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The Signet Ring Stromal Tumor in a 13-Year-Old Girl: Case Report

Tajossadat Allameh[1] and Behnoosh Mohamadi[2*]

1. *Department of Obstetrics and Gynecology, Isfahan University of Medical Sciences, Isfahan, Iran*
2. *Departments of Pathology, Isfahan University o Medical Sciences, Isfahan, Iran*

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Corresponding Information:

Behnoosh Mohamadi

Departments of Pathology, Isfahan University o Medical Sciences, Isfahan, Iran

Email: behnoosh.mohamadi.mui@gmail.com

ABSTRACT

Signet ring stromal tumor is a rare benign ovarian neoplasm, of which only about 17 cases have been reported since 1996. The signet ring appearance of this tumor may mimic a Krukenberg tumor and result in a diagnostic challenge in some cases. The previous cases occurred in adult or in old patients. We have reporteda Signet ring stromal tumor in a 13-year-old girl.

Keywords Signet ring stromal tumor, Ovary, Neoplasm, Sex cord stromal tumor

Introduction

Signet ring stromal tumor is a rare benign ovarian neoplasm which was reported by Ramzy in 1996 for the first time **(1)**. Only about 17 cases have been reported since 1996, listed in Table 1 **(1-14)**. The signet ring appearance of this tumor may mimic a Krukenberg tumor and result in a diagnostic challenge in some cases, specially if the neoplasm involves both ovaries **(13)**.

Table 1. Clinical features of signet ring stromal tumor from review of literature

Author	Presentation	Laterality	Macroscopy	Size	Procedure	Outcome
Ramzy, 1976 **(1)**	28Yrs, Abdominal pain	Right-sided	Solid	8 cm	Total abdominal hysterectomy and right salpingo-oophorectomy, wedge resection of left ovary	No recurrence until 15 months after surgery
Suarez *et al.*, 1992 **(2)**	50Yrs, Menopause3 years ago , Abdominal pain, Palpable mass at left hemiabdomen		Solid white, with irregular yellowish areas, firm fasciculated in appearance	9 cm	Simple left oophorectomy	No recurrence until 26 months after surgery
Dickersin *et al.*,1995 **(3)**	1.21Yrs,NA ۲/۷۸Yrs,NA ۳/۸۳Yrs,NA		Solid Hemorrhagic Solid cystic Solid	5 cm 8.5 cm 8.9 cm	NA NA NA	No recurrence for 2 years after surgery

						No recurrence for 1.6 years after surgery No recurrence for 2 months after surgery
Cashel *et al.*, 1999 **(4)**	52Yrs, weight loss, Left sided adnexal mass, No hormonal effect	Conjunction with Brenner tumor	A hard well-defined nodule with a firm mottled yellow to tan surface	2.5 cm	Total abdominal hysterectomy and bilateral salpingo-oophorectomy	No evidence of recurrence in 2 years follow up
Su *et al.*, 2003 **(5)**	76Yrs, Low abdominal pain, Left palpable adnexal mass	Left-sided	Grayto pale-tan solid tumor with Focal stromal edema and scattered chalky-white spots	5 cm	Bilateral salpingo-oophorectomy	No recurrence for 12 months after surgery
Vang *et al.*, 2004 **(6)**	1.34 Yrs,pelvic mass 2.35 Yrs,pelvic mass 3.41 Yrs,Abdominal pain	Left-sided Right sided Right sided	Gray, tan, or yellow. One tumor had a nodular appearance and two were focally hemorrhagic and necrotic.	13 cm 3.5 cm 13 cm	1 and 3:Total abdominal hysterectomy and bilateral salpingo-oophorectomy 2:Right oophorectomy	No recurrence for 17.4 years after surgery No recurrence for 4.7 years after surgery No recurrence for 1month after surgery
Hardisson *et al.*, 2008 **(7)**	54Yrs, G2P2 3month history of low abdominal discomfort, Palpable mass at left hemiabdomen	Left sided	Solid	5 cm	Laparoscopic bilateral salpingo-oophorectomy	No recurrence for 21 months after surgery
Matsumoto *et al.*, 2008 **(8)**	76Yrs, Incidental pelvic mass in examination	Left-sided	Solid light brown focally admixed with white fibromatous tissue	7 cm	Total abdominal hysterectomy and bilateral salpingo-oophorectomy	No recurrence for 9 months after surgery
Forde *et al.*, 2010 **(9)**	69 y, G5 Hx of hysterectomy Abdominal pain of 1 week duration, No findings in Examination secondry to body habitus	Bilateral		5 cm	Exploratory laparatomy, partial omentectomy, bilateral salpingo-oophorectomy and peritoneal washing	-
Sukur *et al.*, 2010 **(10)**	44Yrs, G4P2 Polymenorrheafor 3 months, Palpable right adnexal mass	Right-sided	Semisolid	5 cm	Right salpingo-oophorectomy	-
Kopszynski *et al.*, 2016 **(12)**	79Yrs Abdominal uterine bleeding, Normal pelvic examination	Left sided	A poorly circumscribed, yellowish, firm, solid tumor	1.1 cm	Abdominal hysterectomy and bilateral salpingo-oophorectomy	No recurrence after 11 months
McGregor *et al.*, 2016 **(11)**	64 Yrs, left ovarian mass identified by imaging	Left sided	Yellow-tan and gray-white areas	3.5 cm	Total hysterectomy and bilateral salpingo-oophorectomy	-
2020 **(13)**	70 Yrs, Abdominal pain, distention, bloating and rectal bleeding	Bilateral tumor	Solid homogenous white-yellow, firm cut surfaces	Left:4 Right 5.5 cm	Total abdominal hysterectomy and bilateral salpingo-oophorectomy	No recurrence for 12 months after surgery
Tsai *et al.*, 2020 **(14)**	43y Palpable pelvic mass	Left-sided Collision tumor of sclerosing stromal tumor and signet-ring stromal tumor	Cystic and solid	16 cm	Left salpingo-oophorectomy	-

