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Radiological findings of two neoplasms with perivascular epithelioid cell differentiation

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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

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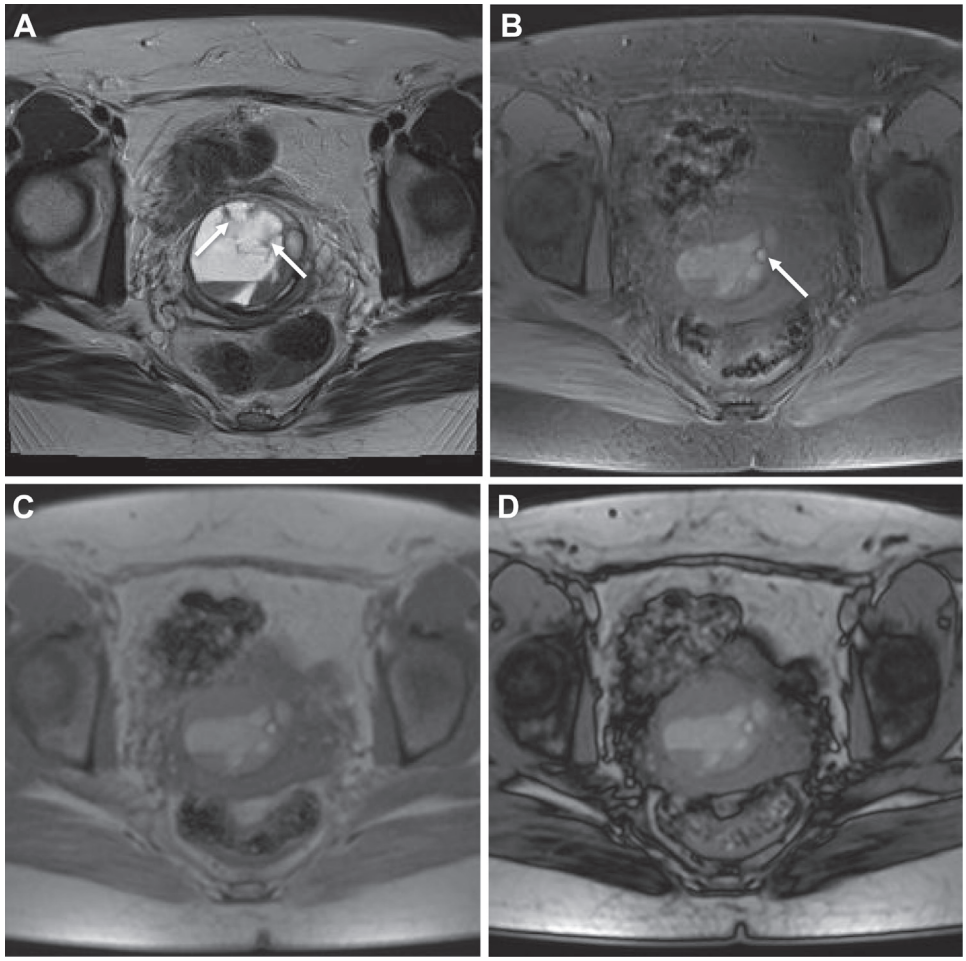

Fig. 1 – (A) Axial T2-weighted image at the level of the pelvis demonstrates a complex cystic mass arising from the anterior wall of the cervix, with internal septations (arrows) of varying thickness. (B) Axial T1-weighted image with fat suppression demonstrates areas of high T1 signal (arrow) within 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.

