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Case Report

Sarcoidosis presenting as ovarian mass with omental and peritoneal nodules with elevated CA 125 level; mimicking malignancy

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ABSTRACT

Sarcoidosis is a systemic inflammatory disease of unknown aetiology with a variety of nonspecific clinicoradiological features making diagnosis challenging. Sarcoidosis commonly involves pulmonary and lymphoreticular systems; ovarian involvement being extremely rare. We present a case of a 50-year Indian postmenopausal female, with a history of abdominal pain and distension, referred from the peripheral hospital for management of a solid complex left adnexal mass and elevated CA 125 levels. Based on ultrasonography and contrast-enhanced computed tomography, ovarian carcinoma was a differential diagnosis. Ascitic and pleural fluid cytology and ovarian fine needle aspiration cytology (FNAC) didn't give any conclusive diagnosis. Hence surgical intervention was carried out. Histopathology revealed noncaseating granulomas in ovaries, peritoneum, and omentum. Tissue polymerase chain reaction (PCR) for tuberculous and nontuberculous mycobacteria was negative. Post-operatively serum angiotensin-converting enzyme (ACE) and calcium levels were elevated. These findings supported the diagnosis of sarcoidosis and the patient responded well to systemic steroids.

Keywords: CA 125, Omental nodules, Ovarian mass, Sarcoidosis

INTRODUCTION

Genital tract sarcoidosis is rare; the most common organ involved being the uterus. Ovarian involvement is extremely rare.[1] Wuntakal et al mentioned seven cases of ovarian involvement in English literature.[2] Ovarian sarcoidosis may be misdiagnosed as a malignancy on radiology. Even peritoneal involvement can present as ascites, peritoneal thickening, and nodularity, mimicking peritoneal carcinomatosis and creating a dilemma.[2]

Diagnosis of sarcoidosis can be suspected with awareness to clinicoradiological findings and histopathological evidence of non-caseating epithelioid granulomas with the exclusion of other granulomatous conditions with the support of serological tests like serum ACE levels.[3,4]

CASE REPORT

A 50-year Indian post-menopausal female G3P3 with a history of abdominal pain and distension for 2 months was referred from the peripheral hospital for management of a complex solid left adnexal mass.

Ultrasonography of abdomen and pelvis revealed a well-defined 4.4 cm; heterogeneously hypoechoic lesion in left adnexa adherent to the pelvic wall, possibly of neoplastic etiology. Omentum was nodular with moderate ascites, suspecting metastatic deposits. Serum CA 125 levels were 383 U/ml while carcinoembryonic antigen (CEA) level was 1.9 ng/ml. All baseline blood tests were normal.

Contrast enhanced computed tomography (CECT) abdomen and pelvis demonstrated 4 cm heterogeneously

enhancing mass in left adnexa with enhancing peritoneal thickening and nodularity predominantly in the pelvis, most likely suggestive of the metastatic process. Omentum showed fat stranding and nodularity. Computed tomography (CT) thorax revealed right side mild pleural effusion, fibrotic changes with collapse involving middle lobe and fibrotic nodules and stands in the left upper lobe, suggesting a sequel to old infection.

Pleural and repeated ascitic fluid cytology for malignant cells revealed lymphocytes and reactive mesothelial cells. Ultrasonography guided FNAC from the left adnexa revealed plenty of mononuclear inflammatory cells only.

Due to suspicion of malignancy, the patient was planned for surgical intervention. Total abdominal hysterectomy with bilateral salpingooophorectomy, omentectomy and peritonectomy was performed.

Histopathology revealed noncaseating epithelioid granulomatous inflammation involving bilateral ovaries, fallopian tubes, parametria, omentum and peritoneum (Figure 1). The granulomas showed absent/minimal lymphocytic cuffing. Special stain for acid-fast bacilli was negative. No foreign body/fungal organisms were identified. Section from uterus and cervix were unremarkable.

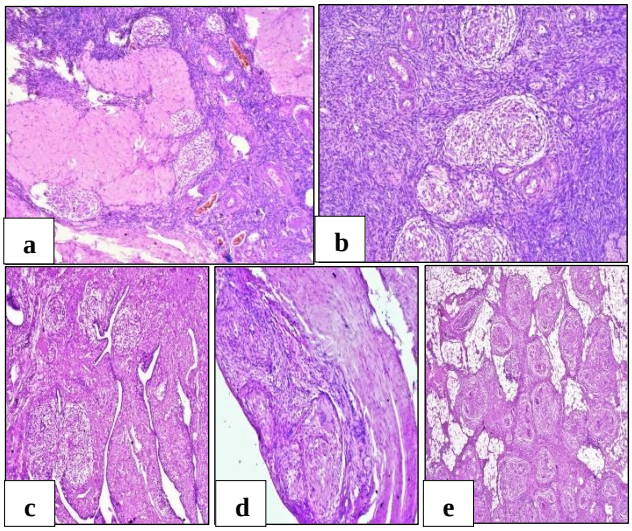

Figure 1: (a) and (b) Ovary showing multiple non-caseating granulomas (H and E, 40x), (c) and (d) fallopian tube and peritoneum showing similar non-caseating granulomas (H and E, 40x), and (e) omentum studded multiple such granulomas (H and E, 10x).

