

Primary pancreatic carcinoid tumour

Sir,

Primary pancreatic carcinoid tumour is extremely rare pancreatic neoplasm constituting only 0.5% of all carcinoid tumors. To date, only 46 cases have been reported in literature.^[1] Given the rarity of these neoplasm coupled with advanced and non-specific presentation, recognition of these neoplasm has immense therapeutic and prognostic implications. We report a case of primary pancreatic carcinoid with unusual clinical presentation and review the literature.

A 67-year-old male patient presented to us with dull aching pain in epigastrium and right lumbar region for last six months. His local and systemic examination revealed no other abnormalities except for mild hypertension. His complete blood count and blood biochemistry was normal. Chest X-ray showed cardiomegaly and echocardiography revealed left ventricle concentric hypertrophy. USG abdomen showed large right retroperitoneal tumor with heterogeneous echotexture measuring $7.8 \times 6.7 \times 10.2$ cm in close approximation to head of pancreas and right kidney. CT scan whole abdomen showed large complex well encapsulated cystic SOL with septation and solid enhancing nodular areas, measuring $7.54 \times 6.55 \times 10.0$ cm at the right side of retroperitoneum with loss of fat planes with adjacent right kidney and head of pancreas [Figure 1].

He underwent complete excision of retroperitoneal tumor with lymphadenectomy from crus of diaphragm to the right renal hilum. The tumor was mostly cystic and adhered to pancreatic head. Tissue planes with adjacent structures such as second part of duodenum and distal CBD were maintained. Histopathology revealed tumor composed of uniform cells with round or oval nuclei and eosinophilic cytoplasm arranged in cords and in trabecular fashion suggesting well differentiated neuroendocrine tumor of pancreas [Figure 2]. Immunohistochemistry showed tumor cell expressing synaptophysin and chromogranin, in favor of well-differentiated carcinoid tumor of pancreas. He had an uneventful post-operative period. Follow-up USG whole abdomen after six months showed no evidence of recurrence.

Primary pancreatic carcinoid tumor is a rare neuroendocrine tumor originating from enterochromaffin

cells and excreting serotonin derivatives. Carcinoid tumors most commonly arise in the midgut organs, less commonly in hindgut organs and rarely in organs derived from embryonic foregut. The colonic carcinoids exhibit the maximum potential to metastasize with metastasis occurring in 60-70% of the tumors. In contrast although appendix constitute the most commonest site of carcinoid, only 2-3% of appendicular carcinoid have metastasis at presentation. Carcinoid tumors commonly metastasize to regional lymph nodes and liver. In the classic histological terminology, carcinoid lesions are widely regarded as malignant neoplasm.^[2] Incidence of metastasis in primary pancreatic carcinoid was about 72% and about 20% of patients presented with carcinoid syndrome.^[3]



Figure 1: CT scan section showing large complex well encapsulated cystic SOL with septation and solid enhancing nodular areas at right side of retroperitoneum with loss of fat planes with adjacent right kidney and head of pancreas

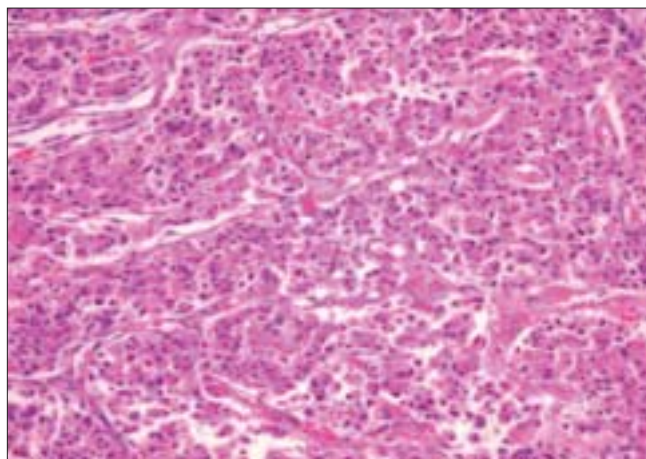


Figure 2: Photomicrograph of a histological section of the excised pancreatic tumor showing monomorphic tumor cells in an organoid pattern characterized by nests and sheets, consistent with a carcinoid tumor (H&E, $\times 200$)

However there are no precise histological criteria to distinguish benign from malignant carcinoid or carcinoid with metastatic potential. Cure for this disease is directly related to an early diagnosis. But as the tumor is often asymptomatic or has non-specific clinical manifestation, as in our case, the diagnosis and treatment is often delayed. Usually these tumors become symptomatic only when hepatic metastasis occurs, which manifest as classical carcinoid syndrome. However absence of the characteristic carcinoid syndrome does not exclude the diagnosis of carcinoid tumor even in patient with liver metastasis.^[4] In most of the reported cases of primary Pancreatic carcinoid, the tumor was located in the tail or the body of Pancreas,^[5] but in our case it was located in the head of pancreas. Although rare, one should be aware of existence of pancreatic carcinoid tumor. For tumors limited to pancreas with no evidence of distant metastasis, a radical resection with lymphadenectomy may be curative.^[6]

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