

Sclerosing Stromal Tumour of Ovary: A Rare Case Report

Dr Neha Singh¹, Dr Jitender Singh Chauhan²

¹Senior Resident, Department of Pathology, BPSGMC, Khanpur Kalan, Sonipat Road, Haryana, India

²Resident, Department of Surgery, PGIMS, Rohtak, Haryana, India

ABSTRACT

Sclerosing stromal tumours (SSTs) are rare, benign, sex cord stromal tumours of the ovary. They affect women, in their second and third decades of life, who complain mainly of menstrual irregularity. We reported a histologically confirmed case of 18 year old patient who presented with chief complaints of pelvic pain and irregular menstruation. She later underwent surgery when a pre-pathology work-up raised the suspicion of a malignancy. It is imperative to consider the differential diagnosis of SST of the ovary in a young woman with an ovarian tumour.

KEYWORDS: Ovary, Sclerosing stromal tumor (SST), Smooth Muscle Actin (SMA).

INTRODUCTION

Sclerosing stromal tumor (SST) is an extremely rare benign subtype of ovarian stromal neoplasm of the sex cord stromal category. It has distinctive clinical and pathological features which differentiate it from other stromal tumors. The tumor occurs predominantly in the 2nd and 3rd decades of life.^{1, 2} It is usually unilateral and well circumscribed; and its recurrence has not been reported.³ Chalvardjian and Scully first described these tumours as having a heterogeneous pattern, often characterised by pseudo-lobulation of cellular areas, a prominent tendency to sclerosis and marked vascularity, which distinguished them from fibromas, thecomas and other more common types of ovarian stromal tumours¹.

CASE REPORT

A 18 year old girl was admitted to the hospital for menstrual irregularity, metrorrhagia and pelvic pain since last four months. Physical examination revealed no palpable abdominal lump nor hepatosplenomegaly. On ultrasonographic examination, there were solid and cystic areas in left ovary of approximately 8X6X4 cm in size. All tumor markers (LDH, AFP, Beta HCG, CA-125, CEA) were below cut-off levels. Serum hormone levels were within normal limit. Ascites was not present. Medical history of the patient was unremarkable. On CT scan, there was bulky left ovary measuring 5X4.4X3.2 cm with multiple cystic areas, largest measuring 3.2X2.4 cm, suggestive of neoplastic etiology. MRI showed minimal ascites in para-colic spaces & pelvis, a left ovarian solid-cystic mass measuring 5.2 cm, suggestive of neoplastic etiology.

The right ovary was normal. The ovarian mass showed benign pathology on frozen section analysis which was subsequently removed by laparoscopic salpingo-oophorectomy. Post-operative recovery was uneventful. The specimen measured 6X5X3 cm with sharply demarcated mass having smooth and intact outer surface. The cut surface revealed solid, cystic and edematous areas. No haemorrhagic or necrotic areas were observed. The specimen was fixed in 10% neutral formalin. The paraffin-embedded tissue sections were stained with haematoxylin and eosin. Immunohistochemical analysis for inhibin, vimentin, smooth muscle actin, desmin, cytokeratin, Ki-67 was performed by using avidine-biotin peroxidase complex method.

On histopathological examination, the tumor showed ovoid to spindle cells, luteinized cells arranged in lobules separated by dense to moderate fibrocollagenous stroma. Intercellular edema was seen with intervening thin walled large blood vessels. Mild nuclear atypia was also seen (Fig. 1). The lobules were composed of two-cell population: rounded polyhedral cells with eosinophilic or vacuolated cytoplasm and spindle shaped fibroblasts (Fig. 2). Mitotic figures were absent. Cellular areas revealed a rich thin-walled vascular network. Immunohistochemical analysis demonstrated positivity for

SMA (Fig. 4) and negativity for vimentin, cytokeratin. Ki-67 index was 1 to 2%. At the periphery of the mass, residual ovarian tissue was seen. Subsequently the diagnosis of sclerosing stromal tumor of the ovary was made. Attached fallopian tube was unremarkable.

DISCUSSION

Sclerosing stromal tumor is a rare, benign subtype of ovarian stromal tumors that differs from the other entities both clinically and pathologically. As a distinct entity, it was first described in 1973 by Chaldvardjian and Scully.¹ The most common presenting clinical symptoms include menstrual irregularity, pelvic pain and non-specific symptoms related to the ovarian mass.^{4,5} Few patients also present with infertility, hirsutism or no symptoms at all which resolved spontaneously once the tumor was removed.⁶

SST usually presents in the 2nd-3rd decade of life, whereas other ovarian stromal tumors present in the 5th-6th decade of life.⁵ Generally, the tumour is unilateral. The common naked-eye appearance is mostly solid and cystic in various proportions.⁷ Microscopic picture of SST is heterogenous and contrasts with the relative homogeneity of other stromal tumors like thecoma and fibroma. The tumor is characterized by cellular pseudo-lobules, prominent interlobular fibrosis, frequent marked vascularity and a dual-cell population, collagen-producing spindle cells and lipid containing round or ovoid cells.⁸ The peculiar finding of a thick rim of compressed residual ovarian tissue at the periphery of the mass suggests a slow growing benign tumor. Thecoma and fibroma have no residual ovarian tissue at the periphery as they generally occur in the fifth or sixth decades of life when the ovaries are atrophic.^{9,10}

