OLGU SUNUMU CASE REPORT

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Intrapancreatic Accessory Spleen Diagnosed by EUS/FNA Cytology: One Can Only Perceive What the Mind is Prepared to Comprehend

EUS/İİA Sitolojisi ile Tanı Almış İntrapankreatik Aksesuar Dalak: Zihin Ancak Kavramaya Hazır Olduğunu Algılayabilir

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ABSTRACT Intrapancreatic accessory spleen is a relatively uncommon congenital lesion, with a prevalence of 2%; reported in an autopsy series. Radiological differential diagnosis for intra and peripancreatic accessory spleen includes pancreatic neuroendocrine tumors, lymphoepithelial cysts, lymphomas, hypervascular metastases, solid pseudopapillary tumors and pancreatic adenocarcinomas. Accessory spleen is a clinically quiescent entity which is usually asymptomatic. It does not require surgical resection or extra follow up. In a patient with an intra-peripancreatic mass, in order to prevent invasive procedures such as unnecessary surgical resections and to decrease patient morbidity and mortality rates, the endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) cytological diagnosis is mandatory. As this is a very rare entity in routine practice, the cytopathologists should keep it in mind, in the differential diagnoses of a pancreatic mass. In this report, we present a rare case of intrapancreatic accessory spleen which is diagnosed by EUS /FNA cytology, discussing its unique cytomorphological and immunohistochemical characteristics.

Keywords: Spleen; pancreas; endoscopic ultrasound-guided fine needle aspiration; cytology

ÖZET Intrapankreatik aksesuar dalak göreceli olarak nadir görülen bir konjenital lezyondur. Prevalansı yayınlanmış otopsi serilerinde %2 olarak tanımlanmıştır. İntra ve peripankreatik aksesuar dalağın radyolojik ayırıcı tanısında nöroendokrin tümörler, lenfoepitelyal kistler, lenfomalar, hipervasküler metastazlar, solid psödopapiller tümör ve pankreatik adenokarsinom bulunmaktadır. Aksesuar dalak klinik olarak asemptomatik seyreden bir antitedir. Cerrahi rezeksiyon ya da ek takip gerektirmemektedir. İntra ya da peripankreatik kitle ile prezente olan bir hastada, gereksiz cerrahi rezeksiyonlar gibi invaziv yöntemleri önlemek, hasta morbidite ve mortalitesini azaltmak amacı ile, endoskopik ultrason eşliğinde ince iğne aspirasyonu (EUS/İİA) sitolojisi ile tanı koymak şarttır. Rutin pratikte çok nadir görülen bir antite olsa da, aksesuar dalak pankreas kitlelerinin ayırıcı tanısında akılda tutulmalıdır. Bu olgu sunumunda EUS/İİA sitolojisi ile tanı almış intrapankreatik aksesuar dalak olgusunu sunmakta ve özgün sitomorfolojik ve immünhistokimyasal özelliklerini tartışmayı amaçladık.

Anahtar Kelimeler: Dalak; pankreas; endoskopik ultrason eşliğinde ince iğne aspirasyonu; sitoloji

Accessory spleen (AS) is a congenital developmental anomaly which is formed as a result of an insufficiency in splenic precursor mesenchymal cell fusion. It occurs in 10-30% of the population.[1,2] AS is mostly seen in the splenic hilus.[2,3] Intrapancreatic AS (IPAS) is a relatively rare entity with a prevalance of 2%, reported in autopsy series.[1] AS is a clinically quiescent entity which is usually asyptomatic and does not require follow up or resection. Thanks to the current advanced radiological tech-

niques, the diagnosis of an AS may be achieved radiologically but the most accurate and reliable method for achieving a diagnosis is the direct sampling of the lesion. In the literature, half of the reported cases have had the diagnosis of an AS after surgical resection.[1,3,4]

Pancreatic endocrine neoplasies (PEN), lymphoepithelial cysts, lymphoma, hypervascular metastasis, solid pseudopapillary tumor and pancreatic adenocarcinoma are included in the differential diagnoses of intra and peripancreatic AS.[3,4]

Eighty percent of the reported IPAS cases have had the radiological diagnosis of PEN.[1,5,6] The typical cytological characteristic of AS is the presence of small lymphocytes representing the white pulpa. Mixed inflammatory cells admixed with the small lymphocytes, endothelial cells and histiocytes are the other components of AS.[5,7] The sinusoidal endothelial cells are immunoreactant with CD8 immunocytochemical antibody and this feature is specific for the diagnosis of accessory spleen. They are better observed in the cell block sections.[1,3,5,7]

CASE REPORT

A 37 year old male patient arrived at the outpatient clinic with complaints of abdominal discomfort. An abdomial ultrasonography was performed revealing a hypoechoic area without nodular configuration, located at the pancreatic tail. An endoscopic ultrasonography (EUS) was performed and a hypoechoic space occupying mass of 26x23 mm, located at the pancreatic tail was detected. It lacked clear demarcating borders with the pancreas. Upon doppler US, the lesion was observed next to the splenic artery and vein, displaying no vascular flow or invasion to the neighboring tissues (Figure 1a). The remaining pancreatic parencyma was homogenous and normoechoic. The lesion had the preliminary radiologic diagnosis of PEN. EUS guided fine needle aspiration (EUS-FNA) was performed revealing a soft pancreatic lesion.

