Case Report

Angioleiomyoma of Uterus and Cervix: A Rare Report of Two Cases

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Abstract

We report two rare cases of genital angioleiomyomas (ALs), one each of uterus and cervix. The uterine AL showed a very rare presentation of endometrial polyp, while the cervical AL presented as an intramural cervical growth. We have also reviewed the literature and enlisted all uterine and cervical ALs reported till now.

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Introduction

Angioleiomyoma (AL) is a common tumor of extremities and the head-and-neck region but only rarely reported in uterus, cervix, ovary, and broad ligament. It is supposedly a morphological variant of leiomyoma, the most common tumor of the uterus. Uterine AL is quite a rare entity, and as per our search, only 38 case reports have been described in the literature so far [Table 1]. Out of them, only a very few have been described as a polypoid uterine angioleiomomatous lesion,[1,2] which makes our case (Case 1) rarer.

Another case we report is a cervical AL, of which we could find only three case reports reported in the literature so far [Table 2].[1,3,4] None of them showed a lipomatous component in contrast to our case (Case 2) where it is present, making it the first cervical AL of its kind.

Case Reports

Case 1

A multiparous patient of known hyperthyroidism presented with discharge per vaginum, heavy vaginal bleeding during periods, and dysmenorrhea for 2 years. Per speculum examination revealed an unhealthy cervix with the presence of discharge and bleeding. Furthermore, a polyp at external os measuring 3 cm × 3 cm was noticed. The rest of the clinical examination was within the normal limits. Her Pap examination showed endocervical squamous metaplasia. Her routine hematological, biochemical, urine, and X-ray chest findings were unremarkable. Venereal Disease Research Laboratory test was negative. Human immunodeficiency virus and Hepatitis C virus antibodies were nonreactive. Ultrasound examination of the abdomen showed an endometrial polyp arising from the anterior uterine wall with increased vascularity and loss of posterior wall differentiation. Bilateral adenexae were normal. Endometrial biopsy done 2 months ago showed proliferative endometrium. The patient had a history of pulmonary tuberculosis 22 years ago which was treated satisfactorily by antituberculous treatment. Hysteroscopic polypectomy was done and polyp sent for the histopathological examination.

Case 2

A 47-year-old known diabetic woman presented with heavy cyclical vaginal bleeding for 3 months. Her abdominal examination was unremarkable. Per speculum examination revealed ectropion of the lower lip of cervix with erosion. Ultrasound examination of the abdomen showed a nabothian cyst on the anterior lip of cervix. A round irregular marginated echogenic mass (? polyp) measuring 22 mm × 20 mm was seen on cervix. Hematological, biochemical, and urine examinations

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Table 1: Cases of uterine angioleiomyomas reported until now

