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Xantogranulomatous Salphingoophoritis- A Rare Case Report

Yoga Lakshmi S.K[1], Sudha C.P[2], Savitha T.S[3], Malathi T[4]
[1]Senior Resident in Department of Obstetrics and Gynaecology, KIMS hospital, Bangalore
[2]Professor and unit chief in the department of Obstetrics & Gynaecology, KIMS hospital, Bangalore
[3]Associate Professor in the department of Obstetrics & Gynaecology, KIMS hospital, Bangalore
[4]Assistant Professor in the department of Obstetrics & Gynaecology, KIMS hospital, Bangalore

***Corresponding author**
Yoga Lakshmi. S.K
Email: yoga.manohar@gmail.com

Abstract: Xantogranulomatous inflammation is a form of chronic inflammation that rarely affects the female genital tract especially the fallopian tubes and the ovaries together. To our knowledge only 5 cases of Xantogranulomatous Salphingoophoritis have been reported till date. We report one such case of Xantogranulomatous Salphingoophoritis.
Keywords: Xantogranulomatous inflammation.

INTRODUCTION

Xantogranulomatous inflammation commonly affects organs like kidney and gallbladder [1]. It is destructive to the affected organ and infiltrates surrounding tissues. It is a benign condition which is treated by surgical excision. Proposed etiological factors are IUCD, Endometritis, Inborn errors of lipid metabolism, immunodeficiency and infection by certain bacteria like E.coli, Bacteroides fragilis, Proteus and Salmonella typhi. Clinically patients present with low grade fever, pain abdomen and mass per abdomen. Histopathologically characterised by foamy histiocytes with mixed inflammatory infiltrate.

CASE REPORT

A 35years old married P2L1 female patient presented with complaint of fever with lower abdominal pain since 1 month and irregular cycles for 8 months. Patient had a history of intrauterine fetal demise at 6 months of amenorrhoea 5years prior to this and she was evaluated for secondary infertility and treated for Pelvic.

Inflammatory disease with doxycycline 5months prior to presentation. On examination patient was febrile, had left iliac fossa tenderness and per vaginal examination revealed a bulky uterus with a tender palpable mass in the left fornix. Endocervical swab culture was taken which showed growth of Escherichia coli sensitive to levofloxacin and gentamicin hence patient was treated with these antibiotics. Pelvic USG revealed a Left adnexal complex lesion measuring 7.7×3.5× 6.5 cms with cystic areas, internal echoes, septations and rim vascularity, CT scan was reported as pyogenic tuboovarian abscess adherent to the bowel. Sigmoidoscopy confirmed the infiltration of the bowel.

Investigations revealed Neutrophilic leucocytosis with anaemia, elevated ESR 132 and CA125 -20.

Patient underwent prophylactic left ureteric stenting followed by Diagnostic laparoscopy. Left sided tuboovarian mass densely adherent to the posterior wall of the uterus and rectum was identified hence Laparotomy with Total abdominal hysterectomy with Bilateral Salphinghingoophorectomy, Adhesiolysis and Rectal mucosal repair was performed as there was infiltration of rectum and breech in rectal mucosa occurred as a result of adhesiolysis. Specimen obtained was sent for frozen section which was reported as oedematous fibro collagenous tissue with thick walled blood vessels with focal collection of histiocytes and neutrophils. Histopathology was reported as Xantogranulomatous Salphingoophoritis.

DISCUSSION

Kunakemakorn [2] was the first to describe Xantogranulomatous inflammation of serosa of uterus, left fallopian tube and ovary in his report of inflammatory pseudotumor in the pelvis in 1976.The Xantogranulomatous lesions are unusual forms of chronic inflammatory process, pathologically characterized by the presence of lipid -filled histiocytes with admixed lymphocytes, plasma cells and neutrophils. Most commonly affected organ is kidney, followed by gall bladder [1]. Other organs in which Xantogranulomatous inflammation has been reported are stomach, anorectal area, bone, urinary bladder, testis, epididymis and female genital tract. Xantogranulomatous inflammation of the female genital tract is unusual and is essentially limited to the endometrium but may extend into the myometrium [3].

Till date, only 15 cases of Xantogranulomatous inflammation of the female genital tract are reported in literature out of which there are 7 cases with unilateral ovary, 5 cases with unilateral fallopian tube, 1 Case with bilateral fallopian tube involvement and only 2 cases with ovary and fallopian tube showing simultaneous involvement [4].

Age distribution

A review of the literature done by Margaret et al., revealed that patients ranged from 23-72 years old.

Aetiology

The pathogenesis is unclear and many theories have been postulated, such as theory of infection, associated microorganisms include Actinomyces, Staphylococcus aureus, Streptococcus (Enterococcus) faecalis, E coli, Viridans streptococci, B fragilis, Torulopsis (Candida) glabrata, and group B streptococci proved by tissue culture. Other causes include endometriosis, intrauterine contraceptive device, inborn errors of lipid metabolism, and drug induced, ineffective clearance of bacteria by phagocytes [5].

Pathogenesis

A combination of factors may be responsible. For example, bleeding and obstruction may predispose to infection; tissue necrosis occurs, followed by the release of cholesterol and other lipids and phagocytosis by macrophages. Amongst the theories, the most accepted theory is of infection.

Histologically, foamy histocytes (xanthoma cells), and chronic inflammatory cells are consistently observed.

Xanthoma cells are histiocytes with abundant lipid laden cytoplasm having vacuolated appearance. It is aggregates of such foam cells, which are responsible for the yellow colour observed on gross examination.

