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Review

Pancreatic plasmacytoma: A rare but important entity for gastroenterologists, oncologists and hematologists

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ABSTRACT

Multiple myeloma (MM) is a plasma cell neoplasia and accounts 10% of the hemopoietic malignancies with mean age of 55 years and 5% of these disorders have extramedullary (EM) disease named as plasmacytomas (Pm). Pm is a solitary tumor of plasma cells and may be primary as solitary masses without bone marrow (BM) involvement or may accompany MM [1–4]. Upper respiratory tract is most common site for EM involvement (80–90%). All the body sites including gastrointestinal tract (about 10%), genitourinary tract, reticuloendothelial system, thyroid, lungs, skin, and testicles can also be involved [4,5]. First case with pancreatic Pm has been reported by Hefferman in 1947.[6] Here pancreatic Pm has been reviewed; clinical, diagnostic prognostic and therapeutic approaches have been discussed.

1. Incidence

Pancreatic involvement is usually the result of a secondary involvement in cases with MM.[7] Relatively limited numbers of pancreatic Pm have been reported. In autopsy studies 2.3% incidence has been reported.[7,8] Primary pancreatic Pm is seen less than 0.1% of all pancreatic tumors.[6,9,10]

2. Age-sex

Mean age is 55 similar to MM and in a study covering 63 cases mean age has been found as 58.5.[10,11] Pancreatic Pms are usually seen in relatively older patients but have been reported in patients as young as 32 years of age.[12,13] There is male predominance as high as 3–5 fold as in other Pms.[5,14] Most commonly focal involvement has been reported but diffuse infiltration may be seen.[2–4,15,16]

3. Clinical presentation

Pancreatic Pm may accompany underlying MM, may precede the diagnosis of MM or may be the recurrence sign of MM. Patients with pancreatic Pm commonly present with obstructive jaundice. The most commonly involved site of presentation is the head of pancreas. For this reason the most common clinical finding is obstructive jaundice and abdominal pain[1–5,7,17–22]. First presentation has been found as jaundice in 70% of the cases in a large review.[11] Other symptoms are weight loss, asthenia, and anorexia.[5] Left-sided portal hypertension (left-sided, sinistral, segmental portal hypertension (LSPH) with bleeding in the upper gastrointestinal tract has been reported in a case.[23] Severe upper gastrointestinal bleeding including life-threatening variceal bleeding may be the first sign.[21,24] Diffuse enlargement or a mass in the tail of the pancreas have also been reported[16,25,26] and cases involving the body of the pancreas have been reported.[1,5,13] Abdominal vessel involvement has been reported from autopsy findings.[13,27–29]

In some cases pancreatic Pm may accompany to other sites of Pms. A case of concurrent gastric and pancreatic Pm has been reported in a case.[30] Clinical presentation mimicking acute pancreatitis has been reported in a 71 year old patient.[11] Jejunal involvement in addition to pancreas has been reported in an another case.[26] In one case pancreatic Pm has been reported 5 years after maxillary Pm as second focus.[13]

4. Radiological findings

Radiological findings are not specific. In cases with focal involvement the solid mass may be homogeneous, heterogeneous or multilobulated with variable enhancement.[1,31]

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Ultrasonographically most of the lesions have been reported as hypoechoic focal masses with low-level echoes.[7,8,21,32]

At computerized tomography (CT), lesions have been defined as well-defined, lobulated, homogeneous soft-tissue masses and are usually hypo-attenuating compared to normal pancreatic tissue. The most typical finding of pancreatic plasmacytoma on CT has been reported as the presence of a focal multilobulated mass with homogeneous contrast enhancement.[1,7,8,16,17,19] Diffuse infiltration of the pancreas has been described in only a few cases.[16,33]

At MRI lesions are hypointense on T1-weighted images and hyperintense on T2-weighted images, compared to normal pancreatic tissue.[34] MR cholangio-pancreatography is very sensitive method for the detection of pancreatic Pm as in other biliary and pancreatic ductal obstructive lesions.[7,8,20,26,30]

In cases with diffuse presentation there is diffuse volumetric enlargement of the pancreas with lobulated contours and predominantly homogenous uptake in the portal phase.[16,35]

In recent times, the use of F^{18} PET/CT has been recommended in patients with MM and also for Pm. This technology provides morphologic evaluation by CT and metabolic tumor activity by PET and also predictive for response. Moderate to intense 18F-FDG uptake is seen as in other plasma cell neoplasias.[1,17,36,37]

The radiological differentiation of pancreatic Pm from other pancreatic tumors including poorly differentiated pancreatic neoplasm, lymphoma, and metastasis is difficult.[5,11,38] Pancreatic Pm must be thought in differential diagnosis of pancreas masses.

