***Case report***

# Calcified Bilateral Ovarian Fibroma in a 15 year old Female: Case report and Literature review

.


## 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-ovariectomy, in whom the tumors turned out to be bilateral calcific ovarian fibromas.
**Discussion**: Ovarian neoplasia is often misdiagnosed because of their non-specific symptoms and similarities to other pathologies on radiological imaging. Their management in adolescents poses a peculiar challenge as a balance needs to be reached between the risk of malignancy and the need to preserve fertility. About 1/3 of pelvic masses in pre-pubescent 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 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 had 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 in, 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 weeks 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 (Figure 1).

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.
She had exploratory laparotomy in conjunction with the gynecology team, intra-operative finding included 1.5 liters of serous ascites, bilateral hard, nodular and pedunculated ovarian tumor which measured13cm in the widest diameter on the right and 12cm on the left. No normal ovarian tissue could be grossly visualized in the tumor on both sides (Figure 2). 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 (Figure 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 (Figures 4 & 5). She remained in good clinical condition with no complains at the last outpatient review which was 1 year after surgery.

## 3. DISCUSSION

Ovarian fibromas are benign tumors which pose a difficult diagnosis pre-operatively. They are mostly misdiagnosed as uterine myoma and sometimes malignant ovarian tumor due to their appearance and the presence of ascites and elevated CA 125. They are the most common solid ovarian neoplasm. These tumours may be functional or non-functional. The functional tumours 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. The ovary may be completely taken over by the fibrous tumor such that it no longer undertakes any hormonal function, resulting in primary amenorrhea if this occurs 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 13cm in the widest diameter on the right and 12cm 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 >5cm in widest diameter present with ascites, the fluid usually escapes from its edematous surface [13,14]. The Ovarian cortex where fibroma arises from does not have lymphatic vessels, this makes large amount of fluid to escape from large tumors [13]. The patient presented has a large tumor 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 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 tumorectomy. 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 tumorectomy is the preferred goal for younger patients, salpingo-oophorectomy is advisable for the perimenopausal and post-menopausal women [7,8]. The patient in this report however had bilateral salpingo-ovariectomy 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 tumorectomy or surgical excision which can be done by laparotomy or laparoscopically.

## COMPETING INTERESTS

Authors have declared that no competing interests exist, as regards this article.