Year	Author	Number of cases	Size of AL (cm)	Age of patient (years)	Presentation	Management
2018	Pierro A *et al*.	1	32	37	Abdominal distension, menorrhagia, and dyspepsia	TH
	Kim H *et al*.	1	5	36	Lower abdominal pain	Laparoscopic myomectomy
	Gupta M *et al*.	4	3–15	32,37,44,46	Abnormal uterine bleeding	Polypectomy in one, myomectomy in two and TH with BSO in one case
2017	Hong JA *et al*.	1	12.5	33	Menorrhagia, anemia, and a palpable lower abdominal mass with headache, vertigo, and vomiting in an unmarried woman	Myomectomy
	Singh S *et al*.	1	5.7	40	Menorrhagia, dysmenorrhea, and abdominopelvic mass in a nullipara	TAH with BSO
	Junainah EM *et al*.	1	14	42	Severe dysmenorrhea and cyclical lower abdominal pain	Myomectomy
2016	McAdams CR *et al*.	1	2.5	51	Menorrhagia	Hysteroscopic resection
	Sikora-Szczęśniak DL	9	13 (largest)	43–60	Abnormal uterine bleeding and anemia	TAH +/- resection of appendages in six, removal of AL in two and myomectomy in one case
	Demiray H *et al*.	-	-	-	Details not found	-
2015	Diwaker P *et al*.	1	4.5	39	Polymenorrhagia and pain in the lower abdomen	TAH
	Zizi-Sermpetzoglou A *et al*.	1	6.8	50	Lower abdominal pain and abnormal uterine bleeding	TAH with BSO
	Gomez FJT *et al*.	1	6	46	Right corneal pedunculated AL in a known case of leiomyomatosis	TAH with BSO
	Laxminarayana B *et al*.	1	9	37	Dysfunctional uterine bleeding, urinary retention, and pain abdomen	TAH
2014	Sharma C *et al*.	1	30	19	Abdominal mass, episodic abdominal pain and menorrhagia	Subtotal hysterectomy
	Grigoriadis C *et al*.	1	25	53	Severe abnormal uterine bleeding with palpable pelvic mass	TAH with BSO, total omentectomy, and pelvic lymph node dissection
	Kamath MS *et al*.	1	18	37	Menorrhagia, abdominopelvic mass with easy bruising	Myomectomy
	Bommanahalli BP *et al*.	1	6	45	Lower abdominal pain and menorrhagia	TAH with BSO
2013	Jin CH *et al*.	1	5	41	Abnormal uterine bleeding in a nullipara	Laparoscopic myomectomy
	Manimekhala P *et al*.	2	9, 2	28, 45	Oligomenorrhoea followed by secondary amenorrhea; excessive uterine bleeding and leucorrhea	TAH; Vaginal hysterectomy
	Sikora-Szczęśniak DL *et al*.	1	6	51	Painful cycles with profuse bleeding	TAH with BSO
2012	Sahu L *et al*.	1	25	49	Menorrhagaia	TAH with BSO
	Thomas S *et al*.	1	18	47	Abdominal distension, menorrhagia, dyspepsia, weight loss, pseudo Meigs syndrome with raised CA-125 levels	TAH with BSO
	Handler M *et al*.	1	26	38	Progressive abdominal distension and early satiety, along with acute abdominal cramps, menorrhagia and deranged coagulation profile	TAH with BSO
2011	Lazarov N *et al*.	1	-	35	AL in a diabetic patient with chronic renal insufficiency on hemodialysis	Details not found
2009	Hakverdi, S *et al*.	Multiple	-	69	Progressive abdominal distension	TAH with BSO

Contd...

Table 1: Contd...

Year	Author	Number of cases	Size of AL (cm)	Age of patient (years)	Presentation	Management
2008	Koshy AK *et al*.	1	30	45	Abdominal lump	TAH with BSO
	Byun JH *et al*.	1	--	--	Massive vaginal bleeding and severe anemia	Details not found
2007	McCluggage WG *et al*.	3	1.5–6	33, 44, 49	Abnormal uterine bleeding	TAH with BSO in first two cases, myomectomy in the third
2006	Culhaci N *et al*.	1	6.5	45	Lower abdominal pain, nausea, vomiting	TAH with BSO
2003	Hsieh CH *et al*.	3	14, 20, 22	50	Menorrhagia, lower abdominal mass	TAH with BSO
2002	Prabha V *et al*.	-	-	-	Details not found	-
2001	Agorastos T *et al*.	1	11.8	50	Menorrhagia, lower abdominal discomfort	TAH with BSO
1999	Hennig Y *et al*.	1	12	41	Not mentioned	Hysterectomy
1995	Gyure KA *et al*.	1	Not mentioned	29	Left lower extremity phlebitis, deep vein thrombosis, pulmonary embolism in a nulligravida patient of tuberous sclerosis associated with uterine lymphoangiomyomatosis (with sarcomatous transformation), papillary serous carcinoma ovary, uterine angiomyoma and renal angiomyolipoma	TAH with BSO with tumor debulking, pelvic lymph node dissection and resection of sigmoid colon with colorectal anastomosis
1990	Jameson C *et al*.	1	4	32	Acute intermittent right abdominal pain in a patient of tuberous sclerosis, ovarian endometriosis and infertility	Resection of angioleiomyomatous mass with right oopherectomy
1980	Konichezky M *et al*.	Multiple	7 (largest)	33	Urinary frequency and urgency with increased uterine size on examination	TAH with BSO
1964	Salasc P *et al*.	-	-	-	Details not found	-
1953	Catsaras J *et al*.	-	-	-	Details not found	-

TH: Total hysterectomy, BSO: Bilateral salpingo-oopherectomy, TAH: Total abdominal hysterectomy, AL: Angioleiomyoma

were unremarkable. A clinical diagnosis of dysfunctional uterine bleeding with cervical polyp was made. Total abdominal hysterectomy was done and specimen sent for the histopathological examination.

