

Vanishing ovarian mass: Sarcoidosis

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A woman was referred to our hospital with the working diagnosis of ovarian malignancy. While she was undergoing both clinical and radiological evaluation and monitoring, a decrease in the size of the ovarian mass was noted. After further evaluation via laboratory findings and tissue biopsy, we arrived at a final diagnosis of sarcoidosis, which is very unusual in the ovaries. Our case places emphasis on the importance of considering rare entities, such as ovarian sarcoidosis, and the importance of radiologic changes in solid ovarian mass dimensions over time.

Introduction

Sarcoidosis is a chronic multisystem disorder of unknown etiology, characterized (histologically) by the presence of noncaseating epithelioid granulomas. The disease can affect every organ system; however, ovarian involvement is extremely rare. According to Wuntakal et al, seven cases of ovarian involvement have been reported in the English language literature (1). Ovarian sarcoidosis may be misdiagnosed by radiologists as a solid ovarian malignancy. In this case report, we present a case of ovarian mass which, despite the presence of imaging features suggestive of an ovarian malignancy, demonstrated a significant decrease in size over a relatively short period of time, a phenomenon more consistent with ovarian sarcoidosis.

Case report

A 50-year-old Caucasian woman with a history of abdominal discomfort and pain for three months was referred from an outside hospital. She needed a recommendation

for further management of a unilateral, complex, solid-mass lesion (9cm x 6.5cm x 5cm) in the left adnexum and for multiple enlarged lymph nodes found on abdominal ultrasonography (US). She had a medical history of right salpingo-oophorectomy, and her case was managed within the gynecology/oncology multidisciplinary framework.

MRI demonstrated a left adnexal mass accompanied by a complex cyst (Fig. 1). Multiple enlarged lymph nodes appeared in the paraaortic, left common and internal iliac

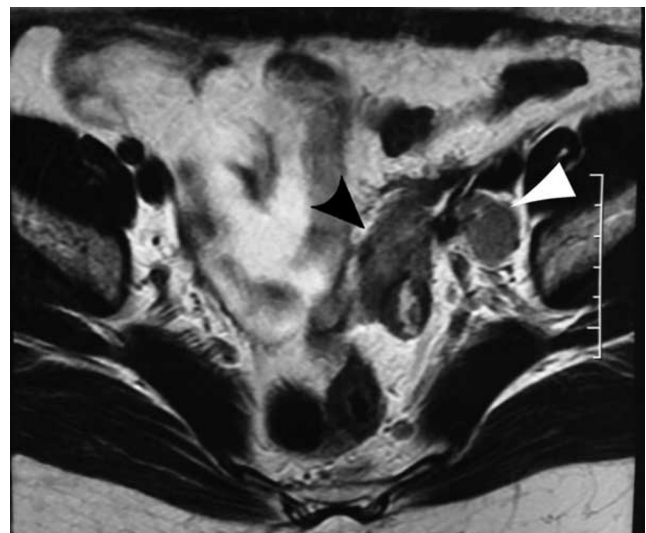


Figure 1. 50-year-old woman with ovarian sarcoidosis. T2-weighted MR image of the pelvis depicting left adnexal mass (black arrowhead) concomitant with posteriorly localized complex cystic lesion and enlarged lymph node (white arrowhead) in the left internal iliac chain.

Citation: Turkay R, Bakir B, Golabi UC, Topuz S, Ilhan HR. Vanishing ovarian mass: Sarcoidosis. *Radiology Case Reports*. (Online) 2012;7:685.

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

DOI: 10.2484/rcr.v7i3.685

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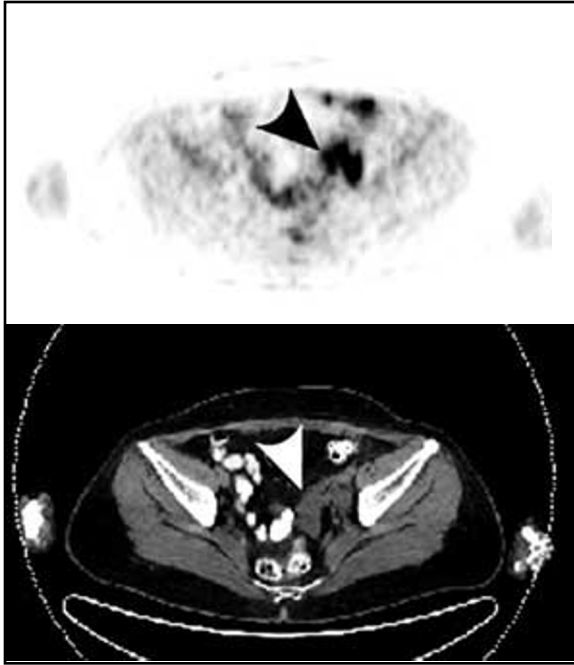


Figure 2. 50-year-old woman with ovarian sarcoidosis. PET/CT scan depicts a left adnexal mass (arrowheads) with left internal iliac lymph nodes, both demonstrating increased FDG uptake.