## 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.

## REFERENCES


1. Spinelli C, Pucci V, Strambi S, et al. Treatment of ovarian lesions in children and adolescents: a retrospective study of 130 cases. Pediatr Hematol Oncol. 2015;32(3):199-206.
2. Gonzalez DO, Cooper JN, Aldrink JH, et al. Variability in surgical management of benign ovarian neoplasms in children. J Pediatr Surg. 2017;52(6)944-950.
3. Kelleher CM, Goldstein AM. Adnexal masses in Children and Adolescents. Clin Obstet Gynecol. 2015;58 (1):76-92.
4. Liu H, Wang X, Lu D, et al. Ovarian masses in children and adolescents in China: analysis of 203 cases. J Ovarian Res. 2013;6:47-52.
5. Chen VW, Ruiz B, Killeen JL, Cote TR, Wu XC, Correa CN. Pathology and Classification of Ovarian tumors. CANCER. 2003 (97);10 :2631-2642
6. Horta M, Cunha TM. Sex cord stromal tumors of the ovary: a comprehensive review and update for radiologists. Diagn Interv Radiol. 2015. 21(4):277-286
7. Kouach J, El Fadel FA, Moukit M, El Hassani ME, Rahali DM, Dehayni M et al. Bilateral Ovarian Fibroma. A case report and brief literature review. Eur J Pharm Med Res. 2016,3 (12), 554-555
8. Leung SW, Yuen PM. Ovarian Fibroma: A review on the clinical characteristics, Diagnostic Difficulties and Management options of 23 cases. Gynecol Obstet Invest 2006; 62 :1-6
9. Singh V, Mishra B, Sinha S. A Rare case of Ovarian Fibroma in a Teenage Girl. Journal of South Asian Federation of Obstetrics and Gynaecology. 2017; 9(2): 131-133
10. Cho YJ, Lee HS, Kim JM, Joo KY, Kim M. Clinical characteristics and Surgical Management options for ovarian fibroma/fibrothecoma: A study of 97 cases .Gynecol Obstet Invest 2013 ; 76:182-187
11. Silvanesaratman V, Dutta R, Jayalakshmi P. Ovarian Fibroma, clinical and histopathological characteristics. Int J Gynecol Obstet , 1990; 33 (3):243-247
12. Kasapoglu I, Turk P, Atalay F, Uncu G. Lapararoscopic Surgery for Bilateral Multiple Calcified Ovarian Fibromas: A case report. Reprod Syst Sex Disord. 2017.6(4):218. 1-4.
13. Son CE, Choi JS, Lee JH, Jeon SW, Hong JH, Bae JW. Laparoscopic Surgical Management and Clinical Characteristics of Ovarian Fibromas. JSLS J Soc Laparoend. 2011. 15(1);16-20
14. Parwate NS, Patel SM, Arora R, Gupta M. Ovarian fibroma: A clinic-pathological study of 23 cases with review of Literature. J Obstet Gynaecolo India. 2016. 66(6): 460-465
15. Schultz KA, Ness KK, Nagarajan R, Steiner ME. Adnexal masses in infancy and childhood. *Clin Obstet Gynecol.* 2006;49(3):464-479.
16. Stepanian M, Cohn DE. Gynecologic malignancies in adolescents. *Adolesc Med Clin.* 2004;15(3):549-568.
17. Yen P, Khong K, Lamba R, Corwin MT, Gerscovich EO. Ovarian Fibromas and Fibrothecomas - Sonographic correlation with Computed tomography and magnetic resonance imaging: a 5-year single-institution experience. J Ultrasound Med.2013. 32 (1): 13-8
18. Christman JE, Ballon SC. Ovarian Fibrosarcoma associated with Maffuci syndrome. Gynaecol Oncol. 1990. 37(2)290-291

