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Calcified Bilateral Ovarian Fibroma in a 15 Year Old Female: Case Report and Literature Review

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Authors' contributions

This work was carried out in collaboration among all authors. Authors AMA and JOI assessed the patient clinically and performed laparotomy, they also wrote the initial draft of the manuscript with substantial contribution from AOY and MAA. AOY performed the radiologic diagnosis while MAA performed the histologic diagnosis. All authors read and approved the final draft of the manuscript and certified it for publication.

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Case Report

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ABSTRACT

Aim: To highlight the potential for misdiagnosis of ovarian fibromas and need for careful evaluation especially when fertility altering decisions need to be taken in the young adolescent.
Presentation of Case: The authors here review literature and present the case of a 15 year old pre-menarchal patient with bilateral, solid hard ovarian tumors with marked ascites, who had bilateral salpingo-oophorectomy, in whom the tumors turned out to be bilateral calcific ovarian fibromas.
Discussion: Ovarian neoplasia are often misdiagnosed because of their non-specific symptoms and similarities to other pathologies on radiological imaging. The management of adolescents with ovarian tumors poses peculiar challenges as there is need for a balance between the risk of

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malignancy and the need to preserve fertility. About 1/3 of pelvic masses in pre-pubertal girls are malignant. This fact in addition to the non-specific features of malignancy in this patient such as weight loss, ascites, necessitated further evaluation which included laparotomy.
Conclusion: Ovarian fibromas occur in adolescents and can pose a diagnostic dilemma; a high index of suspicion is required to plan fertility-sparing and cancer-limiting management.

Keywords: Cancer; fertility; fibroid; fibroma; ovary; tumors.

1. INTRODUCTION

Ovarian neoplasia present with non-specific symptoms that can be due to pathologies in other abdominal or pelvic organs, thus posing diagnostic dilemma and delay in definitive care in many instances. Lower abdominal or pelvic pain is one of the earliest complaints; chronic pain is often the commoner presentation than acute abdominal pain [1]. Chronic pain has however a higher likelihood to be associated with malignant lesions of the ovary [2]. Precocious puberty may be the first sign in some pre-menarchal girls due to hormone-producing functional neoplasms [3]. The adolescent may also present with abnormal uterine bleeding or dysmenorrhea [4]. Tumors can also be classified as Surface epithelial tumors (serous, mucinous, endometroid, clear cell, and transitional cell), Sex cord stromal tumors (granulosa, thecoma, Fibroma, Sertoli cell, Sertoli-Leydig, Steroid) and Germ cell tumors (dysgerminoma, yolk sac, embryonal carcinoma, choriocarcinoma, teratoma) [5].

The sex cord stromal tumors are further sub-classified into: pure stromal tumors (Fibroma, cellular fibroma, thecoma, fibrosarcoma, sclerosing stromal tumor, Leydig cell tumor, steroid cell tumor), pure sex cord tumors(adult granulosa cell tumor, juvenile granulosa cell tumor, Sertoli cell tumor) and Mixed sex cord-stromal tumors(Sertoli -Leydig cell tumors) [6].

Ovarian Fibroma was first mentioned in medical literature by J. Astrue in 1743 [7,8]. They are sex cord stromal tumors of the pure stromal variety; they originate from excessive growth of the stroma and connective tissue of the cortex of the ovary. Fibromas and thecomas can coexist in the same tumor as fibrothecomas [6,9].

Ovarian fibromas can either be cystic or solid although, cystic tumors are commoner while solid tumors are rare. Ovarian fibromas are the most common solid ovarian neoplasm and accounts for 1-4.7% of all ovarian tumours [10]. They are mostly benign however focal fibro-sarcomatous changes have been reported in less than 1% of cases [8].

These tumors are commonly seen in peri-menopausal and post-menopausal women (median age of 48 years) and are rare in children [8]. Less than 10% of ovarian fibromas are seen in individuals younger than 30 years of age [9]. They can be unilateral or bilateral; the tumors are unilateral in 90% of cases 70% of which occur on the left [11-13].

Ovarian tumors especially fibromas are uncommon in pre-menarchial adolescents, often misdiagnosed, resulting in delayed treatment and their management have potential to alter reproductive life of the patients. We here we present the case of a 15 year old adolescent with bilateral calcified ovarian fibroma.