NA: not available

Case report

A 13-year-old virgin girl came to the office complaining about abdominal pain from several months ago. No abnormal findings were detected in past medical history and physical examination. Abdominal ultrasoundwas performed and revealed a hypo echogenic heterogeneous lesion measuring 33×29 mm in the medial portion of the left ovary. Color Doppler ultrasoundshowed low vascularity index of lesion. Due to lack of typical appearance of the mass, the radiologist offered more evaluation by MRI. The MRI examination revealed a lobulated complex solid cystic mass in the left ovary measuring 51×44×26 mm. The solid component showed low T1w and T2w signal intensity with avid enhancement on post contrast images with nor hemorrhagic neither fatty signals, and Sertoli Leydig cell tumor was suggested as the first diagnosis. Ascites or lymphadenopathy was not found in imaging studies. Serum LDH, CEA, CA19-9, CA125, Beta HCG, and AFP level were within normal range. The patient underwent left salpingo-oophorectomy. Specimenwas received fresh for frozen section in pathology laboratory. In Macroscopic examination, the left ovary was measured as 6×5×4 cm. Cutting revealed a solid, homogenous, firm, white-tan mass measured as 5cm in maximum diameter without hemorrhage, necrosis or calcification. Microscopically, the neoplasm composed of fibroma-like areas that merged with signet ring component (About 50% of tumor volume). In the latter component, large sheets of round cells with single or multiple intracytoplasmic clear vacuoles and eccentrically located small nuclei without obvious atypia, mitotic activity or necrosis were present. In immunohistochemical staining, the cells were diffusely positive for vimentin and negative for CK, CEA, and CD10. Only focal expression of inhibin-A and calretinin were seen. The diagnosis of Signet ring stromal tumor was confirmed by immunohistochemical staining. Now after 2 years, the patient is well without recurrence or metastasis.

Discussion

Signet ring stromal tumor is a rare ovarian stromal neoplasm **(7)** Up to now only 17 cases have been reported (1-14). All the reported cases have occurred in adults with a range between 21 and 83 years of age (Mean age of 54 years) **(7)**. The clinical features are nonspecific,and abdominal pain is the most common symptom. On imaging,the mass may be solid or cystic ,nevertheless there is no specific features for distinguishing this tumor from other neoplasms.