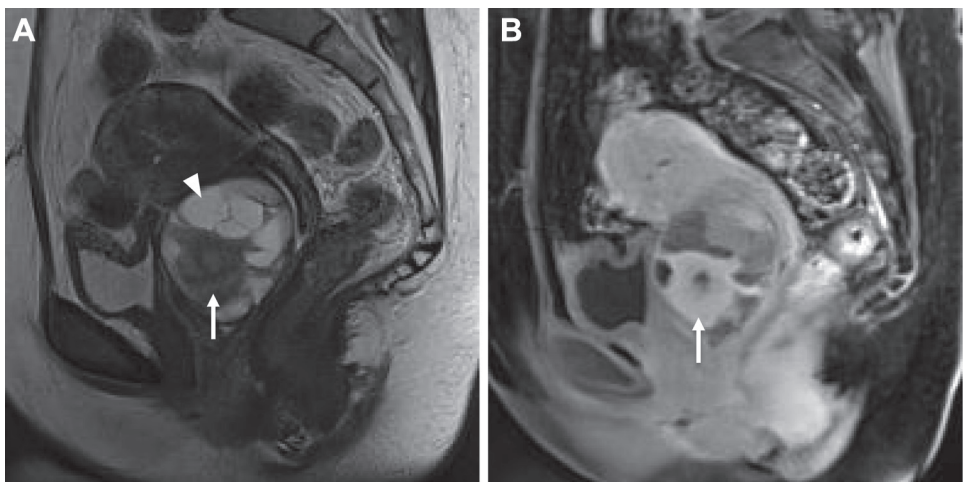

Fig. 2 – (A) Sagittal T2-weighted image in the same patient again shows the large mass with cystic (arrowhead) and solid (arrow) components arising from the anterior cervical wall. (B) Postcontrast fat-saturated T1-weighted sagittal image demonstrates avid enhancement within the solid component (arrow).

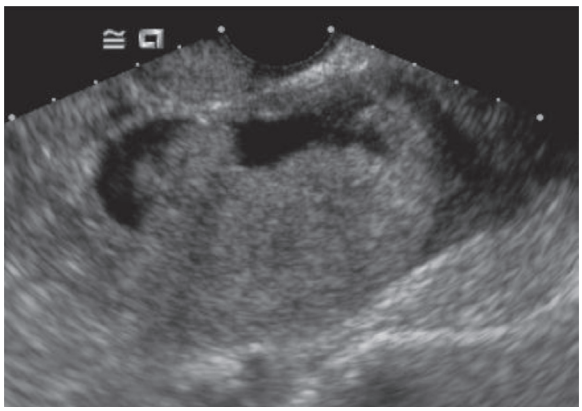

Fig. 3 – Sagittal grayscale image of the right adnexa from the transvaginal portion of the study shows a complex mass with solid and cystic components.

circumscribed 6-cm complex mass with solid and cystic components originating from the anterior cervical wall (Figs. 1 and 2). Internal septations of varying thickness were present within the mass, in addition to solid avidly enhancing components. Small locations of high T1 signal intensity within the mass correlated to hemorrhagic or proteinaceous fluid. No loss of signal intensity within the mass was observed on chemical shift imaging or upon application of fat suppression to indicate presence of microscopic or macroscopic fat. There was no free fluid in the pelvis, no inguinal or pelvic lymphadenopathy, or involvement of the adjacent organs. Both ovaries and the remainder of the uterus showed no abnormalities, with exception of an intrauterine device within the endometrial canal and postsurgical changes related to prior cesarean section.

Case report 2

A 39-year-old woman with a past medical history of colitis presented to the emergency department with complaints of abdominal pain and diarrhea that were intermittent for the past month and a half. Additionally, she reported a 10 lb unintentional weight gain, bloating, and early satiety for the past month. She denied difficulty with urination or bowel movements. The remainder of the history and physical examination were noncontributory.

Sonographic examination of the pelvis with transabdominal and transvaginal approach demonstrated a solid and cystic mass in the right adnexa with foci of arterial and venous flow on color Doppler assessment (Fig. 3). The right ovary was not visualized. A normal left ovary was identified, and small myomata were also noted in the uterus.

Subsequently, a computed tomography examination of the abdomen and pelvis with oral and intravenous contrast material was performed (Fig. 4). This study confirmed presence of a complex circumscribed mass in the right adnexa, interposed between the posterior uterine wall and the rectosigmoid colon. Solid and cystic components were present in the mass with avid enhancement in the solid components. Trace free fluid was present in the pelvis, in addition to mild circumferential wall thickening of the sigmoid colon.

MRI examination of the pelvis without and with intravenous contrast was performed later the same day for further evaluation and characterization of the adnexal mass, with similar findings as on computed tomography examination (Fig. 5). No microscopic or macroscopic fat was identified within the mass, which contained areas of high T1 signal intensity, representing hemorrhagic or proteinaceous fluid; avidly enhancing solid components were seen. Multiple cervical nabothian cysts were incidentally noted. Wall thickening and enhancement of the rectosigmoid colon persisted suggesting mild colitis and explaining clinically reported diarrhea. The above described mass focally abutted portions of the sigmoid colon and the uterus, making the assessment of the site of origin difficult.