2. PRESENTATION OF CASE

The patient is a 15 year old nulliparous female who presented to the general surgery outpatient clinic of the Babcock University Teaching Hospital, with lower abdominal mass of 10 years duration, which had progressively increased in size. It was initially painless however 7 years before presentation she developed intermittent dull aching pain over the mass and the severity of the pain increased for about 2 months before presentation. She was yet to attain menarche, not sexually active and had no gastrointestinal or urinary symptoms.

On examination, she looked well but in intermittent painful distress, she had no gross feature suggestive of congenital anomaly or other recognized syndromes.

Abdominal examination revealed a 16 week size suprapubic mass arising from the pelvis. The mass was hard, nodular, tender and not mobile, there was marked ascites. Rectal examination showed an extra-luminal hard nodular mass in the anterior wall of the rectum. Plain abdominal X-ray showed multiple calcific oval shaped masses in the pelvis (Fig. 1).

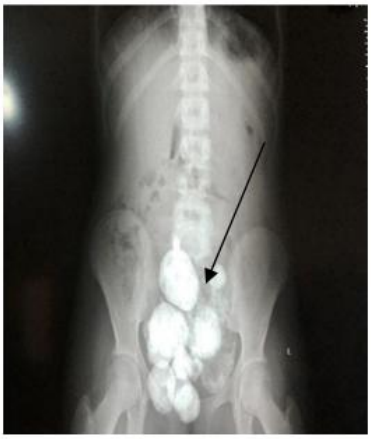


Fig. 1. Plain abdominal X-ray showed multiple calcified oval shaped masses (arrow) in the pelvis

Abdomino-pelvic ultrasound scanning report revealed multiple round calcified pelvic masses extending to the lower aspect of the anterior abdominal wall. The uterus was hypoplastic and both ovaries could not be visualized separately from the mass. The chest X-ray revealed essentially normal findings.

Computerized tomography (CT) scan of the abdomen and pelvis showed multiple oval shaped calcified masses of various sizes located in the pelvis and extending to the lower anterior abdominal wall. A hypoplastic uterus was also seen, but both ovaries could not be independently observed. Intravenous Urogram (IVU) revealed normal kidneys and urinary tract. She had CA-125 and CEA done and the levels of both tumor markers were <3.5 MIU/ml, which are normal for laboratory reference range. Exploratory laparotomy was done and intra-operative findings included; 1.5 liters of serous ascites, bilateral hard, nodular and pedunculated ovarian tumor which measured 13 cm in the widest diameter on the right and 12 cm on the left. No normal ovarian tissue could be grossly visualized in the tumor on both sides (Fig. 2).

Fig. 2. Exploratory laparotomy procedure: Findings at exploratory laparotomy

The uterus was grossly normal morphologically, but hypoplastic in size. The fallopian tubes, liver and the intestine were grossly normal and no enlarged lymph node was seen.

She had bilateral oophorectomy and drainage of the ascites. The ovaries could not be spared because the tumor has taken up both ovaries and no normal tissue could be seen (Fig. 3).

She made satisfactory clinical progress postoperatively. Histology of the right and left ovarian masses showed proliferating spindle cells disposed in a whorled arrangement. The tumour cells have bland wavy nuclei and moderate eosinophilic cytoplasm admixed with variable amount of extracellular collagens. Focal areas of dystrophic calcification were seen. Features were consistent with bilateral ovarian Fibroma (Figs. 4 and 5). She remained in good clinical condition with no complains at the last outpatient review which was 1 year after surgery.

