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Case Report

Xanthogranulomatous Salpingitis in a Case of Mullerian Duct Anomaly and Ipsilateral Renal Agenesis (OVHIRA Syndrome): A Rare Case Report

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Abstract

Xanthogranulomatous inflammation is described as the presence of lipid-laden foamy macrophages admixed with other inflammatory cells, however, it being present in the female genital tract is rather uncommon. OVHIRA syndrome is a rare Mullerian duct anomaly associated with hemi-vagina which obstructs the free flow of menstrual blood, hence creating a compatible environment for the growth of bacteria like *E. coli*. We report a rare case of an 11-year old female with OVHIRA syndrome who developed bilateral Xanthogranulomatous Salpingitis. This report is the first case of its kind in literature, to the best of our knowledge.

Keywords: *Xanthogranulomatous Salpingitis; OVHIRA Syndrome; E. coli; Fallopian Tubes; Hematosalpinx; Hematocolpos*

Introduction

Xanthogranulomatous inflammation in the gall bladder, kidney and urinary bladder are well described and studied entities. The female genital tract, however, has been described as a rare site for this type of inflammation, with endometrium being the most common location according to various reports. In literature, only a few cases involving the ovary and fallopian tube have been described [1]. Xanthogranulomatous inflammation is commonly characterized by the accumulation of lipid-laden foamy macrophages (foamy histiocytes with small dark nuclei and clear cytoplasm) intermixed with lymphocytes, giant cells, and plasma cells [2].

We report the case of an 11-year old patient who presented with premenstrual dysmenorrhea and symptoms of cyclical lower abdominal pain.

Case Report

An 11-year old female presented with the chief complaints of lower abdominal pain, which was cyclical in nature and radiated to the lower back, for the past 1 year. The patient also experienced severe dysmenorrhea. On examination, the abdomen was found to be distended corresponding with 16 weeks of gestation. Ultrasonographic examination showed a large 75 x 58 mm hypoechoic collection in the lower uterine segment extending into the cervical canal indicating hematometra along with vaginal distention. CECT abdomen, showed hematometra along with bilateral hydrosalpinx with mild right hydroureteronephrosis due to compression of the lower ureter by hematosalpinx. The CECT also revealed an absent left kidney. The MRI pelvis performed, revealed an obstructed hemivagina with a transverse

vaginal septum, along with hematocolpos, hematometra, bilateral hematosalpinx, and absent left kidney (Figure 1a and 1b). Vaginal canalization and septal removal procedure with hematometra drainage was done. Microscopy of the vaginal tissue and the septum showed fibromuscular tissue with mild inflammation and congestion. The cervical tissue showed acute inflammatory exudate. This was followed by mold insertion, 5 days after which the patient developed high-grade fever along with per abdominal tenderness and foul-smelling discharge per vaginum. A USG performed thereafter, showed prominent small bowel loops with mild ascites following which an exploratory laparotomy was done. During the laparotomy, the bowel loops were found to be adhered to the uterus and bilateral fallopian tubes. Hence, partial colectomy with a subtotal hysterectomy and bilateral salpingectomy was done. Grossly, both the tubes were distended, which on cut showed the presence of grey-brown mass measuring 0.5 x 0.3 x 0.2 cm in the left tube and 0.6 x 0.4 x 0.1cm in the right tube. The bowel walls were gangrenous with necrosis on microscopy. Microscopy of the endometrium showed proliferative endometrium with unremarkable myometrium. The bilateral tubes showed dense chronic inflammation comprising of lymphocytes, plasma cells with sheets of lipid-laden foamy macrophages, and few neutrophils along the walls of the tubes with congested blood vessels (Figure 1c and 1d). Hence, based on the clinical, radiological, and histopathological findings, the final diagnosis of bilateral Xanthogranulomatous salpingitis (XGS) in a case of OVHIRA syndrome was given.