Discussion

Few descriptions of the radiological appearance of PEComas exist to define specific diagnostic imaging characteristics. Of

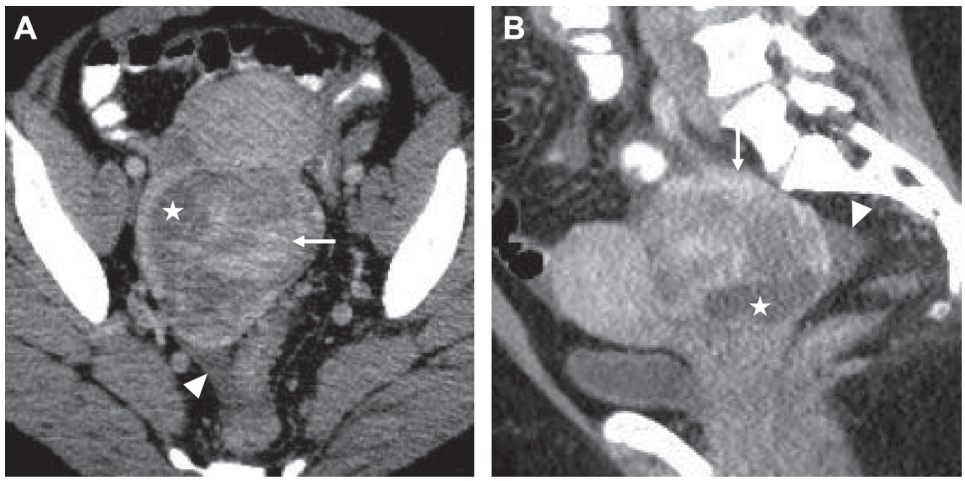

Fig. 4 – (A) and (B) Contrast-enhanced axial and sagittal CT images show a complex right adnexal mass with cystic (stars) and avidly enhancing solid (arrows) components. Trace free fluid is identified in the pelvis (arrowheads).