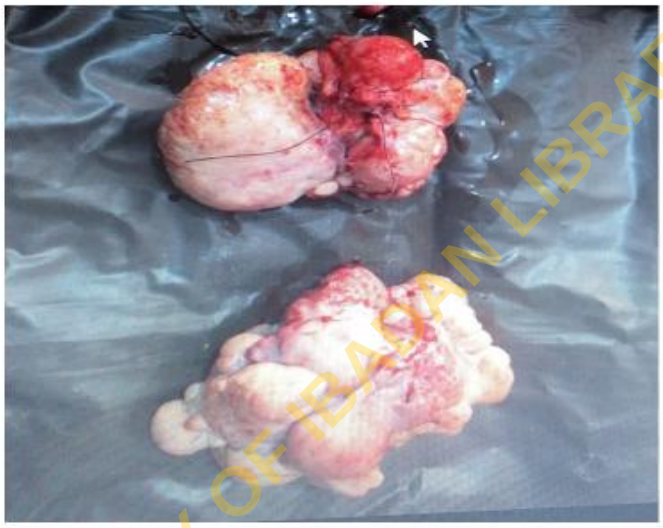
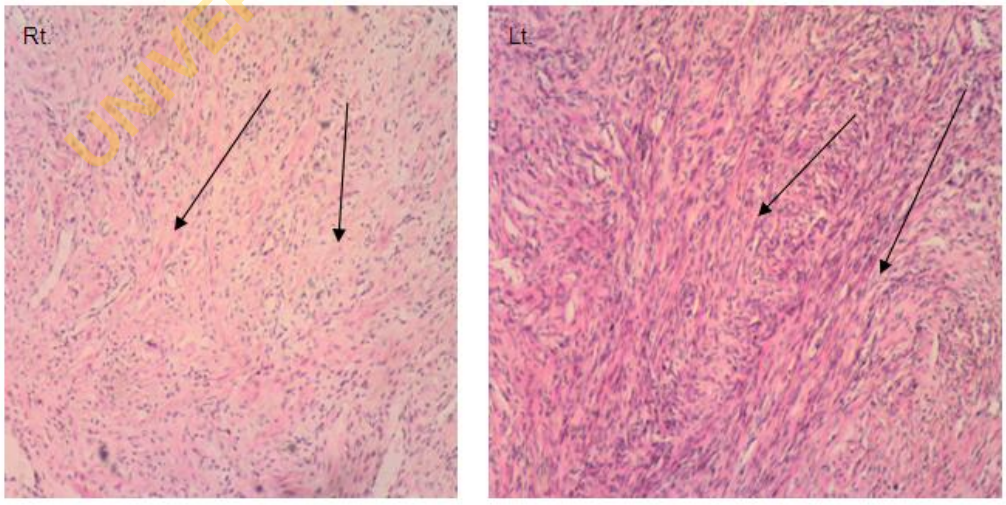


Fig. 3. Right and left ovaries with gross multiple tumor nodules that were firm to hard in consistency



Figs. 4 and 5. Photomicrograph from the right (Rt.) and left (Lt.) ovarian masses showed proliferating spindle cells disposed in a whorled arrangement with variable amount of extracellular collagen (arrows). (Haematoxylin and eosin, X100)

3. DISCUSSION

The expected quality of life from the benign nature of ovarian fibromas can be marred by diagnostic dilemma encountered in their management, with a high risk of decisions which may adversely affect future reproductive prospects. They are mostly misdiagnosed as uterine leiomyomata and sometimes as malignant ovarian tumors due to their appearance and the presence of ascites and elevated CA 125; features which are not tumor specific. They are the most common solid ovarian neoplasm. These tumours may be non-functional or functional (which are usually associated with endocrine abnormalities, especially due to oestrogen secretion) [11].

The patients with ovarian fibroma typically present with palpable abdominal masses, abdominal distension, lower abdominal discomfort, lower abdominal pain/pelvic pain especially in pedunculated tumors that have undergone torsion. They may also present with menstrual abnormalities, which include, inter-menstrual bleeding and metrorrhagia, which are due to hormonal changes associated with functional tumors. The ovary may be completely taken over by the fibrous tumor such that it no longer undertakes any hormonal function, resulting in primary amenorrhea in the pre-pubertal girl, as found in this case presentation. The pressure effects on surrounding structures may give lower gastro-intestinal and urinary symptoms. However it can be asymptomatic and may only be an incidental finding on routine gynecologic examination and evaluation, therefore a high index of suspicion is required for accurate pre-operative diagnosis [13,14]. In this case report the patient had an incidental ultrasound scanning finding of an ovarian mass as a pre-pubertal girl, which had progressively increased in size and become symptomatic overtime. Her tumor had increased in size to 13 cm in the widest diameter on the right and 12 cm diameter on the left observed at laparotomy, with associated bilateral adnexal torsion. Reports from studies reveal that 5-35% of pelvic masses in pre-pubescent girls are malignant [15,16]. This in addition to the non-specific features of malignancy; weight loss, ascites, necessitated further evaluation which included laparotomy.

Ovarian fibromas can be associated with ascites especially in large tumors in which about 40-50% of tumors >5 cm in widest diameter present with ascites, the fluid usually escapes from its edematous surface [13,14]. The Ovarian cortex, from which the fibroma arises, does not have lymphatic vessels, this makes large amount of fluid to escape from large tumors [13]. The patient presented had bilateral large ovarian tumors with associated ascites.

