

Interesting Images

Primary Angiosarcoma of the Abdominal Aorta Following Endovascular Aneurysm Repair Mimicking Vascular Graft Infection

Marcel Wehmann ¹ , Martin Freesmeyer ^{1,*} , Anika Biermann ², Jürgen Zanow ³ and Mika Henkenjohann ³

¹ Clinic of Nuclear Medicine, Jena University Hospital, 07747 Jena, Germany; marcel.wehmann@med.uni-jena.de

² Section Pathology, Institute of Forensic Medicine, Jena University Hospital, 07747 Jena, Germany; anika.biermann@med.uni-jena.de

³ Department of General, Visceral and Vascular Surgery, Jena University Hospital, 07747 Jena, Germany; juergen.zanow@med.uni-jena.de (J.Z.); mika.henkenjohann@med.uni-jena.de (M.H.)

* Correspondence: martin.freesmeyer@med.uni-jena.de; Tel.: +49-3641-9329801

Abstract

A 64-year-old woman presented with recurrent abdominal pain and rapid weight loss of 10 kg. Five years prior, an abdominal endovascular aneurysm repair (EVAR) was performed, followed by an open repair with proximal graft interposition due to an endoleak type IA two years ago. Laboratory tests revealed only moderately elevated infection markers (CrP 66 mg/L, WBC 14 GPt/L). ¹⁸F-FDG PET/CT showed a hypermetabolic peri-aortic mass initially suspected to be inflammatory. However, the patient did not exhibit signs of fever. Because of the equivocal constellation, a fine-needle biopsy was conducted, revealing malignant cells. The patient underwent complete tumor resection including aortic replacement. Histopathology confirmed the diagnosis of an angiosarcoma. The treatment was then supplemented with chemotherapy using doxorubicin and ifosfamide.

Keywords: angiosarcoma; prosthetic infection; surgery; vascular replacement; ¹⁸F-FDG positron-emission tomography; immunohistochemistry

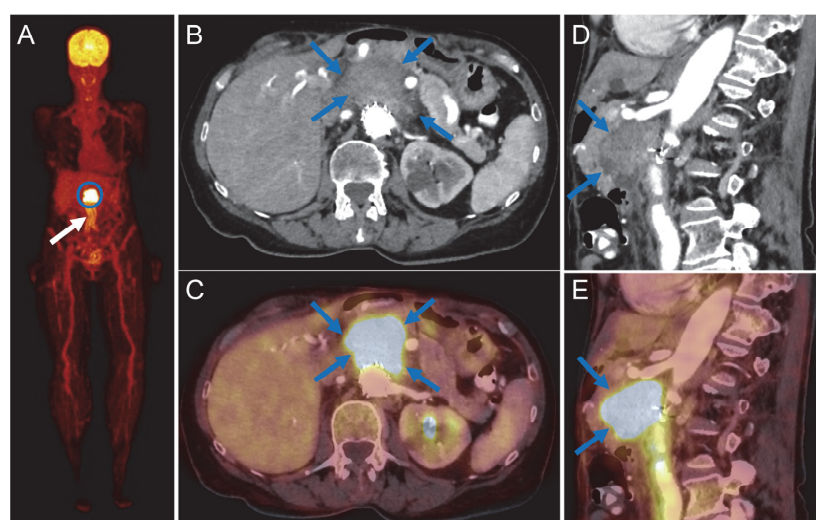


Figure 1. The PET ((A) thick-slice maximum intensity projection (MIP), ap-view) revealed a homogeneous hypermetabolism along the implanted aorto-iliac prosthesis (white arrow) and a markedly



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tracer-accumulating peri-aortic mass (blue circle), without compression of the aorta itself, adjacent to the bare spring of the EVAR. The maximum standardized uptake value (SUV_{max}) of the lesion was 23.6. No additional hypermetabolic lesions were present. Given the potential for both highly inflammatory processes and malignant causes to result in highly elevated SUV_{max} values, a definitive distinction could not be made [1–3]. This diagnostic uncertainty is not exclusive to our case. Integrated, multiparametric imaging approaches, combining anatomical modalities such as ultrasound, CT, or MR with ^{18}F -FDG PET, have recently been proposed and validated to increase diagnostic certainty in similarly equivocal constellations, where morphological imaging alone remains inconclusive [4]. Such combined strategies have been shown to improve the accuracy of detecting malignancy when compared with the use of any individual modality. This supports their application in cases where an elevated SUV_{max} cannot be reliably distinguished from an inflammatory or malignant process [5]. The contrast-enhanced CT images in axial (B) and sagittal view (D) with the corresponding ^{18}F -FDG-PET/CT fusion images (C,E) provide a detailed depiction of the tumor extent (blue arrows), demonstrating the space-consuming aspect and the association to the residual part of the EVAR prosthesis. Based on the distribution of metabolic activity observed around the aorta, a preliminary diagnosis of prosthetic infection with formation of hypertrophic inflammatory tissue was made. To confirm the diagnosis, endoscopic ultrasound with fine-needle aspiration (FNA) was performed, which revealed malignant cells. Consequently, the patient underwent a surgical procedure.