Clinical history and findings of cases are summarized in Table 3.

Gross specimen of Case 1 showed a polyp measuring 6 cm × 3 cm × 2 cm. Its cut surface was grayish brown and solid. Gross specimen of Case 2 showed uterus and cervix measuring 10 cm × 3 cm × 2 cm. Cut section of the uterus was unremarkable; endometrium measured 1.2 cm. Cervix was elongated measuring 3 cm length. Cut surface of cervix showed a well-defined grayish-white round growth on the lower end measuring 3.2 cm in diameter. Microscopic examinations of polyp (Case 1) and lower cervical end growth (Case 2) showed almost similar picture, comprising spindle-shaped cells, arranged in fascicles or evenly distributed in compact form. Cells had spindle-shaped nucleus with no atypia or mitosis. In between were areas having thick-walled arterioles, sheathed by spindle cell layers and showing areas of vessel wall hyalinization. Hemorrhagic areas as well as few fibrin clots were also seen in and around vessels. Occasional fat cells with areas of hyalinization were noticed in the smooth muscle component areas in the cervical lesion [Figure 1a-f]. Microscopic examinations of the uterus and cervix of Case 2 were unremarkable.

Immunohistochemically, both the cases were diffusely positive for smooth muscle antigen (SMA) in fascicles encircling blood vessels as well as in the smooth muscle component in the background and CD 34 positivity in endothelial lining of blood vessels. Estrogen receptor staining was weekly positive to negative while progesterone receptor staining was negative. CD 10, S-100, Sudan Black, and HMB-45 were negative in all cases [Figure 2a-f].

Discussion

ALs are benign tumors which are considered a distinct variant of leiomyomas. They are predominantly

Table 2: Cases of cervical angioleiomyomas reported until now

Year	Author	Number of cases	Size of AL (cm)	Age of patient (years)	Presentation	Management
2018	Gupta M *et al*.	2	5–10.5	36–39	Lower pelvic pain, bleeding per vaginum	Polypectomy; myomectomy
2011	Al Sannaa GA *et al*.	1	6.5	29	Presenting as a cervical polyp	Excision of polyp
2009	Koleskas D *et al*.	1	3	48	Menorrhagia, intermenstrual bleeding, and postcoital bleeding	Polypectomy

AL: Angioleiomyoma

Table 3: Clinico-pathological summary of cases

Cases	Age (years)	Clinical presentation	Preoperative diagnosis	Histopathological sample sent	Gross findings	Histopathological diagnosis
Case 1	32	Menorrhagia with lower abdominal pain. Ultrasound showed increased vascularity in the polyp	Leiomyomatous polyp	Polypectomy tissue	Measured 6×3 × 2 cm Cut surface grayish brown and solid	Angioleiomyomatous polyp of the uterus (venous type)
Case 2	47	Menorrhagia	DUB with? cervical polyp	Uterus and cervix	Cervix lengthened, measuring 3.5×2 × 2 cm. Cut surface showed a solid grayish-white growth on anterior lip measuring 3.2 cm	Angioleiomyoma of cervix (venous type with lipomatous component)