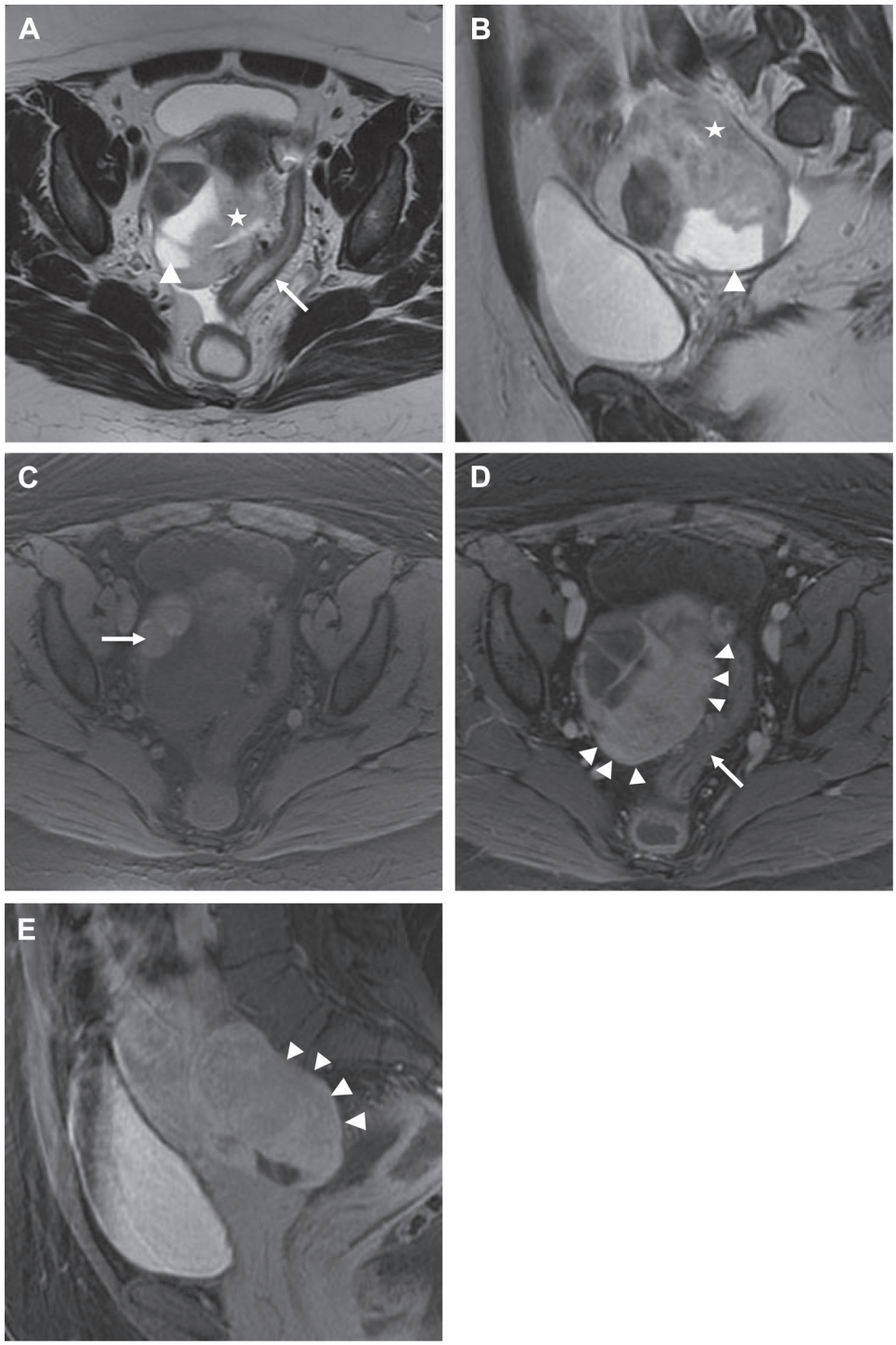

Fig. 5 – (A) and (B) T2-weighted axial and sagittal images demonstrate a complex right adnexal mass with cystic (arrowheads) and solid (star) components. There is also mild circumferential wall thickening of the rectosigmoid colon (arrow). (C) Small areas of high T1 signal (arrow) within the mass on T1-weighted image with fat saturation represent hemorrhagic or proteinaceous fluid. (D) and (E) Following intravenous contrast administration, fat-saturated T1-weighted axial and sagittal images show avidly enhancing solid components (arrowheads), as well as wall thickening and enhancement of the rectosigmoid colon.

the reports available in the literature, many tumors have a non-specific imaging appearance, precluding generalization and definitive recognition [2]. Presence of microscopic fat is diagnostic of angiomyolipomas of the kidney, which belong to this new group of tumors; however, this finding has not been seen in PEComas at other organs and locations. The

heterogeneous appearance of these tumors is in part related to areas of cystic degeneration and/or necrosis and explains heterogeneous enhancement on cross-sectional studies with avid enhancement in solid portions of the tumors; this description has been utilized in multiple case reports and large cohort studies of PEComas [2–4]. Encapsulation has been reported with some of the tumors, especially with nonaggressive histology, resulting in description of circumscribed mass with solid and cystic components [2–4]. These varied reports indicate the significance of pathologic classification in the diagnosis of PEComa as imaging characteristics are nonspecific.

In the two cases presented here, significant imaging similarities exist. First, both masses are circumscribed, without involvement of the adjacent organs or structures. Additionally, both masses contained solid and cystic components, as has been commonly reported [4,5], with areas of high T1 signal intensity internally, representing hemorrhagic or proteinaceous fluid; this description has not been utilized in prior case reports and studies. Neither mass contained microscopic or macroscopic fat. Solid components of both masses showed avid enhancement, with both tumors exhibiting similar MRI appearance and overall heterogeneous enhancement. Both tumors were located in the pelvis, in close proximity to the uterus, abutting the anterior and/or posterior wall of the uterus, possibly explaining similarities in imaging appearance.

These two cases have striking imaging similarities; they both presented as circumscribed solid and cystic masses with avid enhancement in the solid components. This description is in agreement with imaging characteristics in prior reports in the literature. Both masses contained loculations of hemorrhagic or proteinaceous fluid, a finding not previously reported in PEComas. Both of the masses were located in the pelvis, and neither of them contained microscopic or macroscopic fat. Overall, PEComas are a new subset of mesenchymal tumors that may be included in the differential diagnosis of solid and cystic masses, with final diagnosis confirmed with histological and immunohistochemical confirmation of perivascular epithelioid cells.

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