5. Laboratory findings

There is no specific laboratory finding in cases with pancreatic Pm. Elevated transaminases may be the first sign of pancreatic Pm in a case with MM.[2] Slightly elevated pancreatic enzymes may be the first sign and initial working diagnosis may be acute pancreatitis.[8] However increased level of lipase above 2000 UI/L accompanying cholestatic icterus and elevated liver function tests have been reported.[11] High levels of leukocytes, C-reactive protein and very high levels of serum amylase (921IU/l) have been reported.[13] Tumour markers including CA125, CA199, CEA, and AFP generally have been found within normal limits.[18] In cases with normal tumor markers in a case with pancreatic mass hemopoietic neoplasms including Pm, extramedullary myeloid tumor and lymphoma must be considered.[39]

6. Diagnosis

Biopsy is the standard of care in cases with pancreatic Pm. atypical plasma cells, which are positively immunostained for CD79a, CD138, and the k light chain are helpful for differential diagnosis in a case with pancreatic mass. Most cases of plasma cell infiltration of the pancreas are microscopic, and well-formed masses are unusual.[5]

The route of biopsy may be percutaneous, endoscopic, or surgical.[14] Endoscopic ultrasound fine-needle aspiration (EUS-FNA) is a fast and reliable technique as high as 70–90% for diagnosis.[11,17,40] However, experience of gastroenterologist is very important for EUS and its value varies and also has risks of pancreatitis, bleeding, and perforation.[7,18,41]

About one third of the cases have been diagnosed by EUS-guided FNA. However EUS-guided FNA may be non diagnostic and ascites cytology showing malignant plasma cells may be diagnostic.[11] EUS-FNA has been found to be safe regarding the risk of pancreatitis, bleeding, and perforation. Major complications in 355 cases performed EUS-FNA for solid pancreatic tumors have been found as 2.5%. The most common complications have been found as acute pancreatitis, infection, but complication has been found to be decreased in recent years.[42,43]

The other problem is seeding of the tumor cells during EUS-FNA but it has been reported as a rare event and it has been found as 2.2% in cases with pancreatic adenocarcinoma performed EUS-FNA. So far seeding of a pancreatic Pm during EUS-FNA is controversial. It has been proposed as a very safe method and not cause to seeding but has been reported. However this rate is very low as compared with CT or transabdominal ultrasonography-guided percutaneous biopsy (16.3%).[5,8,19,38,44,45]

About one fifth of the cases have been diagnosed by CT guided percutaneous FNA and about one forth of the cases requires surgical biopsy (Williet N et al., 2017). In some cases these imaging modalities may not show a mass lesion and CT may show significant but unspecific infiltration around the pancreas head, without dilatation of biliary ducts. EUS may not be useful in these cases.[11]

Pancreatic Pm should be considered in the differential diagnosis of a patient with multiple myeloma and a pancreatic mass. An interesting presentation may simulate pancreas neuro-endocrine tumor and differential diagnosis is very important in these cases.[1,2]

7. Treatment

There appears to be no standardized treatment for extramedullary Pm of the pancreas. Pm is sensitive both to radiation and chemotherapy. The combination of local radiation, chemotherapy, and, in selected cases surgery is necessary for diagnosis and treatment.[4] Treatment with surgery, chemotherapy or radiotherapy response rate in 63 cases and in other cases reported from different sites have been reported as 90–100%. High dose chemotherapy with stem cell transplantation is important in cases with secondary pancreatic Pm. Biliary stenting is another choice in some cases.[1,5,7,11,13,20]

8. Prognosis

Approximately 70% of patients with isolated pancreatic Pm and these cases remain disease free at 10 years. Regional recurrence has been reported in up to 25% of patients [7,46,47]. The course is more favorable in cases with primary Pm than that of that of Pm accompanying MM or solitary Pm the bone.[18]

Declaration of competing interest

There is no conflict of interest.

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