Radiology Case Reports 12 (2017) 845–849

Available online at www.sciencedirect.com

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journal homepage: http://Elsevier.com/locate/radcr

Oncology

Radiological findings of two neoplasms with perivascular epithelioid cell differentiation

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ARTICLE INFO

Article history:
Received 1 April 2017
Accepted 1 June 2017
Available online

Keywords:
PEComa
Perivascular
Epithelioid
Mesenchymal

ABSTRACT

Perivascular epithelioid cell tumors (PEComas) constitute a rare subset of mesenchymal neoplasms classified by the World Health Organization in 2002. We present two cases of PEComas; the first is a cervical PEComa in a 35-year-old woman with no known past medical history who presented with a palpable pelvic mass; the second is an adnexal PEComa in a 39-year-old woman with a history of colitis who presented with abdominal pain and diarrhea. The rarity of these tumors has led to little information about imaging characteristics which we hope these two cases will help expand.

Introduction

Neoplasms with perivascular epithelioid cell differentiation, commonly abbreviated as PEComas, are defined as mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells [1]. This relatively new family of neoplasms, as defined by the World Health Organization Classification of Tumors in 2002, includes angiomyolipomas of the kidneys; clear cell "sugar" tumor of the lung; lymphangioleiomyomatosis; clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres; and unusual clear cell tumors of the pancreas, rectum, abdominal serosa, uterus, vulva, thigh, and heart [1,2]. The rarity of these tumors has led to little information about imaging characteristics which we hope these two cases will help expand.

Case report 1

A 35-year-old woman with no known past medical history presented to the gynecologic oncology office after a pelvic mass was palpated during routine pelvic examination. She also reported pelvic pressure and stated that she noted a bulging mass during bowel movements. No additional complaints or abnormalities were elicited during the review of medical history or upon physical examination.

Contrast-enhanced MRI examination of the pelvis obtained to evaluate the patient's complaints demonstrated

Competing Interests: The authors have declared that no competing interests exist.

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https://doi.org/10.1016/j.radcr.2017.06.001

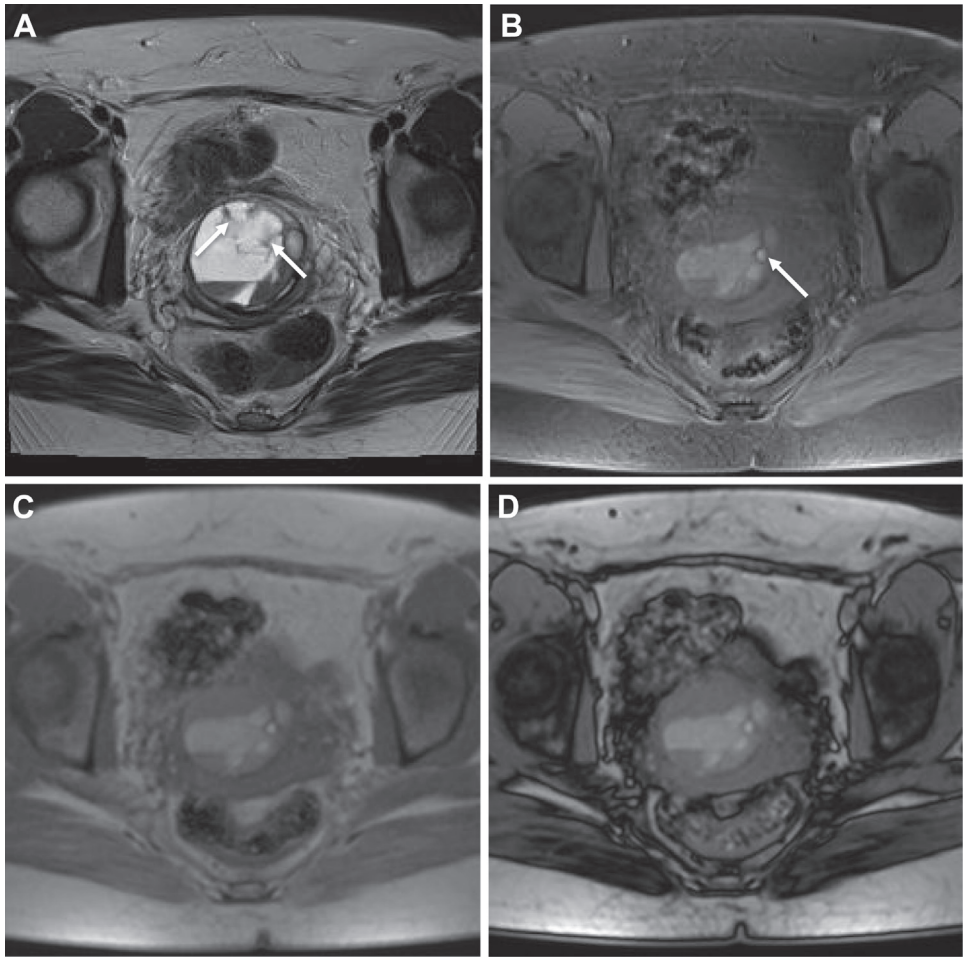

Fig. 1 – (A) Axial T2-weighted image at the level of the pelvis demonstrates a complex cystic mass arising from the anterior wall of the cervix, with internal septations (arrows) of varying thickness. (B) Axial T1-weighted image with fat suppression demonstrates areas of high T1 signal (arrow) within the mass, consistent with hemorrhagic or proteinaceous fluid. (C) and (D) Axial in-phase and out-of-phase images demonstrate absence of microscopic fat within the mass.

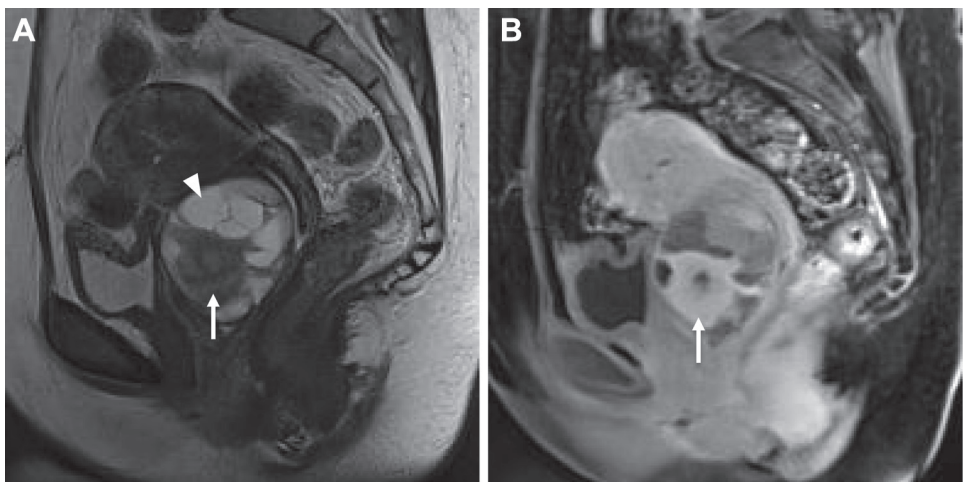

Fig. 2 – (A) Sagittal T2-weighted image in the same patient again shows the large mass with cystic (arrowhead) and solid (arrow) components arising from the anterior cervical wall. (B) Postcontrast fat-saturated T1-weighted sagittal image demonstrates avid enhancement within the solid component (arrow).