Journal of Gastrointestinal Cancer (2019) 50:935–938
https://doi.org/10.1007/s12029-018-0123-2

CASE REPORT

Primary Angiosarcoma Pancreas: a Case Report of an Exceptional Localization

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Published online: 2 June 2018

Introduction

Pancreas sarcomas are very rare and represent less than 1% of all pancreas cancers [1]. Angiosarcoma is uncommon and has defavorable prognosis [1, 2]. The diagnosis of angiosarcoma is based on histological analysis and differentiated from aciae hemangiopericytoma and Kaposi's sarcoma [3]. We reported a case of primary non-metastatic pancreas angiosarcoma in a 41-year-old man in Togo, confirmed by immunohistochemistry. We described epidemiological, morphological, and immunohistochemical aspects of primary pancreas angiosarcoma.

Case Report

A 41-year-old Togolese man with a history of chronic alcohol consumption was admitted to the Sylvanus Olympio Surgery Department in Lomé for acute epigastria pain, jaundice, nausea, and vomiting. The biological assessment revealed an elevation of amylase and lipase of 1012 and 1160 U/l, respectively. The liver function test showed an elevation of enzymes, bilirubin (total bilirubin 15.78 mg/dl, direct bilirubin 12.86 mg/dl, indirect bilirubin 0.99 mg/dl) and alkaline phosphatase (398.64 U/1). Ultrasound examination revealed an heterogeneous tumor mass of the pancreas body, without bile duct abnormalities. A contrast-enhanced CT scan noted a hypo-attenuated heterogeneous tumor of the pancreas body, measuring 63 mm × 46 mm with an infiltration of the para-glandular fat and circumferential entanglement of the superior mesenteric artery (SMA) and the superior mesenteric vein (SMV) (Fig.1). There was no evidence of bile duct dilatation. There was no distant spread location. Due to the impossibility of tumor resection, a trans-parietal biopsy was performed, guided by CT scan for histopathological study.

The histological sections showed tumor proliferation of vascular layers with tumor cells exhibiting cyto-nuclear atypia and hyperchromatic endothelial cells (Fig. 2). The hemangiopericytoma diagnosis was retained without eliminating angiosarcoma. The paraffin blocks were sent to an anatomic pathology laboratory in France for immunohistochemistry. Immunohistochemestry study showed the positivity of CD34, CD31, and factor VIII and the negativity of vimentin markers, including S100 (Figs. 3, 4, and 5). These markers had eliminated hemangiosarcoma and retained the diagnosis of primary non-metastatic angiosarcoma. The patient was sent to the medical oncology unit for chemotherapy. Due to lack of financial resources, the family was unable to afford the chemotherapy cures. The patient died 2 weeks after the diagnosis was established.

Discussion

Epidemiology

Angiosarcoma represents a particular form of sarcomas developed from endothelial cells of blood and lymphatic vessels

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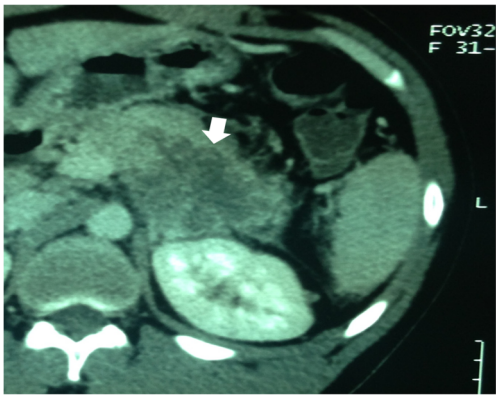

Fig. 1 Contrast-enhanced CT scan showing the tumor of the body of the pancreas (white arrow)

[1]. These are very rare malignant tumors, less than 1% of soft tissue sarcomas but with very aggressive and a metastatic potential [1, 3]. Primary angiosarcoma of the pancreas is an extremely rare cancer. Seth et al. reported an incidence of 0.1% of pancreatic sarcoma after examining 5000 cases of pancreatic cancer [3]. According to Kim et al., pancreatic sarcomas occur frequently in younger subjects. Our subject was 41 years old, similar to that described in literature [1, 4].