Radiological imaging techniques are the modalities most frequently used before surgery; plain abdominal radiograph detects areas of calcification in the tumor as observed in the patient being presented. This modality is limited in use when calcification is absent. Ultrasound scanning shows ovarian fibromas as having hypoechoic solid masses with significant posterior shadowing on ultrasound [8]. However mixed echogenic appearance with isoechoic to hypoechoic features can also be seen due to edema and cystic degeneration of the tumor [17]. Ultrasound scanning can be used to differentiate these tumors from uterine fibroid by the appearance of a pelvic mass that is close to but not directly connected to the uterus and non-visualization of a normal ovary on the affected side. Doppler ultrasound will also show a poorly vascularized tumor with low velocity flow in ovarian fibroma as opposed to uterine fibroid [8].

CT scan appearance of ovarian fibroma is that of solid ovarian masses, homogenous in appearance with delayed contrast enhancement [6]. Although pathological changes such as necrosis, infarction, hemorrhage, degeneration and calcification may affect and alter the overall appearance [15]. The patient presented has significant calcification of the tumor, which caused an increase in weight of the tumor, causing recurrent torsion and made it appear hard to touch suggesting malignancy during laparotomy.

On Magnetic Resonance Imaging (MRI), ovarian fibromas usually appear as marked T1 and T2 weighted hypo-densities. There can also be delayed enhancement on gadolinium administration [6,10].

The tumor marker CA 125 levels are usually normal or mildly elevated in ovarian fibroma but very high levels are rare [7]. Higher levels are mostly seen in women with ascites and these usually resolve after removal of the tumor [13]. The patient in this case report had normal CA-125 levels.

The diagnosis can only be confirmed on histology. Microscopically these tumors consist of fibroblastic cells that are spindle shaped with

cytoplasm producing abundant collagen [8,9,14] however fibrothecoma contains a small proportion of theca cells containing intracellular lipids in addition to the spindle shaped fibroblastic cells and this may show estrogenic activity [17].

Fibromas can occur as an isolated lesion or as part of a syndrome. These syndromes include Meig Syndrome, where the benign ovarian fibroma is associated with ascites and unilateral right sided pleural effusion and this is seen in 1% of ovarian fibromas [14]. Others include; Gorlin-Golt syndrome is an uncommon multi-systemic autosomal dominant disorder also known as Nevoid basal cell carcinoma and its features includes Bilateral Calcified Ovarian fibromas, multiple basal cell carcinoma of the skin, keratocystic odontogenic tumours, anomalies of the vertebral and skull, calcified dural fold, hypertelorism, cardiac fibromas, fetal rhabdomyomas and rhabdomyosarcomas [7,12]. About 75% of calcified bilateral ovarian fibromas are associated with Gorlin-Golt syndrome [12], however the patient presented did not have any of these syndromic features. Ovarian fibromas have also been described in association with Maffucci syndrome. This is an anomaly in which there is widespread mesodermal dysplasia associated with hemangiomas or lymphangiomas [18]. In Soto's syndrome, there is excessive growth with macrocephaly, cerebral gigantism, dolichocephaly and delay in developmental milestones in addition to other congenital anomalies [11,12]. Other syndromic associations includes Peutz-Jeghers syndrome, Gardner and Richard Syndrome with Familial polyposis [7]. The patient presented did not have any gross or imaging feature suggestive of these syndromes.

The treatment of choice for ovarian fibroma is surgery and this includes surgical excision or oophorectomy. Although laparoscopy has been shown to be beneficial in adolescents especially for small and medium sized tumors [7,13], laparotomy was done in this patient because of the large size of the tumor). Age and fertility prospects are important factors that determine definitive surgery for ovarian fibroma, while tumor excision and ovarian conservation is the preferred goal for younger patients, salpingo-oophorectomy is advisable for peri-menopausal and post-menopausal women [7,8]. The patient in this report however had bilateral salpingo-oophorectomy because the entire ovary was diseased and there was no gross healthy ovarian tissue to spare, more so the tumor was bilateral, she had ascites and the malignant potential of the tumor could not be ascertained at laparotomy. She had since been placed on hormone replacement therapy.

4. CONCLUSION

Ovarian fibromas are rare benign solid tumors of the ovary commonly seen in peri-menopausal women and rare in children. Pre-operative diagnosis is usually difficult as this tumor can mimic uterine fibroid and other gynecological tumors. Radiological imaging is the major diagnostic modality confirmed by histology. The treatment of choice is tumor excision and conservation of healthy ovarian tissue which can be done either by laparotomy or laparoscopy.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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