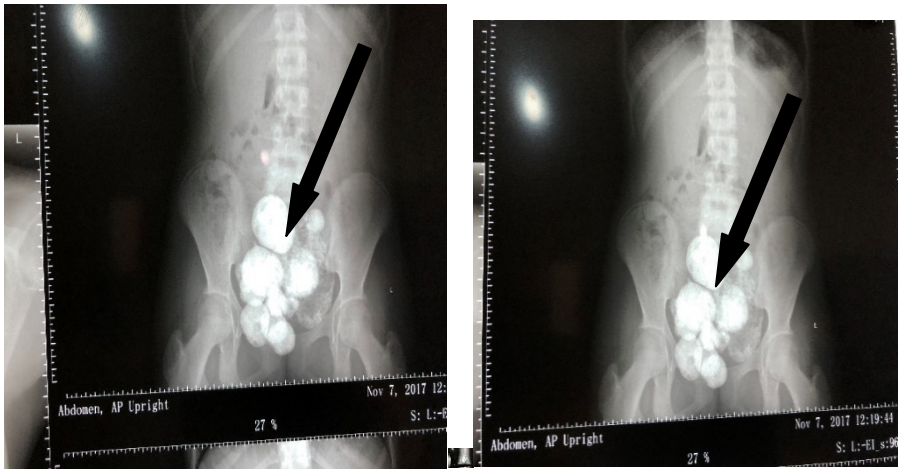


**Figure 1:** Plain abdominal X-ray showed multiple calcified oval shaped masses (arrow) in the pelvis

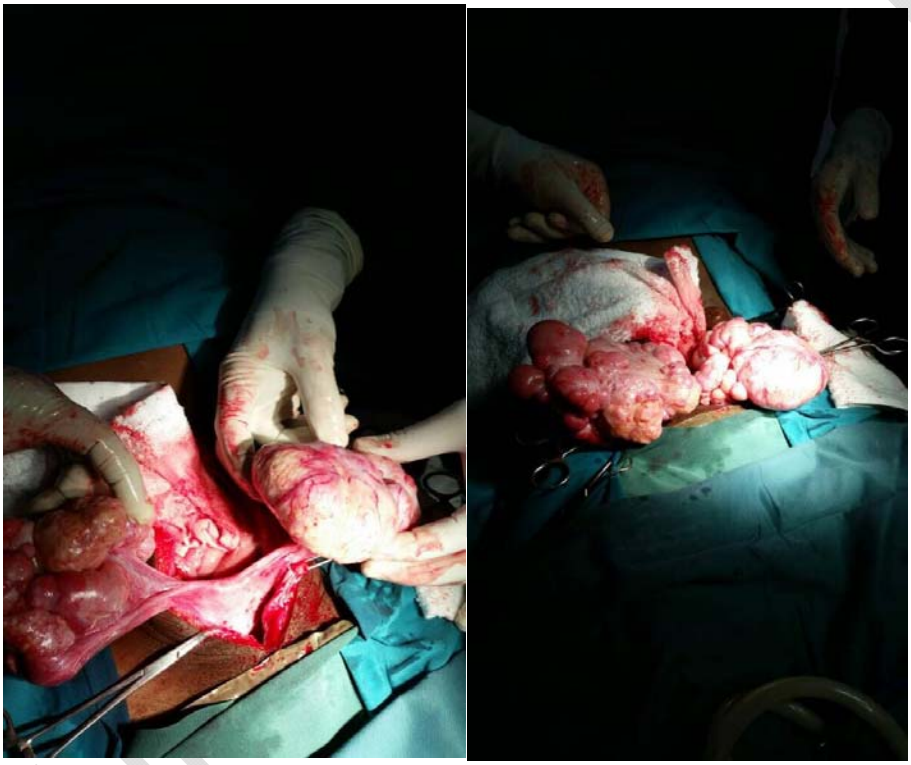

**Figure 2**: Exploratory laparotomy procedure: findings at exploratory laparotomy.

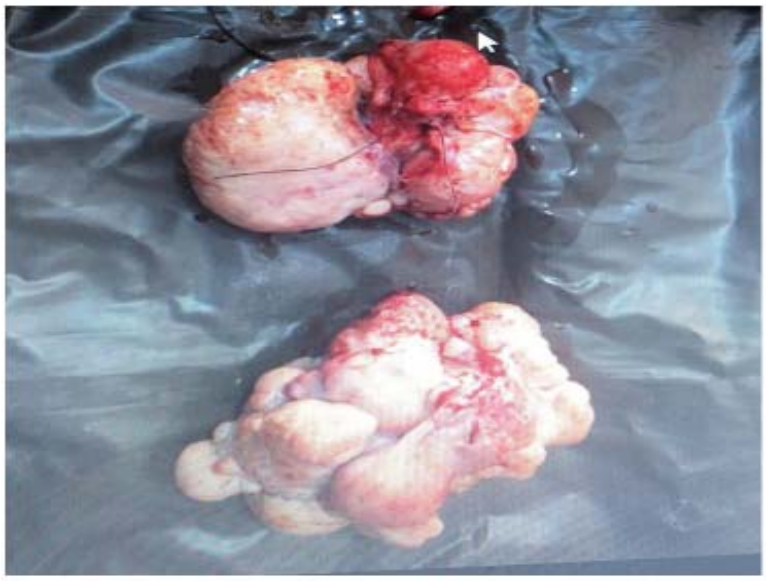

**Figure 3**: Right and left ovaries with gross multiple tumor nodules that were firm to hard in consistency

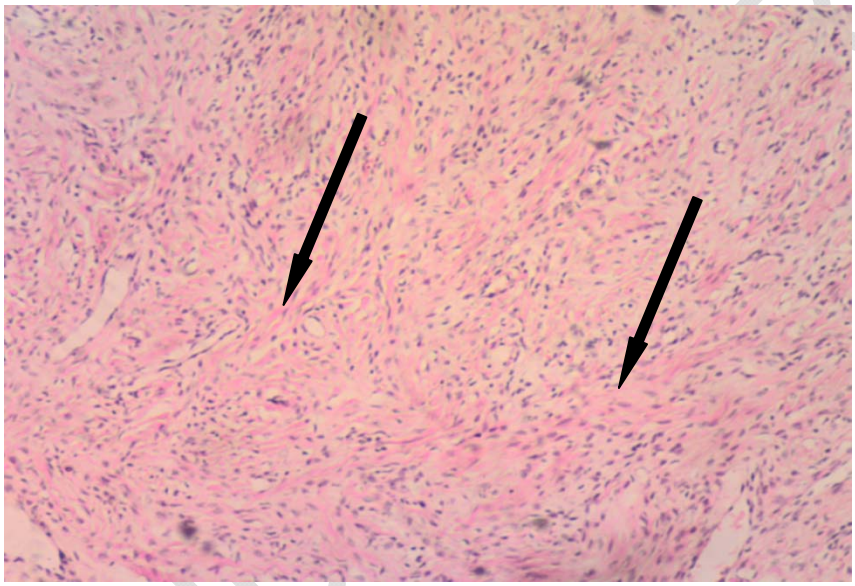