Clinical Features

The clinical manifestations of angiosarcoma vary significantly. Upper abdominal pain is the most common manifestation [3]. Other symptoms include weakness or fatigue, shortness of breath, fever, chest pain, weight loss, and anorexia [2]. A minority of patients are asymptomatic and angiosarcoma is discovered incidentally. These results are similar to those of other pancreatic pathologies and, therefore, are not specific to angiosarcoma. Seth and many other authors consider jaundice as a sign of an advanced lesion [3]. This was true for the case we presented. The patient had an advanced tumor, so curative resection was not feasible. Pancreatic angiosarcoma is often diagnosed at advanced stage where the unique treatment is chemotherapy [5].

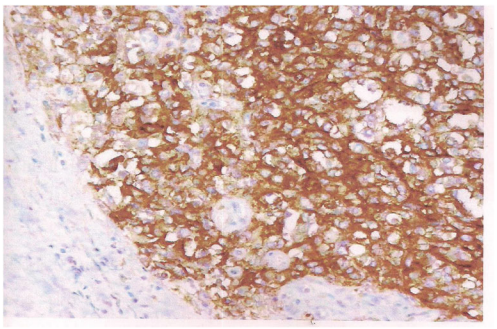

Fig. 3 Note the positivity of tumor cells to markers CD31 in immunohistochemistry (G × 100)

Imaging

Imaging studies including CT, magnetic resonance imaging (MRI), positron emission tomography (PET) scan, and ultrasound can be used to define the extent of abdominal angiosarcoma. Abdominal ultrasound is the first-line examination of any abdominal pain in our context although it is often insufficient for the analysis of a pancreatic mass, leading to the use of CT scan. While CT has a high sensitivity for diagnosing pancreatic masses, it has limited diagnostic value in reliably distinguishing adenocarcinoma from other cancers [6]. CT findings for pancreatic sarcomas can range from a finely heterogeneous solid mass to a highly heterogeneous enhancing lesion with vascularization and necrosis [2, 6]. CT scan is used to guide biopsies of the pancreatic masses [6].

Pathologic Findings

The diagnosis of pancreas angiosarcoma may be very challenging due to the non-specific clinical, radiological, and histopathological features [1, 5].

Histological characteristics of angiosarcoma may provide useful clues for diagnosis. Angiosarcoma can be classified into different subtypes based on the cytologic appearance: spindle-shaped endothelial cells, epithelioid with large rounded or polygonal cells, and pleomorphic or mixed phenotypes as in most angiosarcoma [7]. Areas of irregular anastomosing vasculature lined by atypical

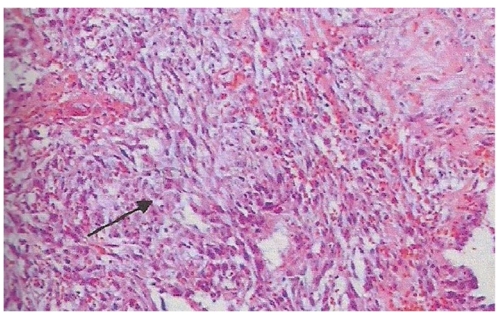

Fig. 2 Note the presence of multiple vascular elements with little differentiation (arrow) and mitoses (H&E, G × 200)

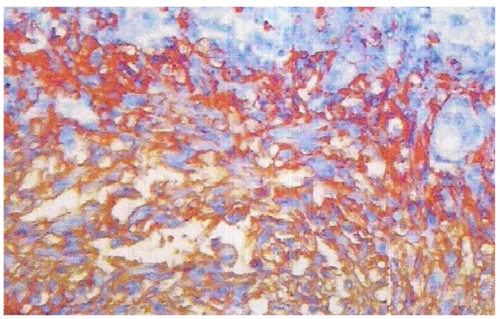

Fig. 4 Note the positivity of tumor cells to CD34 markers in immunohistochemistry (G × 100)