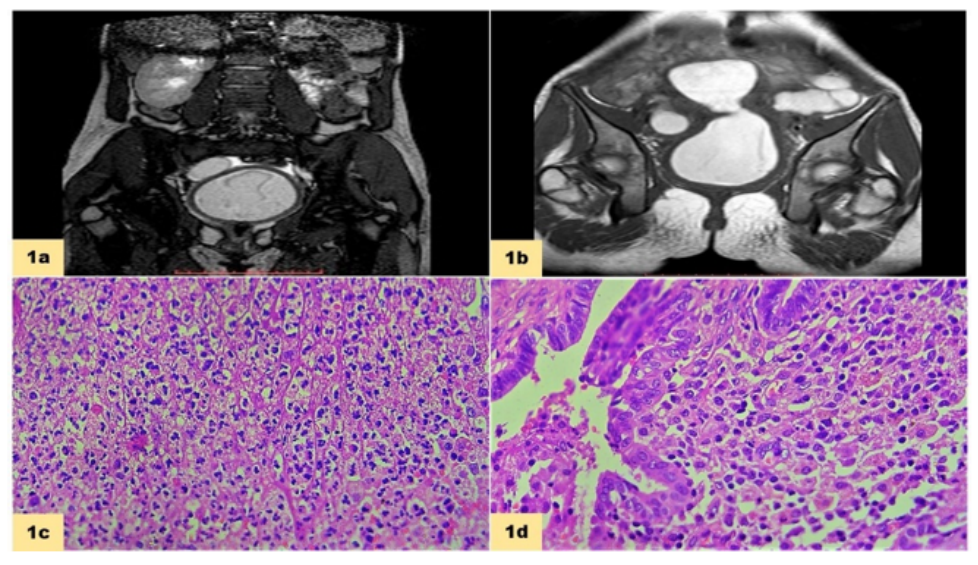

Figure 1: (a): *Coronal BFFE MRI image of abdomen showing absent left kidney, dilated right fallopian tube and cystic structure in pelvis suggestive of a distally obstructed, distended vaginal cavity. (b): Coronal T1W MRI image of pelvis showing hyperintense blood distending bilateral fallopian tubes, uterine cavity and vagina suggestive of hematosalpinx, hematometra and hematocolpos. (c): Section from fallopian tube, showing massive infiltration by foamy macrophages in sheets along with lymphocytes, plasma cells and few neutrophils. (H&E x10). (d): Infiltration of plicae and fibrovascular core of the fallopian tube by foamy macrophages, plasma cells and lymphocytes. (H&E x40).*

Discussion

Xanthogranulomatous inflammation of the female genital system is rare. When involved, fallopian tubes mostly show a unilateral presentation. Occasionally bilateral involvement is seen, similar to our case. It is defined as a form of chronic inflammation showing sheets of foamy lipid-laden macrophages along with plasma cells and neutrophils. The inflammation in the fallopian tube causes thickening of

the wall, which may often appear as a mass-like lesion [3]. As studied in literature only a few cases of xanthogranulomatous inflammation involving the fallopian tubes have been described [4]. The causes for the development of xanthogranulomatous inflammation being multiple, commonly include infection, ineffective antibiotic therapy leading to incomplete clearance of bacteria by phagocytes and occasionally radiotherapy. A prolonged and chronic history of pelvic inflammatory disease and the presence of an IUD is more than often a common cause [5]. *E. coli* is the most common causative organism, followed by *Proteus* spp., *Staphylococcus aureus, Bacteroides fragilis* and *Salmonella typhi* [3]. Our case also showed positivity for *E. coli* on culture of the vaginal discharge fluid.

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare syndrome that is characterized by Mullerian and renal anomalies. Renal agenesis is classically considered to be part of the definition of OHVIRA syndrome [6] similar to our case in which the left kidney was absent. Although there is no established literature of xanthogranulomatous salpingitis in a case of OHVIRA syndrome, the possible cause in our case could be an obstructed hemivagina due to transverse vaginal septum, following which the patient developed hematocolpos and bilateral hematosalpinx. After the septal removal procedure was done, the mold that was inserted along with the collected blood could have acted as a nidus for the growth of *E. coli* bacteria. The prolonged infection then caused tissue necrosis, thereby leading to the release of cholesterol and other lipids and subsequently leading to its phagocytosis by macrophages. To the best of our knowledge, this is the first case report of bilateral XGS in the case of OHVIRA syndrome.

Conclusion

XGS is a rare form of chronic inflammation of the fallopian tube. The association of xanthogranulomatous inflammation with Mullerian duct anomalies like OVHIRA syndrome is very likely owing to the obstructive nature of the syndrome and the development of hematocolpos and hematosalpinx, which can very strongly act as a nidus for infection. Although very rare, OHVIRA syndrome should be evaluated in a female presenting with xanthogranulomatous salpingitis.

Disclosure Statement

The authors report no conflict of interests. This was a case report hence no financial assistance was needed. The authors state that the ideas and views expression are original and not influenced by any official position or institution or funder.

Study Registration

No registration was required as the article submitted is a case report.

Conflict of Interest

The authors report no conflicts of interest.

Ethical Clearance

No ethical clearance was required as the article submitted is a case report.

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