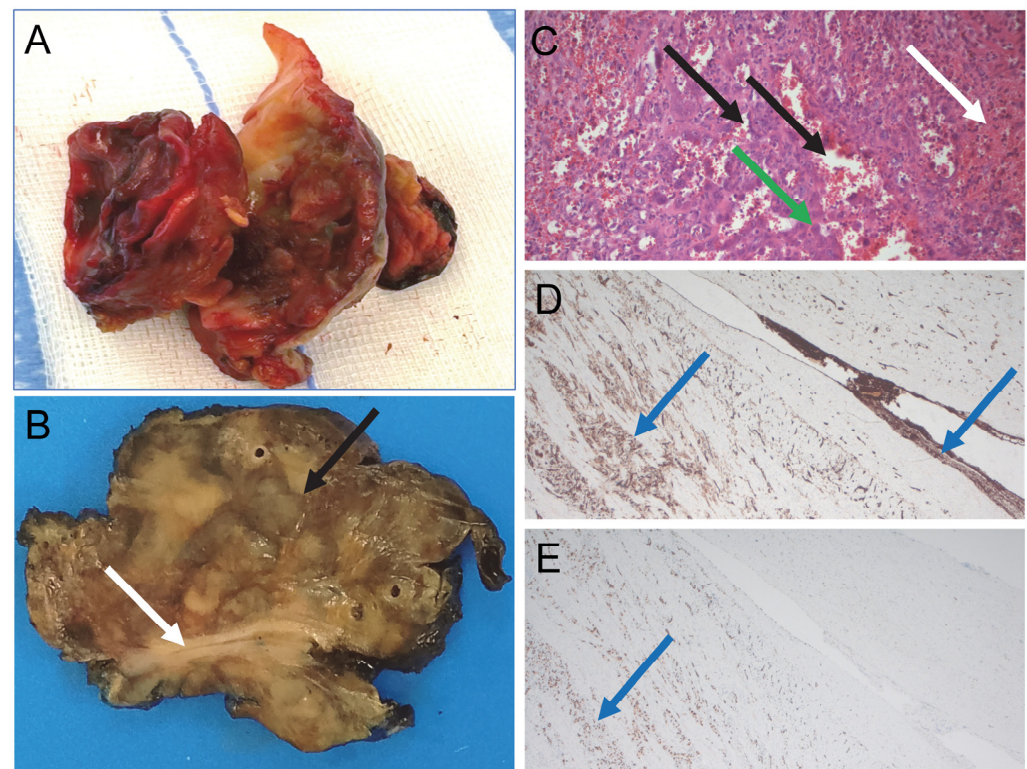


Figure 2. Histopathological and immunohistochemical characterization of the resected epithelioid angiosarcoma. (A) Photo of the resected angiosarcoma taken in the operating room, (B) Tumor specimen after laminar sectioning, (C) Lesion in HE, 200× magnification, (D) Vascular wall stained with CD31, 40× magnification, (E) Vascular wall stained with ERG, 40× magnification. The in toto resected angiosarcoma (A) measured 5.6 × 5.3 × 3.4 cm. For a curative therapeutic approach, it is necessary to perform complete surgical removal [6]. Laminar sections of the space-occupying lesion (B) showed multiple nodules (black arrow) and parts of the vessel wall (white arrow). Microscopically, enlarged cells with an increased nuclear–cytoplasmic ratio forming irregular cavities filled with blood cells are seen ((C) haematoxylin and eosin, black arrows). As these neoplastic structures form no proper vessels, erythrocyte extravasations occur, resulting in hemorrhages (white arrow) and leading to the diagnosis of angiosarcoma. Since the neoplastic cells resemble epithelial cells due to their abundant eosinophilic cytoplasm (green arrow) the subtype of an epithelioid angiosarcoma is

diagnosed. Immunohistochemistry of endothelial markers CD31 ((D) cytoplasmic expression) and ERG ((E) nuclear expression) are primarily utilized to confirm differentiation and classification and illustrates the highly infiltrative nature of the neoplasm (blue arrows) [7,8]. Despite the low incidence of angiosarcomas, which account for 2% of soft-tissue sarcomas, some cases have been reported in which the malignancy has also occurred subsequent to the implantation of a vascular graft [9,10]. Furthermore, prosthesis infection was frequently suspected initially in similar situations [11]. A subsequent PET/CT scan, conducted four months later, revealed the presence of soft-tissue metastases in the gluteal region, in addition to multifocal bone metastases. The patient underwent chemotherapy with doxorubicin and ifosfamide.

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Data Availability Statement: The original contributions presented in this study are included in the article. Further inquiries can be directed to the corresponding author.

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Abbreviations

The following abbreviations are used in this manuscript:

EVAR	Endovascular aneurysm repair
¹⁸ F-FDG	Fluorine-18 fluorodeoxyglucose
PET/CT	Positron-emission tomography/computed tomography
SUV _{max}	Maximum standardized uptake value

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