**Figure 4:** Photomicrograph from the right ovarian mass showed proliferating spindle cells disposed in a whorled arrangement with variable amount of extracellular collagen (arrows). (Haematoxylin and eosin, X100)

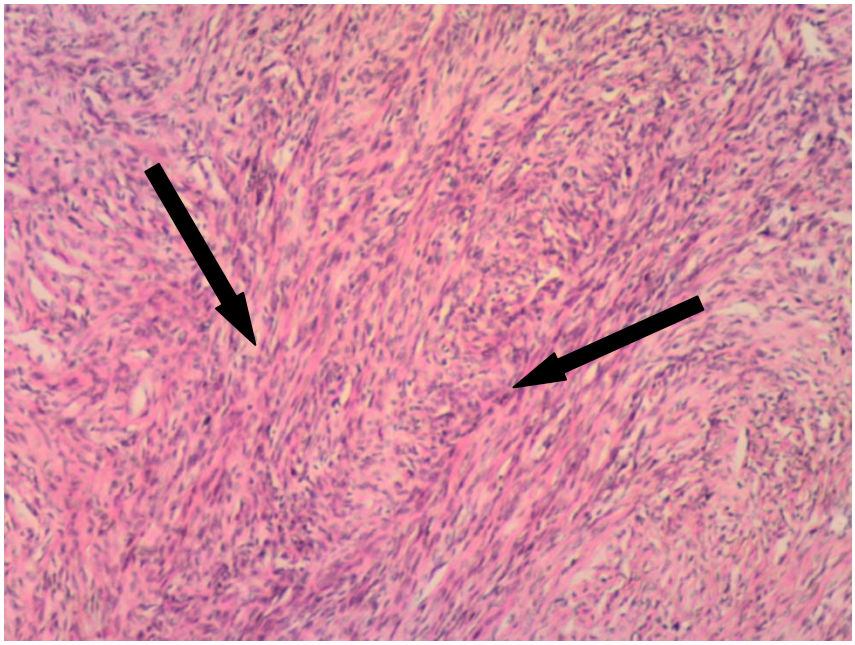

**Figure 5:** Photomicrograph from the left ovarian mass showed proliferating spindle cells disposed in a whorled arrangement (arrows). (Haematoxylin and eosin, X100)