Sarcoidosis associated with pseudopapillary pancreatic tumor

Elhadidy Tamer, Morsy Nesreen Elsayed, Abdelwahab Heba Wagih, Refky Basel, Zalata Khaled

ABSTRACT

Introduction: The sarcoidosis is an idiopathic multisystem inflammatory disease characterized by the presence of non-caseating granulomas in the affected organs. A clear association between sarcoidosis and malignancies has been reported. Cancer can occur in patients with an established diagnosis of sarcoidosis and sarcoidosis can subsequently develop in a cancer patient. Malignancy can also be associated with the occurrence of sarcoïd reactions.

Case Report: We report the case of sarcoidosis/sarcoïd-like reaction associated with pseudopapillary pancreatic tumor.

Conclusion: This case report emphasizes the need to add sarcoidosis in the differential diagnosis of lung lesions associated with pancreatic tumors.
Sarcoidosis associated with pseudopapillary pancreatic tumor

Elhadidy Tamer, Morsy Nesreen Elsayed, Abdelwahab Heba Wagih, Refky Basel, Zalata Khaled

ABSTRACT

Introduction: The sarcoidosis is an idiopathic multisystem inflammatory disease characterized by the presence of non-caseating granulomas in the affected organs. A clear association between sarcoidosis and malignancies has been reported. Cancer can occur in patients with an established diagnosis of sarcoidosis and sarcoidosis can subsequently develop in a cancer patient. Malignancy can also be associated with the occurrence of sarcoid reactions. Case Report: We report the case of sarcoidosis/sarcoid-like reaction associated with pseudopapillary pancreatic tumor. Conclusion: This case report emphasizes the need to add sarcoidosis in the differential diagnosis of lung lesions associated with pancreatic tumors.

Keywords: Pancreatic Neoplasms, Pseudopapillary neoplasm, Pseudopapillary neoplasm, Sarcoidosis

INTRODUCTION

Solid pseudopapillary neoplasia of the pancreas is an extremely rare epithelial tumor of low malignant potential and accounts for less than 1–2% of exocrine pancreatic tumors [1]. This tumor was described by using various names including ‘solid cystic tumor’, ‘papillary cystic tumor’, ‘papillary epithelial neoplasia’, ‘solid and papillary epithelial neoplasia’, ‘papillary epithelial tumor’ and ‘Frantz’s tumor’, ‘solid and papillary tumor’, ‘solid-cystic papillary epithelial neoplasm’, ‘benign or malignant papillary tumor of the pancreas’ until it was defined by the World Health Organization in 1996 as ‘solid pseudopapillary tumor’ of the pancreas [2]. Sarcoidosis is a multisystem inflammatory disease that mainly affects the intrathoracic lymph nodes, the lungs, the skin and the eyes. The clinical pictures include systemic and organ-specific symptoms. However, in the majority of cases it is diagnosed in asymptomatic patients, based on the finding of hilar adenopathy on chest radiography performed for other reasons [3]. Malignancy can be associated with the occurrence of sarcoid reactions. Problems may also arise in distinguishing between tumor-related sarcoidosis and true systemic sarcoidosis. In this study, we report, to our knowledge, the first case of sarcoidosis associated with pseudopapillary pancreatic tumor.
CASE REPORT

A 44-year-old female presented to oncology center Mansoura University with a one month history of vague abdominal pain and bilateral edema lower limbs. Abdominal ultrasound revealed well defined soft tissue mass at splenic and left renal area with area of cystic degenerations. Further etiological investigations were performed, including abdominal computed tomography (CT) scan which showed large enhanced soft tissue mass in left hypochondriac region with cystic degeneration and foci of calcifications inside. Anteriorly it was seen in contact with greater curvature of the stomach with no clear fat plane in between. Medially, it is seen contacting and displacing pancreatic tail. Chest CT scan showed multiple enlarged pretracheal, aortopulmonary, subcarinal and hilar lymph nodes. The largest seen was subcarinal lymph node measuring 3.5x2.8 cm. Both lung parenchymas showed bilateral perilymphatic nodules. A metastatic cancer was initially suspected then ultrasound-guided Tru-cut biopsy of the abdominal mass showed sheets of small uniform tumor cells surrounding delicate hyalinized fibrovascular stroma forming pseudopapillae. Some cells have eosinophilic others have vacuolated cytoplasm with grooved nuclei. Infrequent mitosis was detected. No significant immunohistochemical staining was observed for CD10/PR picture consistent with pseudopapillary pancreatic tumor (Figure 1). The surgical removal of pancreatic mass was done and sent for pathological evaluation which confirms the result of previous tru-cut biopsy. She was discharged and transferred to chest department Mansoura University for assessment of CT chest. Fiber optic bronchoscopy was done from which bronchoalveolar lavage and transcarinal needle aspiration was taken but showed inflammatory cells without atypical or giant cells.

Follow-up CT scan of chest 10 months later showed bilateral perilymphatic nodules with disappearance of previously described lymphadenopathy (Figure 2). Thoracoscopic lung biopsy then taken and histological examinations revealed non-caseating epithelioid granuloma.