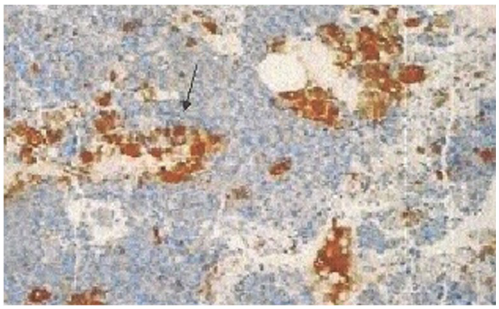

Fig. 5 Note the positivity of tumor cells to vimentin in immunohistochemistry (G × 100)

endothelial cells are readily identifiable in histology study. However, the existence of anaplasia in most of angiosarcomas makes it challenging to distinguish such tumors from other undifferentiated ones such as hemangiopericytoma melanoma [8]. Therefore, immunohistochemistry is mandatory to establish the definitive diagnosis of angiosarcoma. Expression of endothelial markers CD31, CD34, von Willebrand factor (vWF), *Ulex europaeus* agglutinin 1 (UEA-1), Friend leukemia integration 1 (Fli-1), endothelin-1, vascular endothelial growth factor (VEGFR), and erythroblast transformation-specific-related gene (ERG) can help identify angiosarcomas, each with different sensitivity and specificity, among which the vWF, UEA-1, and CD31 are considered the most useful for the diagnosis of poorly differentiated cases [9, 10]. However, some of these markers may be lost due to progressive tumor dedifferentiation. Otherwise, angiosarcoma may express cytokeratins, which makes it difficult to distinguish it from poorly differentiated carcinomas [10]. Therefore, a number of other markers with different tissue specificity including lymphatic endothelial (lymphatic vessel endothelial hyaluronan receptor-1/Lyve-1 or podoplanin/D2-40), smooth muscle (desmin), neural (S100, SOX10), epithelial (keratin), stromal (CD117, DOG1), melanocytic (HMB-45, melan-A), human herpes virus 8 (HHV-8), and mesothelial (calretinin, CK5/6, HBME-1 and WT-1) can be used to distinguish angiosarcoma from other tumors such as melanoma [11, 12]. In our clinical case, the search for Ac anti-HHV8 that is specific to Kaposi's sarcoma was negative. The hemangiopericytoma is positive for certain markers specific for angiosarcoma as CD31 and CD34. It is a tumor that does not have specific markers such as Kaposi's sarcoma [11]. In our case, the strong positivity of factor VIII and the morphology of the lesions allowed us to make the diagnosis of angiosarcoma [11, 12].

Treatment

The best treatment option for angiosacoma of the pancreas remains controversial. Surgery appears to be the most effective treatment approach [13]. Anderson et al. [13] suggested that post-operative adjuvant radiotherapy may contribute to local control in a manner similar to angiosarcoma of other sites; however, Salti et al. [14] held a contrary view and observed that radiotherapy does not seem to prolong survival and chemotherapy should be added to the treatment. Sindhu et al. [15] suggested that chemotherapy may be used for palliative treatment, although the response is likely to be short.

Prognosis

Primary angiosarcoma of the pancreas is a very aggressive tumor with a poor prognosis. The prognosis of angiosarcoma is dismal with a 5-year survival rate lower than 30% for non-metastatic angiosarcoma, and an overall survival of 8 months for the metastatic stage [13, 14].

Conclusion

Primary angiosarcoma of the pancreas is extremely rare and of poor prognosis. It does not have any imaging-specific features. It raises real problems in countries with limited resources such as Togo, especially in terms of diagnosis because of the lack of immunohistochemistry. It is therefore important to strengthen pathology units by providing immunohistochemistry services in order to facilitate early diagnosis. Policy makers should act to ensure that population had adequate and affordable health insurance that covers chemotherapy.

Data Availability Statement All data generated or analyzed during this study are included in this published article.

Authors' Contributions TD is responsible for the conception of the study, participated in the study design, performed the laboratory analysis and interpretation, and wrote the paper. MT, BT, FA, and SD were involved in the clinical and therapeutic management of the patient; they have reviewed the paper. GNK was responsible for the overall scientific management of the study and the preparation of the final paper. All the authors have read and approved the final paper to be submitted for publication.

Compliance with Ethical Standards

Competing Interests The authors declare that they have no competing interests.

Ethics Approval and Consent to Participate This study received approval from the head of the laboratory department to be conducted (Ref No. 08/2017/LAP/CHUSO).

Consent for Publication Written informed consent for publication of their clinical details and clinical images was obtained from the patient. A copy of the consent form is available for review by the Editor of this journal.

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