During the on-site adequacy evaluation, polymorphic lymphocytes including small, mature lymphocytes and lymphocytes with blastic morphology were detected. Smear slides were prepared,

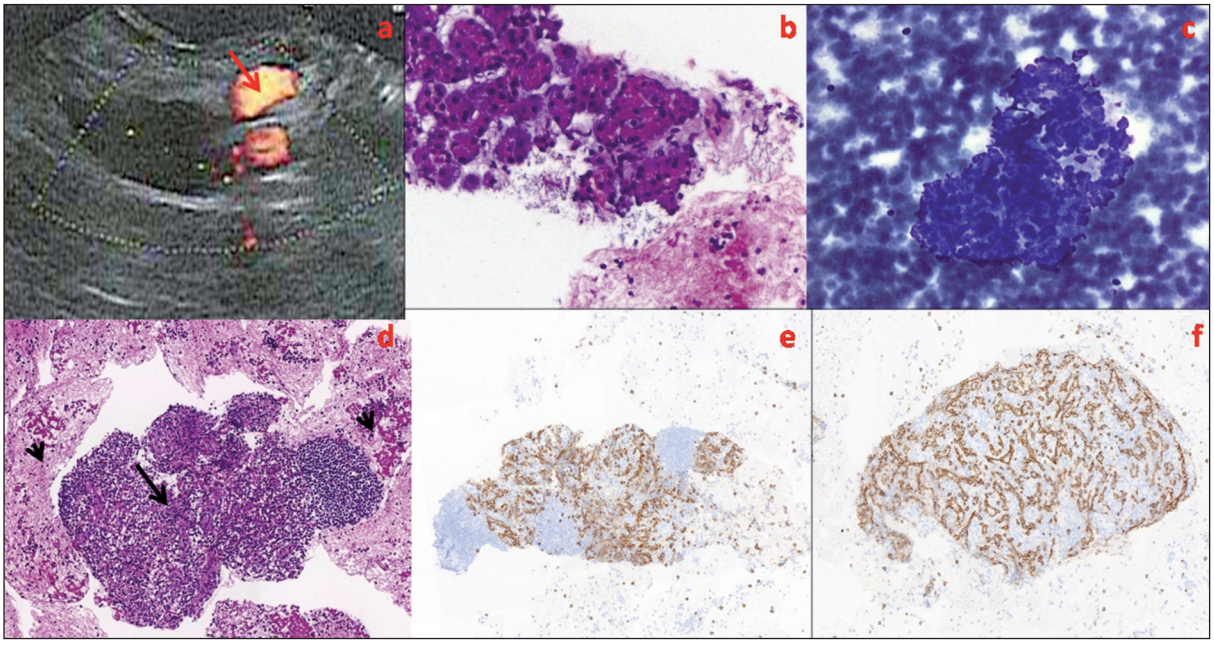

FIGURE 1: a) A lesion located at the pancreatic tail with a longitudinal diameter of 26 mm. Vascular flows from splenic artery and vein (arrow). **b)** Pancreatic acini, blood components, and lyphocytes (H&E, 200x). **c)** Crushed cells with lymphoid morphology, with inexplicit cytologic details on a hemorrhagic background (MGG, 200x). **d)** Splenic white pulp (arrow) and red pulp (arrowheads) demonstrated on the cell block section (H&E, 20 x). **e)** The sinusoidal structures showed positive immunoreaction with anti-CD8 antibody. There was no reaction in the white pulp (immunohistochemistry, 100x). **f)** The sinusoidal endothelial cells showed positive immunoreaction with anti-CD8 antibody, higher magnification (immunohistochemistry, 200x).

air-dried at room temperature and stained with May-Grunwald Giemsa (MGG). A second aspiration was suggested and a cell block was prepared. On MGG slides, sparse mature, small lymphocytes, capillary fragments, histiocytes and pancreatic acini were observed. Accompanying cohesive cellular clusters whose cytological details were not clearly identified were also observed (Figure 1b,c). The predominating cells observed on the cell block sections were lymphocytes while some areas contained histiocytes (Figure 1d).