Tissue PCR for *Mycobacterium tuberculosis* and non-tuberculous mycobacteria was negative. Our attention was then directed to an inflammatory disorder. Postoperatively patient was evaluated for serum ACE levels (73.3 U/L). Serum anti-nuclear antibodies were negative. An elevated erythrocyte sedimentation rate (44 mm/hour) was noted. Serum calcium levels were high (12.3 mg/dl). Based on these findings, we arrived at the extremely rare diagnosis of isolated sarcoidosis of ovaries, fallopian tubes and peritoneum.

The patient was started on steroids and responded well to treatment. The patient has followed up for 4 months with good symptomatic relief.

DISCUSSION

Correct diagnosis and differentiation of peritoneal sarcoidosis from ovarian carcinoma and peritoneal tuberculosis in middle-aged females is significantly challenging. The onset of sarcoidosis is most commonly seen between 20 to 40 years of age; though it has been occasionally reported in the elderly and childhood. Sarcoidosis can affect males and females of all races but it is more prevalent in European, African-American, Swedish and Danish individuals, but rare in Indians, possibly due to lack of screening programs and predominance of other common granulomatous diseases like tuberculosis, fungal infections and leprosy.[1,5]

Ovarian sarcoidosis presenting as adnexal mass is extremely rare whether as a component of systemic disease or as an isolated feature.[1] Most cases occurred in the reproductive age group, although a few have been reported in postmenopausal women.[1] Presenting symptoms of ovarian sarcoidosis can be nonspecific like fever, malaise, abdominal pain, or can show symptoms of ovarian tumors, such as adnexal mass, ascites, peritoneal nodules and omental thickening, as seen in our case.[6] Involvement of fallopian tubes occurs in association with sarcoidosis of other parts of the female genital tract.

CA 125 can be seen elevated in ovarian tumors and some nongynecological malignancies and benign conditions as well.[6] In sarcoidosis CA 125 elevation is probably from increased production by epithelioid cells or by peritoneal cells in response to inflammatory mediators secreted by sarcoid granulomas.[6] Our case also had an elevated CA 125 level (383 U/ml).

Epithelioid cells in sarcoid granulomas are also thought to produce ACE, leading to raised serum ACE levels in approximately 60% cases, proportionate to granuloma load, indicating sarcoid activity.[4] In a study by Ungprasert et al, the sensitivity and specificity of elevated ACE levels for sarcoid diagnosis were 41.4% and 89.9% respectively.[7] In our case, ACE levels were elevated (73.3 U/l) even postoperatively.

An association between sarcoidosis and hypercalcemia has been noted in 5-10% of cases.[2] The calcium levels may fluctuate depending on the disease activity. In our case, calcium levels were raised even in the postoperative period (12.3 mg/dl). Hypercalcemia primarily results from increased synthesis of 1,25dihydroxy vitamin D3 and parathyroid hormone-related protein in sarcoid granulomas.[1,8]

According to the American Thoracic Society Clinical Practice Guidelines diagnosis of sarcoidosis is not standardized, and is based on three major criteria: a compatible clinical presentation, non-necrotizing granulomatous inflammation, and exclusion of alternative causes of granulomatous disease.[10] Non-necrotizing epithelioid cell granulomas are characteristic but not a specific finding. Many infections like tuberculosis, atypical mycobacteria, fungi, parasites and non-infectious etiologies including foreign body granulomas, Crohn's disease, medications, lymphomas, and post-surgical granulomas can show similar granulomas mimicking sarcoidosis.[9,10] Hence it is important to rule out commoner conditions with different treatment protocols, before arriving at a diagnosis of sarcoidosis.[10] Administration of systemic steroids can be hazardous if the underlying cause is infection. Wuntakal et al demonstrated the importance of FNAC and biopsy in the case of ovarian sarcoidosis with a multidisciplinary team approach for management to avoid unnecessary surgery.[1]

CONCLUSION

To conclude, sarcoidosis of ovaries is very rare and presentation and clinicoradiological findings can mimic malignancy. Identification of non-caseating granulomas and excluding other causes of granulomatous diseases is crucial in diagnosis. Serum ACE levels may be supportive. Hence we also want to suggest that though sarcoidosis of ovaries is rare, it should also be considered in the preoperative differential diagnosis of ovarian masses so that extensive surgical interventions can be avoided.

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