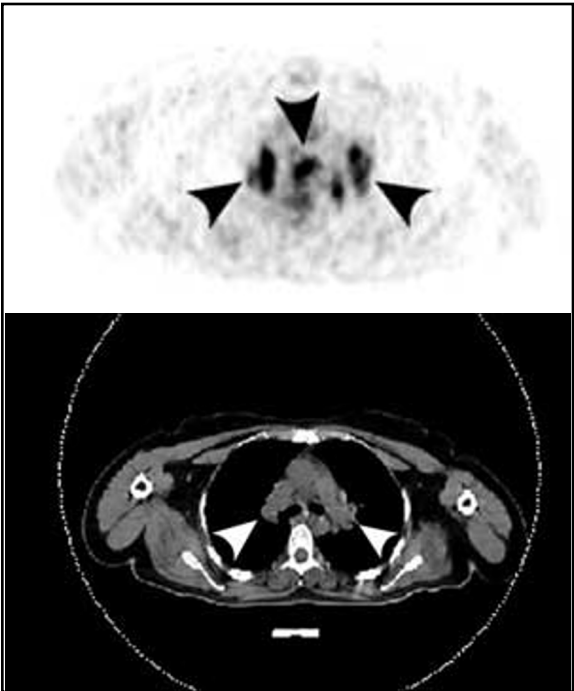


Figure 3. 50-year-old woman with ovarian sarcoidosis. PET/CT scan demonstrates bilateral hilar and subcarinal enlarged lymph nodes (arrowheads) with increased FDG uptake.

(Fig. 1), and inguinal regions, measuring up to 2.5cm on the short axis.

PET/CT revealed a left adnexal mass with increased FDG uptake (SUD max=5.2) (Fig. 2). Multiple left iliac, para-aortic, and inguinal enlarged lymph nodes (SUD max=6.2) were noted, along with mediastinal, hilar, and bilateral supraclavicular enlarged lymph nodes (SUD max=7.9 in the mediastinum) (Fig. 3). In light of these findings and the patient's clinical picture, the gynecological/ oncologic multidisciplinary team recommended clinical review and imaging-guided lymph-node biopsy.

After a two-month period of evaluation and multidisciplinary care, the decision was made to perform an ultrasound to determine the most appropriate lymph node to biopsy for further evaluation. A subsequent decrease in ovarian mass size over this two-month period prompted further imaging with MRI. This revealed marked regression of the mass to 4x3x2.5cm; however, the previously present lymph nodes persisted on exam (Fig. 4). There was

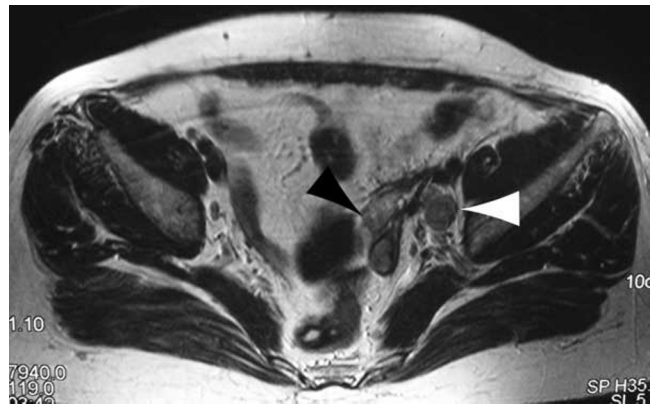


Figure 4. 50-year-old woman with ovarian sarcoidosis. T2-weighted axial pelvic MRI depicts regression of the left adnexal mass (black arrowhead) despite persistence of the left internal iliac lymph node (white arrowhead) with the complex cystic lesion.

no interval progression or development of new lymph-node involvement. The woman underwent US-guided right internal iliac lymph-node biopsy, following a nondifferentiating biopsy of inguinal nodes. Histology revealed noncaseating granulomatous lymphadenitis with negative stains for acid-fast bacilli and fungi. No foreign body was seen. Given the histological findings, our attention was then directed to an inflammatory process. With a working diagnosis of sarcoidosis, the patient was subsequently referred to a pulmonologist who performed bronchoscopy with bronchoalveolar lavage, bronchial biopsy, transbronchial biopsy, and mediastinal lymph-node biopsy. A transbronchial right-lower-lobe biopsy revealed noncaseating granulomatous inflammation, and lymph-node biopsy revealed epithelioid histiocytes with noncaseating granulomatous inflammation (Fig. 5). Based on these clinical, radiological, and histologic