DUB: Dysfunctional uterine bleeding

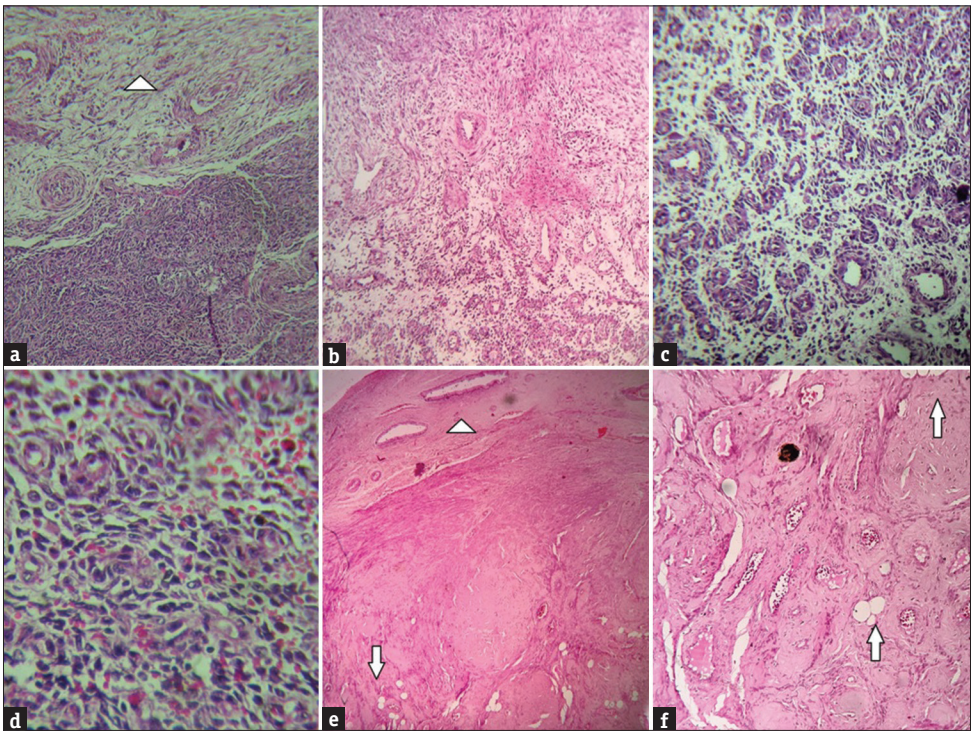

Figure 1: Uterine angioleiomyoma with perivascular swirling of smooth muscles and adjacent normal uterine tissue (arrow head) (H and E, ×50, ×100, ×100, ×400, a-d); cervical angioleiomyoma with occasional clusters of fat cells (arrows) and adjacent normal cervical tissue (arrow head) (H and E, ×50, e-f)

present in subcutis of lower extremities and rarely in the head and neck region and trunk.[5] Very rarely, they occur in the female reproductive tract, mostly in middle aged women and usually present with abnormal uterine bleeding and pain in the abdomen. They may also present with atypical symptomatology such as acute abdomen, massive bleeding per vaginum, Pseudo-Meigs Syndrome,[6] puberty menorrhagia, coagulopathies,[7] or catastrophic events such as rupture of the uterus.[8]

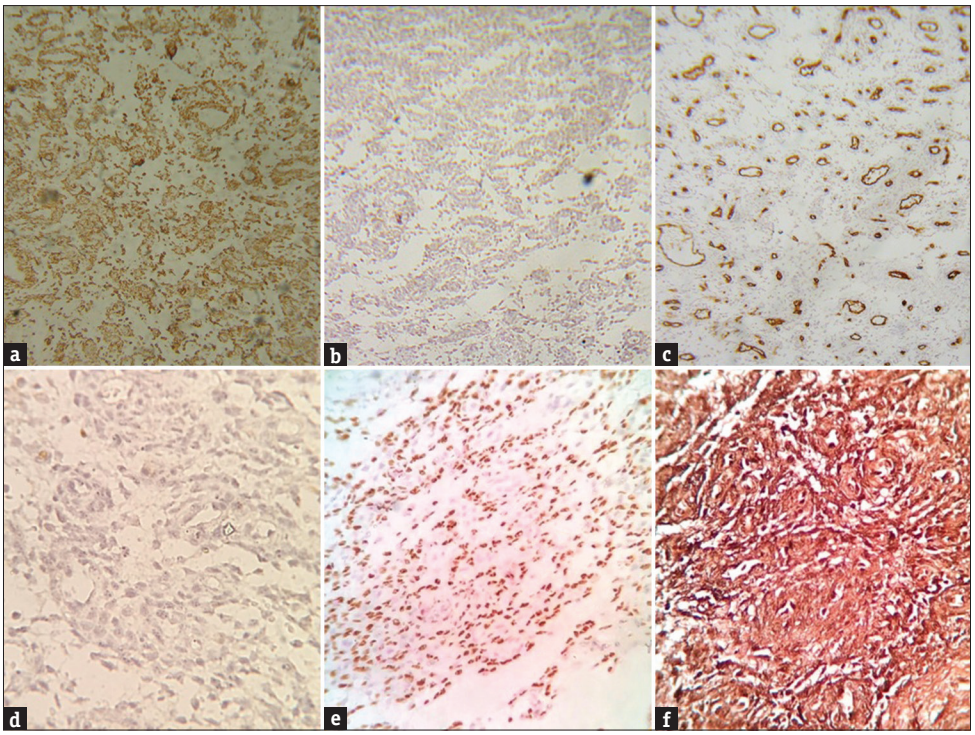

Figure 2: Immunohistochemistry of uterine angioleiomyoma showing strong smooth muscle antigen positivity in perivascular smooth muscle fascicles (×100, a); negative reaction with CD 10 (×100, b); positive CD 34 staining in vascular endothelium (×100, c); negative Estrogen receptor staining (×100, d); focally positive progesterone receptor staining (×100, e); negative staining with reticulin stain (×100, f)

