinomas, with a low-grade malignant behaviour; (c) poorly differentiated endocrine carcinomas (small cells carcinomas), with a high-grade malignant behaviour; (d) mixed endocrine-exocrine carcinomas, with characteristics of both endocrine and exocrine tumours. Each category includes functioning and non-functioning tumors. The pathological criteria of malignancy are lymph node invasion, distant metastasis or invasion to adjacent organs. Metastatic disease can be present at the time of diagnosis in approximately 50% of the cases⁴. Invasion of the spleen represented metastasis in our case. Due to the vagueness of the symptoms,

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diagnosis can be delayed. In general, malignant metastatic pancreatic endocrine tumors are slow growing neoplasms. Survival from the time of diagnosis usually ranges from two-ten years⁵.

Usually, neuroendocrine tumors involving the spleen can represent direct invasion from adjoining organs such as the pancreas but Pancreatic neuroendocrine tumour presenting as a primary splenic mass is exceptionally rare^{6,7}. Imaging has an important role in localizing the primary tumor and identifying metastatic sites. Investigations include CT and MRI, and newer modalities, like endosonography and somatostatin receptor scintigraphy which are highly sensitive for detecting carcinoids presenting 80–100% sensitivity⁸.

Angiosarcoma is a malignancy of vascular origin, and is characterized by masses of endothelial cells with cellular atypia and anaplasia⁸. Angiosarcoma may occur anywhere in the body, but most often in the skin, soft tissues, breast and liver. Splenic involvement is exceedingly rare. Clinical manifestations include abdominal discomforts, splenomegaly and signs of Immune thrombocytopenic purpura⁹, which are not specific to the disease. Pathogenesis remains obscure. Prognosis is extremely poor, with a six-month survival rate of 20%¹⁰. The tumor commonly metastasizes to the liver, lung, bone, lymph nodes, omentum, or peritoneum.

The uniqueness of this case was the presentation of neuroendocrine tumor as a splenic mass with radiological imaging favoring splenic angiosarcoma. In our case, presenting complaints were not suggestive of pancreatic neuroendocrine tumor and imaging features were suggestive of primary splenic mass probably non lymphoreticular malignancy angiosarcoma. The reason why it could mimic the picture of angiosarcoma could be explained by hypervascular component within the tumor, which absorbed the contrast medium in radiological examination. The age, clinical and operative findings of the mass were going in favor of angiosarcoma, but its non-aggressive nature went against it. However, an overreliance on radiological diagnosis led to managing the tumor mass as primary splenic angiosarcoma until histopathology examination revealed a pancreatic neuroendocrine tumor.

CONCLUSION: Neuroendocrine tumor of the tail of the pancreas should be kept in mind in the differential diagnosis of splenic mass and thus be evaluated for the same, since it is rare and has different biological behavior.

ACKNOWLEDGMENTS: Dr Mahesh, Dr Reehan, Department of Surgery & Dr Narayan, Dr Prakash Jain, Department of Radiology SIMS & RC.

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Fig. 1: CT abdomen showing a heterogeneous mass in left hypochondrium in close relation to tail of pancreas infiltrating splenic parenchyma.



Fig. 2: Cut section of spleen showing infiltrating yellowish, solid tumor with compressed splenic tissue.

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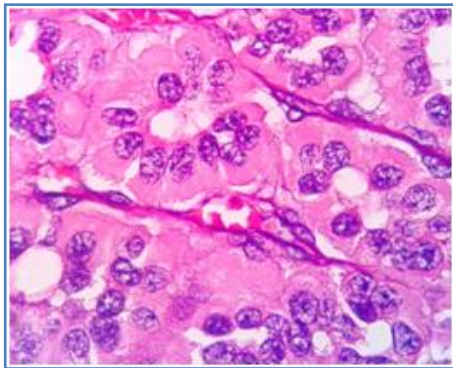


Fig. 3: 40X H&E Uniform tumor cells arranged in glandular pattern showing stippled chromatin.

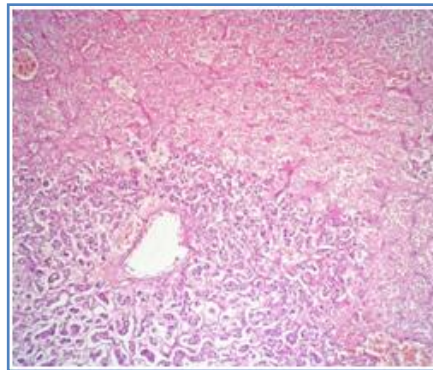


Fig. 4: 10x H&E Tumor cells infiltrating spleen

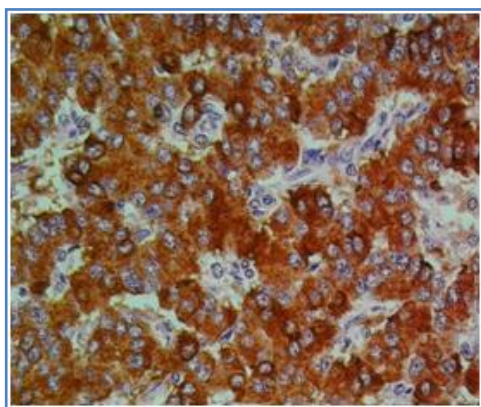


Fig. 5: Immunoreactivity of chromogranin within the tumor cells

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Date of Submission: 20/11/2013.
Date of Peer Review: 21/11/2013.
Date of Acceptance: 06/12/2013.
Date of Publishing: 19/12/2013