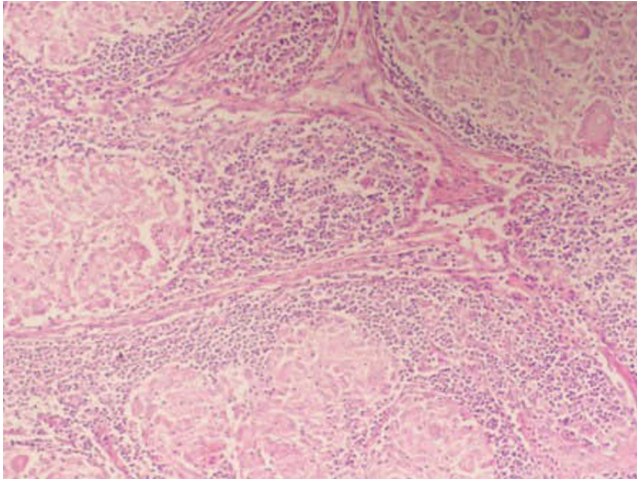


Figure 5. 50-year-old woman with ovarian sarcoidosis. Non-necrotizing granuloma with epithelioid histiocytes artery (x100).

findings, we arrived at the extremely rare diagnosis of sarcoidosis.

Discussion

Sarcoidosis has a variety of clinical and radiologic manifestations. It commonly affects young and middle-aged adults, and frequently presents with bilateral hilar lymphadenopathy, which is the most common radiologic finding and may occur with associated pulmonary infiltrates, and ocular and skin lesions (2). Other organs may also be involved. Genitourinary tract sarcoidosis is seen in 5% of patients in autopsy series (2). Renal manifestations of sarcoidosis are interstitial nephritis, glomerulonephritis, and nephrocalcinosis due to hypercalcemia (2, 3). Renal functions are usually protected, and radiologic imaging is possible. In such cases, contrast-enhanced CT can depict striated nephrograms (2, 3). The epididymis is the most affected organ in genital sarcoidosis, followed by testicular involvement (which is very rare and particularly accompanied by the epididymis) (2, 3, 4). Acute epididymo-orchitis or painless masses in the scrotum and testicular swelling can be clinical manifestations (4). Characteristically, testicular involvement of sarcoidosis is multiple and bilateral (2, 3). US depicts multiple hypoechogenic nodules that demonstrate low signal intensity on T2-weighted MRI and show enhancement after contrast injection (2). Sarcoidosis of the female genital tract is extremely rare, with the most common site of involvement being the uterus (2, 4). One of the major concerns of female genital sarcoidosis is its differentiation from other lesions like tuberculosis. Amenorrhoea, menorrhagia, metrorrhagia, and postmenopausal menorrhagia are clinical manifestations of female genital sarcoidosis (4). Cervix and fallopian involvement are also reported (4). Cases of reported ovarian sarcoidosis are even rarer (1).

The unusual clinical presentation of the disease, and the imaging features mimicking ovarian malignancies, present a problem in the differential diagnosis for an ovarian mass.

A majority of the cases of ovarian sarcoidosis were diagnosed after laparoscopy (1, 5). Our case is one of the few cases of ovarian sarcoidosis that was diagnosed via a non-surgical approach.

Sarcoidosis spontaneously remits in up to 33% of patients (6). During the period of evaluation, we were able to recognize the specific regression of the adnexal mass over time; this caused us to reconsider our differential diagnosis of this patient's ovarian mass. Spontaneous regression of solid ovarian malignancies is a rare (7, 8) and extremely unexpected situation. As far as we know, among differential diagnosis of solid ovarian malignancies, spontaneous regression is limited only to germinal ovarian tumors in the reported English literature (7, 8). Regression of ovarian masses may be a distinctive feature, and clinicians may use it to evaluate ovarian masses in the future.

To date, no specific radiological findings best describe the occurrence of ovarian sarcoidosis, nor are there any nuclear medicine findings specific to the diagnosis. FDG uptake in sarcoidosis, for example, is nonspecific in both intensity and pattern. In fact, the FDG uptake pattern of ovarian sarcoidosis can mimic other processes, including that of (but not limited to) ovarian malignancy, diffuse metastatic disease, and lymphoma (3, 6).

Given that sarcoidosis frequently involves multiple organ systems, familiarity with the radiologic and clinical features of the disease in various organs can play a crucial role in its diagnosis and management for patients afflicted with the disease. While ovarian sarcoidosis is rarely reported in the literature, we speculate that ovarian sarcoidosis may be more common than we think, especially in the diagnostic workup of patients with ovarian masses. We also intend to remind clinicians that, in the differential diagnosis of solid ovarian masses, sarcoidosis should be considered despite its reported rarity within the published literature.

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