Pathogenesis of AL is unclear. Few authors have found karyotypic abnormalities in such patients.[9] Histogenetically, most authors believe these tumors to be hamartomatous, akin to renal angiomyolipomas. This view is substantiated by the presence of mature fat cells in some of the cases, as in our case (Case 2).[5] Abnormal uterine bleeding often results from dysregulated growth factors and their receptors, which affect vascular morphology and regulate angiogenesis,[10] while pain may be caused by ischemia or vascular contraction.[8]

Microscopically, three histological types are distinguished in uterine or cervical AL, capillary or solid, cavernous, and venous. Tumors of the solid type are composed of abundant small-sized vascular channels in the background of compact and intersecting smooth muscle bundles which also sheath these channels. Tumors of the cavernous type have dilated vascular channels with smaller amounts of smooth muscle tissue in their wall, merging imperceptibly with the surrounding smooth muscle bundles. Tumors of the venous type consist of vascular channels with thick muscular walls of venous type with not so compact smooth muscle background. Hyaline and myxoid degeneration may be present, as seen in our cases also. Both of our cases are of venous variant.

The differential diagnosis of uterine AL depends upon the histopathological picture. In our case 1, it was confused with endometrial stromal tumor (EST) because of strikingly similar arrangement of blood vessels. However, on closer scrutiny, other features of EST such as resemblance with endometrial stroma, hyalinization, infiltrative margins, and individual cell encircling by reticulin-positive cells were missing. CD-10 negativity along with diffuse SMA positivity confirmed the diagnosis of AL, as CD-10 positivity is usually strong in EST but negative or weak in AL, as in our case. Another differential diagnosis is perivascular epithelioid cell tumor (PEcoma), distinguished by its clear/eosinophilic cytoplasm and HMB 45 positivity, which is negative in AL. Vascular leiomyoma can be reasonably distinguished by its myometrium-like capillaries and few arterioles unlike thick sheathed blood vessels of AL. Moreover, sheer number and density of AL are very different from that of vascular leiomyomas. Fibrin deposition in the vessel walls is also an important feature of AL (as seen in our cases too) but unusual in vascular leiomyomas.[11]

Mitosis is sometimes seen in AL but is usually <2 per 10 HPF. In cases with higher mitotic count, differential diagnosis of leiomyosarcoma should be considered if it coexists with necrosis, infiltration, and atypical nuclear features. Rarely, atypical ALs may also be present.[6] Hence, the importance of thorough sampling of the specimen cannot be overemphasized.

In our experience, endometrial or cervical polyps are sometimes replete with proliferating vascular channels,

with spindly compact stroma. Misdiagnosis of AL in such cases can be avoided by SMA staining which is positive in vascular walls but negative in stroma, unlike AL where both tissues are strongly positive.

AL of uterus can rarely be diagnosed in patients without histopathology although infrequently, an alert ultrasonologist can suspect it by increased vascularity in a lesion showing echogenicity different from typical leiomyomas,[2] as in our Case 1. Sometimes, contrast-enhanced computed tomography may also indicate its possibility by revealing the presence of multivascular branches within the tumor mass and uterine artery hypertrophy.

Treatment is hysterectomy. Procedures such as myomectomy are known to have increased rate of incomplete removal and recurrence[5] or even intra-operative failure of surgeon to reach plane of cleavage and uncontrolled hemorrhage.[12] However, sometimes angiomyomectomy with free margins may be attempted in unmarried of childless women.

At present, the WHO does not recognize AL of uterus or cervix as a distinct tumor. In this context, we agree with McCluggage and Boyde[11] who proposed that the WHO should include AL among benign variants of uterine leiomyomas.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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