PEN, high-grade small cell neuroendocrine neoplasm and AS were included in the differential diagnosis.

Pancytokeratin, chromogranin-A, synaptophysin and CD8 immunohistochemical antibodies (Ventana-Roche, USA) were performed on the cell block sections. Isolated lymphocytes and sinusoidal endothelial cells showed positive immunoreaction with anti-CD8 antibody (Figure 1e, f). There was a negative immunoreaction with pancytokeratin, chromogranin-A, and synaptophysin. Upon these cytomorphological and immunohistochemical findings, the case had the diagnosis of IPAS.

Informed consent was obtained from the patient.

DISCUSSION

In the literature, it is stated that intra and peripancreatic AS may mimic various neoplasms radiologically; and this may lead to unnecessary surgical resections.[2,3,8,9] Besides its radiological diagnostic pitfalls, intra or peripancreatic AS also has several cytological diagnostic pitfalls. The most important one is not considering this entity in the differential diagnosis. Arkadopoulos N, et al stated that a case which had the preliminary radiological diagnosis of PEN by MRI and EUS was misdiagnosed cytologically because the cells showed positive immunoreaction by anti-chromogranin-A, anti-glucagon, anti-gastrin, anti-somatostatin anti bodies. The histopathological evaluation of the resection material revealed an IPAS. The non-neoplastic endocrine cells aspirated from the pancreatic islets had shown positive immunoreaction with endocrine markers, leading the cytopatholgist to the misdiagnosis.[3]

In a patient with an intra or peripancreatic mass, in order to prevent invasive procedures such as unnecessary surgical resections and to decrease patient morbidity and mortality rates, the EUS-FNAC diagnosis of the lesion is mandatory.[1] Therefore, when dealing with a case with radiological preliminary diagnosis of PEN, cytopathologists should keep in mind the entity of AS, especially when they see polimorphic lymphoid cells, histiocytes and vascular fragments. In the literature, it is stated that the sinusoidal endothelial cells displaying positive immunoreaction with anti-CD8 antibody is specific for AS. Endothelial cells from systemic vasculature or hemangiomas show negative immunoreaction with anti-CD8.[5]

In an EUS-FNAC, if the material is entirely composed of lymphocytes and does not include any other splenic components, lymphoproliferative disorders should also be included in the differential diagnosis. A seperate aspiration should be advised to obtain extra material for flowcytometry and/or immunohistochemistry.[2]

In this case, there was a little number of polimorphic lymphoid cells on the smear slides. There were no findings of a high grade lymphoma or Hodgkin's lymphoma. Immunohistochemistry revealed negative reaction with pancytokeratin, chromogranin-A and synaptophysin and the Ki-67 proliferation index was <%1.

The lymphocyte groups and histiocytes observed on the slide sections of the cell block represented the white and red pulps, respectively. The most specific finding for the diagnosis of an AS were the sinusoidal structures, positive with anti-CD8 antibody.

In cases with preliminary diagnosis of PEN at EUS, if the on-site evaluation is not possible, the cytopathologic report should imply the possibility of an AS. The cytopathologists' recommendation of a repeat EUS FNA could be effective in the preclusion of unnecessary surgical resection.

When the lymphocytes are the predominating cells observed in EUS-FNA of a pancreatic or a peripancreatic lesion, the differential diagnosis of an AS should be kept in mind and the material should not be evaluated as "inadequate" during on-site evaluation. The triage of the cytologic material should be correctly performed since a cell block preperation for immunohistochemical analysis is mandatory for achieving an accurate diagnosis.

As one cannot perceive what the mind is not prepared to comprehend, the cytopathologists should keep in mind the rare entity of IPAS when dealing with cases with a preliminary radiologic diagnosis of PEN.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Payam Hacısalihoğlu, Davut Şahin, Yıldıran Songür;* ***Design:*** *Davut Şahin, Payam Hacısalihoğlu, Yıldıran Songür;* ***Control/Supervision:*** *Payam Hacısalihoğlu, Davut Şahin, Yıldıran Songür;* ***Data Collection and/or Processing:*** *Payam Hacısalihoğlu, Davut Şahin, Yıldıran Songür;* ***Analysis and/or Interpretation:*** *Davut Şahin, Payam Hacısalihoğlu, Yıldıran Songür;* ***Literature Review:*** *Davut Şahin;* ***Writing the Article:*** *Payam Hacısalihoğlu, Davut Şahin;* ***Critical Review:*** *Payam Hacısalihoğlu, Davut Şahin;* ***References and Fundings:*** *Davut Şahin, Payam Hacısalihoğlu;* ***Materials:*** *Davut Şahin.*

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