DISCUSSION

A solid pseudopapillary neoplasm (SPN) of the pancreas was described firstly by Dr. Frantz in 1959. It is a rare pancreatic tumors which have a relatively low malignant potential and are mostly diagnosed in young women. The treatment is surgical resection; the prognosis is favorable after resection [4, 5]. Sarcoidosis is a multisystem disease of unknown etiology that can affect any organ. It is characterized by non-caseating granulomatous lesions involving the lungs, skin, eyes, salivary glands and internal organs [6]. The question of whether there is a causal relationship between sarcoidosis and cancer has been debated for years. Sarcoidosis is associated with malignancy more than can be explained by chance. Cancer can occur in patients with an established diagnosis of sarcoidosis and sarcoidosis can subsequently develop in a cancer patient. Malignancy can also be associated with the occurrence of sarcoid reactions. Problems may also arise in distinguishing between tumor-related sarcoid reactions and true systemic sarcoidosis [7]. So, in this study revision of pathological specimen was then carried out in order to make a differential diagnosis between pancreatic tumor associated with sarcoidosis, or the presence of a pancreatic granuloma as a part of a systemic sarcoidosis. Pathological revision showed epithelioid granuloma of thoracoscopic biopsy and solid pseudopapillary pancreatic tumor of pancreatic mass biopsy. The strongest association between sarcoidosis and solid tumors is described with adenocarcinoma of the lung, although other cancers have also been reported. In most cases, the diagnosis of sarcoidosis preceded the detection of neoplasm, leading to the hypothesis that the immune system dysfunction and the tissue chronic inflammation characterizing sarcoidosis can facilitate cancer development. However, it has been also reported cases in which diagnosis of cancer precedes the development of sarcoidosis, as well as cases of concomitant diagnosis. Sarcoid-like reaction occurs more frequently in regional lymph nodes of neoplasm (“typical sarcoid-like reaction”), and is believed to represent a T cell-mediated immune response to soluble antigenic factors shed by the tumoral cells.
However, cases of (“atypical sarcoid-like reaction”) in distant lymph nodes have been observed [3]. Other studies such as Mastroroberto et al. reported the first case of association of sarcoidosis and pancreatic neuroendocrine tumor [3] and Zambrana et al. also reported a case with both sarcoidosis and pancreatic cancer [7]. In this study, we report, to our knowledge, the first case of sarcoidosis associated with pseudopapillary pancreatic tumor.

**CONCLUSION**

This case report summarizes the association between sarcoidosis and pancreatic tumors to be one of the differential diagnoses in our mind while dealing with pancreatic cancer in addition to metastatic lesions.

*********

**Acknowledgements**

We wish to thank all the clinical staff at Clínicas Americas in Medellin Colombia for their support in the care of the patient.

**Author Contributions**

Elhadidy Tamer – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Morsy Nesreen Elsayed – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Abdelwahab Heba Wagih – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Refky Basel – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Zalata Khaled – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

**Copyright**

© 2015 Elhadidy Tamer et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

**REFERENCES**

1. Martin RC, Klimstra DS, Brennan MF, Conlon KC. Solid-pseudopapillary tumor of the pancreas: a surgical enigma? Ann Surg Oncol 2002 Jan-Feb;9(1):35–40.
2. Kloppel G, Solcia E, Longnecker DS, Capella C, Sobin LH. Histological typing of tumors of the exocrine pancreas. In: World Health Organization International Histological Classification of Tumours. 2ed. Berlin, Heidelberg, New York: Springer; 1996:8452/1.
3. Mastroroberto M, Berardi S, Fraticelli L, et al. Sarcoidosis and sarcoid-like reaction associated with pancreatic malignancy: are you able to read a riddle? JOP 2012 Jul 10;13(4):454–7.
4. Frantz VK. Papillary tumors of the pancreas: Benign or malignant? Tumors of the pancreas. In: Atlas of Tumor Pathology, Section 7, Fascicles 27 and 28. Washington, DC, USA: Armed Forces Institute of Pathology; 1959:32–3.
5. Guo N, Zhou QB, Chen RF, et al. Diagnosis and surgical treatment of solid pseudopapillary neoplasm of the pancreas: analysis of 24 cases. Can J Surg 2011 Dec;54(6):368–74.
6. Iannuzzi MC, Rybicki BA, Teirstein AS. Sarcoidosis. N Engl J Med 2007 Nov 22;357(21):2153–65.
7. Zambrana F, Antúnez A, García-Mata J, Mellado JM, Villar JL. Sarcoidosis as a diagnostic pitfall of pancreatic cancer. Clin Transl Oncol 2009 Jun;11(6):396–8.
Edorium Journals: An introduction

Edorium Journals Team

About Edorium Journals
Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission
We sincerely invite you to submit your valuable research for publication to Edorium Journals.

But why should you publish with Edorium Journals?
In less than 10 words - we give you what no one does.

Vision of being the best
We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services
We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review
All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review
All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version
Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status
From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

Six weeks
You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.

Four weeks
After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.

Most Favored Author program
Join this program and publish any number of articles free of charge for one to five years.

Favored Author program
One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program
Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence
We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...
We request you to have a look at our website to know more about us and our services.

We welcome you to interact with us, share with us, join us and of course publish with us.

CONNECT WITH US

We request you to have a look at our website to know more about us and our services.

Edorium Journals: On Web
Browse Journals

This page is not a part of the published article. This page is an introduction to Edorium Journals and the publication services.