The emergence of foam cells may be attributed to the following factors:

- Inefficient or inappropriate antibiotics applied in the early phase of infection that resulted in ineffective control of bacterial multiplication.
- Presence of a lipid metabolic disorder that induces hyperlipidaemia and the foam cells are formed when the lipid deposited is phagocytised by phagocytes.
- The application of intrauterine contraceptive devices or drugs, involvement of adjacent organs and pelvic
- Peritoneum results in adhesions, hence arousing the suspicion of malignancy.

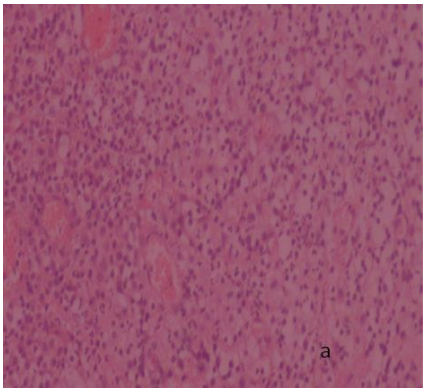

Fig-a: Chronic inflammatory infiltrates

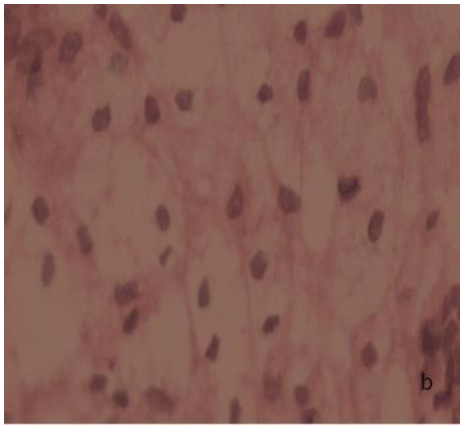

Fig- b: Faomy histocytes

CLINICAL PRESENTATION

Presentation most often includes fever, lower abdominal pain and mass per abdomen, menorrhagia, anaemia, and anorexia. Gynecological examination reveals adnexal mass with tenderness with blood investigations showing elevated ESR and raised white blood cell count. Grossly, the affected ovary is enlarged and appears like a tumor with yellowish appearance, with cystic areas within due to necrosis.

DIFFERENTIAL DIAGNOSIS

Malacoplakia and Xanthogranulomatous pseudotumor of the vagina are closely related entities resulting from infection by gram-negative bacilli, usually Escherichia coli [3].

Malakoplakia is a form of chronic granulomatous inflammation, can involve a variety of organs, most frequently the urinary tract, but it rarely affects the female genital tract. It is recognized by the presence of calcified cytoplasmic or extra cytoplasmic lamellar spherules that is both intracellular and extracellular, concentrically laminated basophilic masses [3] called Michaelis-Gutmann bodies are present in variable numbers.

Xanthogranulomatous pseudotumor of the vagina typically contains yellow polypoidal nodules arise from the vaginal mucosa, at times accompanied by

a discharge. The microscopic findings are identical to those described in other body sites, and include the presence of large collections of histocytes with abundant granular to pale foamy cytoplasm (von Hansemann cells). Walther *et al*. have suggested that malakoplakia and Xanthogranulomatous inflammation may have a common pathogenesis. Malakoplakia is thought to result from a deficit in the ability of phagocytes to kill microorganisms, possibly due to diminished release of b-glucuronidase and low levels of cyclic guanosine monophosphate. Michaelis-Gutmann bodies are thought to arise from phagolysosomes, which contain partly digested bacteria. Infections like tuberculosis, fungal infections which can be ruled out by culture and special stains for the causative organisms.

Xanthogranulomatous oophoritis is often misdiagnosed by pathologists if they don't keep this entity in mind. This may be due to the rarity of the condition. If the lesion is mainly focal scattered lymphocytes, it may be misdiagnosed as secondary lymphoma or leukaemia. If the lymphocytes are scattering diffusely and foam cells are seldom, a diagnosis of malignant small cell tumour with stromal luteinization may be rendered. If there are small amount of obvious fibrosis and foam cells, a diagnosis of sclerosing stromal tumor may be made. However, the right diagnosis is possible to be made as long as pathologists elevate vigilance and master the pathological features. The correct diagnosis may be confirmed by the finding of numerous gram-negative rod-like bacteria on tissue Gram stain, on silver stain, or by electron microscopy [3].Immunohistiochemical stains are helpful in establishing the diagnosis, including CD68 (foam cells positive), CD3 (T lymphocyte marker), CD20 (B lymphocytes marker).Neoplastic lesions should be ruled out with detailed investigations as Xantogranulomatous oophoritis can be easily confused with ovarian malignancy, clinically, radiologically, and pathologically. Immunohistochemistry helps in confirming the diagnosis included, but it is seldom required in the presence of characteristics histopathologi-cal features. Treatment of choice for Xantogranulomatous Salphingoophoritis is Salpingooophorectomy.

CONCLUSION

Though a rare condition, gynaecologists need to consider Xantogranulomatous Salphingoophoritis as one of the differential diagnosis, when a young to middle aged patient presents with a tuboovarian mass and a history of low-grade fever, pain abdomen, PID and infertility, so that appropriate management plan can be made and patients undergo a less radical surgery, hence better prognosis. Neoplastic lesions should be ruled out with detailed investigations as Xantogranulomatous oophoritis can be easily confused with ovarian malignancy, clinically, radiologically, and pathologically. Immunohistochemistry helps in confirming the diagnosis included, but it is seldom required in the presence of characteristics histopathological features. Treatment of choice for Xantogranulomatous Salphingoophoritis is Salpingooophorectomy. Antibiotic therapy has been attempted, but it has not succeeded in reducing ovarian mass.

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