Inhibin positivity differentiates SST from vascular tumors. Massive ovarian edema may be confused with SST, but preserved ovarian tissue within the edematous stroma and absence of heterogeneity favors the diagnosis of massive ovarian edema.⁹ The edema of SST is zonal in contrast to that seen in massive ovarian edema or an edematous fibroma. Sometimes, the vacuolated cells and signet-ring cells in association with edematous stroma may be mistaken for Krukenberg tumor of the ovary. But these malignant tumors occur typically in women in the 6th and 7th decades, are mostly bilateral, and lack the pseudo-lobulated pattern of sclerosing stromal tumor on cut surfaces.¹ The vascular, sclerotic and edematous stromal changes are constant features of these tumours and relate to the local elaboration of some vascular permeability and growth factors (VPF/VEGF).¹¹

In the literature, calretinin, inhibin, CD34 and alpha glutathione S-transferase positivity (a-GST) was reported to be useful to differentiate SST from thecoma, fibroma and other sex cord stromal tumors.^{12, 13} CD 34 stains the endothelium of often dilated and branching vascular architecture, and clearly distinguishes SST from thecoma and fibromas. Alpha-GST positivity within scattered cells appears to be useful in the distinction of SST from diffuse staining thecomas and no staining fibromas.¹² On immunohistochemistry, SSTs are usually vimentin positive, SMA positive, inhibin positive or negative, calretinin positive or negative, desmin positive or negative, and pancytokeratin negative. MRI is useful in making a preoperative diagnosis of SST and distinguishing SST from other malignant ovarian tumors as well as other stromal tumors.¹⁴

CONCLUSION

The definite diagnosis of SST can be made only by pathologic evaluation but at least a diagnosis of benign ovarian tumor is possible intraoperatively via frozen section analysis by examining the background of pseudolobular pattern, heterogeneity of the cellular areas and densely hyalinized or markedly edematous stroma. Due to the rarity of this particular ovarian neoplasm, a possibility of sclerosing stromal tumor should be kept in mind in young patients with ovarian mass, as almost all the sclerosing stromal tumors of the ovary reported in the literature are benign and are treated successfully by enucleation or unilateral ovariectomy.

REFERENCES

- [1] A. Chalvardjian and R. E. Scully, "Sclerosing stromal tumors of the ovary," *Cancer*, vol. 31, no. 3, pp. 664–670, 1973.
- [2] G. Marelli, S. Carinelli, A. Mariani, L. Frigerio, and A. Ferrari, "Sclerosing stromal tumor of the ovary—report of eight cases and review of the literature," *European Journal of Obstetrics Gynecology and Reproductive Biology*, vol. 76, no. 1, pp. 85–89, 1998.
- [3] J. Prat, *Pathology of the Ovary*, Saunders, London, UK, 2004.
- [4] Matsubayashi R, Matsuo Y, Doi J, Kudo S, Matsuguchi K, Sugimori H. Sclerosing stromal tumor of the ovary: radiologic findings. *Eur Radiol* 1999; 9: 1335–38.
- [5] S.M. Ismail and S.M. Walker, Bilateral virilizing sclerosing stromal tumor of the ovary in a pregnant woman with Gorlin's syndrome: implications for pathogenesis of ovarian stromal neoplasms. *Histopathology* 1990; 17: 159-63.

- [6] Cashell AW, Cohen ML. Masculinizing sclerosing stromal tumor of the ovary during pregnancy. *Gynecol Oncol* 1991; 43:281–85.
- [7] Akbulut M, Colakoglu N, Soysal ME, Düzcen E. Sclerosing stromal tumor of the ovary: report of a case and review of the literature. *Aegean Pathology Journal*.2004;1:84-89.
- [8] Khanna M, Khanna A, Manjari M, Sclerosing Stromal Tumor of Ovary: A Case Report, *Case reports in pathology* 2012,1-3.
- [9] Ihara N, Togashi K, Todo G, Nakai A, Kojima N, Ishigaki T, Suginami N, Kinoshita M, Shintaku M. Sclerosing stromal tumor of the ovary: MRI. *J Comput Assist Tomogr* 1999;23: 555–57.
- [10] Kim JY, Jung KJ, Chung DS, Kim OD, Lee JH, Youn SK. Sclerosing stromal tumor of the ovary: MR-pathologic correlation in three cases. *Korean J Radiol* 2003; 4: 194 –99.
- [11] Kawauchi S, Tsuji T, Kaku T, Kamura T, Nakano H, Tsuneyoshi M. Sclerosing stromal tumor of the ovary: A clinicopathologic, immunohistochemical, ultrastructural, and cytogenetic analysis with special reference to its vasculature. *Am J Surg Pathol* 1998; 22: 83 –92.
- [12] Tiltman AJ, Haffajee Z. Sclerosing stromal tumors, thecomas, and fibromas of the ovary: an immunohistochemical profile. *Int J Gynecol Pathol* 1999; 18: 254–58.
- [13] Lam RM, Geittmann P. Sclerosing stromal tumor of the ovary. A light, electron microscopic and enzyme histochemical study. *Int J Gynecol Pathol* 1988; 7: 280–90.
- [14] Yerli H., Agildere A. M., Bilezikci B. and Karadeli E. Sclerosing stromal tumor of the ovary with torsion: MRI features, *Acta Radiologica* 2003;44: 612–15.