Grossly, the tumor size varies from 1.1 to 16 cm **(12,14)**. Although bilaterally have been reported in two cases **(9,13)**, most of them are unilateral. This tumor mostly shows a solid cut surface, cystic and hemorrhagic changes also are noted **(3,6,13)**. All patients have no evidence of recurrence or metastasis after surgery (Duration of follow-up ranges from 1 month to 17.4 years). Cashel *et al.*, reported a signet cell stromal tumor in association with Brenner tumor in 1999 **(4)**. Tsai et al reported a collision tumor of Sclerosing stromal tumor and Signet ring stromal tumor in 2019 **(14)**. Microscopically tumor is composed of different proportion of spindle and round cells which some of them have similar morphology of signet ring cells. Marked cellular atypia, mitosis, and necrosis are absent. In some cases PAS positive intracytoplasmic hyaline globules are noted **(2)**. Special staining for mucin and lipid are negative. Ultrastructural studies have shown, the vacuoles are derived from diffuse cytoplasmic edema, mitochondrial swelling or cytoplasmic pseudoinclusions of extracellular matrix **(2,3,4,5)**. Histochemical and immunohistochemical findings have been shown in Table 2. In all studies tumoral cells are negative for EMA, CK, and CEA. Immunoreactivity for calretinin, inhibin, ER, cyclin D1 and beta-catenin has contradictory.

The most important differential diagnosis of signet ring stromal tumor is Krukenberg tumor. Most of the patients with Krukenberg tumor have no previous history of carcinoma. Though bilaterality has been reported in both tumors, therefore it is not useful for differentiation of these two tumors. Grossly the gelatinous appearance favors krukenberg tumor but, multinodularity, necrosis, and hemorrhage may be seen in both neoplasms. Krukenberg tumor are constantly positive for PAS-D and mucicarmin. On the other hand,all signet ring stromal tumors are negative. Pancytokeratin and Vimentin are so helpful for distinguishing of challenging cases, so that keratin is not expressed by signet ring stromal tumors whereas Vimentin is strongly expressed **(6)**.

Conclusion

Signet ring stromal tumor is a rare stromal neoplasm. Previous reported cases aged more than 21 years old. we report a rare case in a 13-year-oldgirl. This benign neoplasm may be misdiagnosed as krukenberg tumor with aggressive behavior, but Histochemical and immunohistochemical techniques are helpful for differentiation between these tumors.

Acknowledgment

None.

Conflict of Interest

None.

	chemical reaction	Reticulin stain	Keratin	EMA	CEA	Vimentin	SMA	Calretinin	Inhibin	ER	CyclinD1	B catenin
Suarez A, et al 1992	PAS:Neg. Alcian blue:Neg. SudanIII:Neg.		Neg.		Neg.	Neg.	Neg.					
Cashel AW, et al 1999	Pas:Neg. Mucicarmin:Neg.	Fiber surrounding individual signet ring cells	Neg.	Neg.	Neg.	diffusely pos.	diffusely pos for actin in both cells		Positive in most signet ring and half of the remaining stromal cells			
Su RM, et al 2003	PAS,Mucicarmin SudanIII:Neg.	Reticulin fibers enmeshing individual spindle cell were highlighted	Neg.									
Vang R, et al 2004	PAS:Neg.		Neg.			Diffusely pos.		Neg.	Neg.			
Hardisson D, et al 2008	PAS:Neg. Oil-red-O:Neg.	a delicate network of fibers investing the signet-ring cells as well as the surrounding fibrous stromal cells (	Neg.	Neg.	Neg.	diffusely pos.	diffusely pos.	Focal	Focal			
Matsumoto M,et al 2008	PAS , Alcian blue, mucicarmine, and oil-red O stain:Neg.	reticulin fiber surrounding individual signet-ring cells	Neg.			Pos.	Focal pos.		Focal pos.	Neg.		
Forde GK, et al 2010	Pas:Neg Mucicarmin:Neg		Neg.	Neg.				Focal pos.	Focal and weak pos.			
Sukur YE,et al 2010	PAS ,Mucin staining:Neg		Neg.			intensely pos.	Focal pos	Neg.	Focal pos.			
McGregor SM, et al 2016	PAS:Neg		Neg.			Pos.			Neg			Diffuse nuclear
Kopszynski J, et al 2016	PAS, alcian blue, mucine, and oil-red O:Neg		Neg.	Neg.	Neg.	Pos.	Neg.	Neg.	Neg.	Neg.	nuclear positivity	Strong nuclear and cytoplasmic
Chen PH, et al 2019		Regular reticulin fiber network surrounding individual cells	focally pos.	Neg.			diffusely pos	diffusely pos		focally posi	Neg	Cytoplasmic and membranous staining pattern without any nuclearexpression
Tsai TY,et al 2019			Neg.			Pos.		Neg. in signet cell				
Current case			Neg.		Neg.	Strongly pos.		Focally pos.	Focally pos.			

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