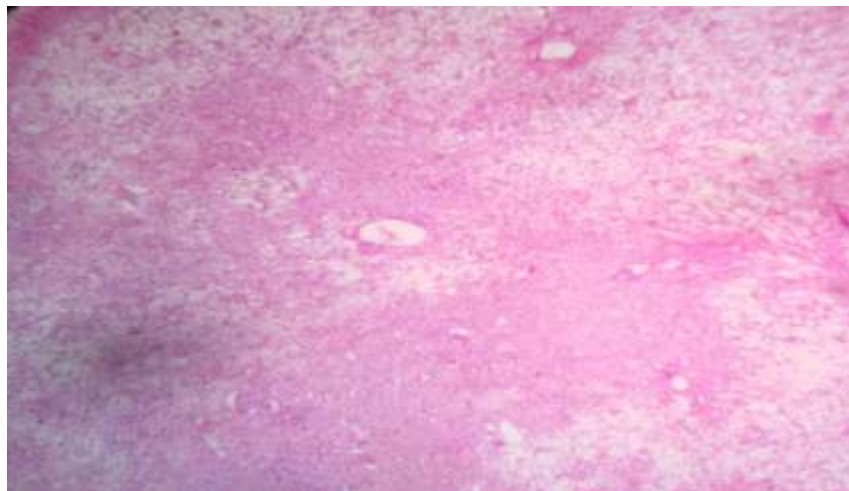


Figure 1: Hypercellular and hypocellular areas with prominent vascularity. (H&E X10)

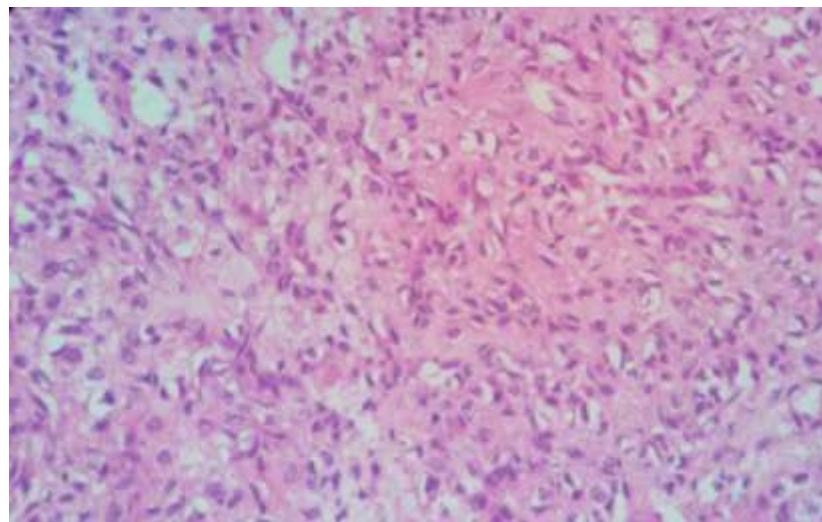


Figure 2: Two populations of tumor cells. One population is of spindle cells and the other population is of round cells with clear cytoplasm (H&E X40)

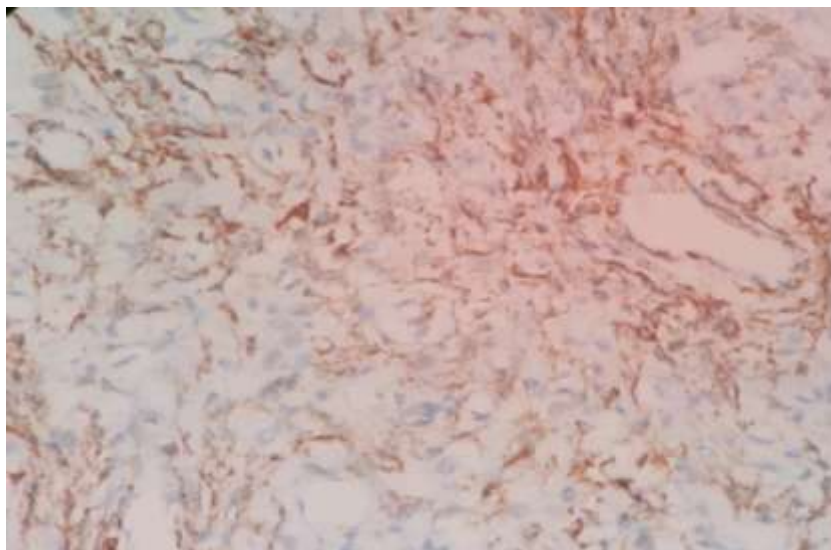


Figure 3: Tumor cells revealing diffuse immunoreactivity to smooth muscle actin (IHC for smooth muscle actin X40).