

# Ovarian Sclerosing Stromal Tumour presenting as acute Abdomen: A report of 2 cases

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## Citation

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## Abstract

Sclerosing stromal tumour (SST) is a rare benign ovarian neoplasm of stromal origin, distinct from the thecoma-fibroma group and steroid cell tumours clinically and pathologically. Sclerosing stromal tumour was first described in 1973 by Chalvardjian and Scully, it predominantly affects young women with a mean age of 28 years. We report two cases of unilateral sclerosing stromal tumour in two nulliparous women aged 29 years and 34 years. They presented with acute lower abdominal pain and right-sided abdominal swelling. Both had laparotomy which revealed right adnexal mass. Tissue diagnosis revealed a cellular tumour arranged in pseudolobular pattern composed of spindle-shaped fibroblastic cells and haemangiopericytoma-like areas.

## INTRODUCTION

Sclerosing stromal tumour (SST) is a rare benign neoplasm of ovarian stromal origin, distinct from the fibroma-thecoma group and steroid (lipid) cell tumours clinically and pathologically.<sup>1,2</sup> Unlike the thecoma-fibroma group of ovarian stromal tumours, which tend to occur in the fifth and sixth decades, the sclerosing stromal tumour predominantly affects young women with a mean age of 28 years.<sup>2</sup>

Since its initial characterization by Chalvardjian and Scully,<sup>3</sup> in 1973, there have been fewer than 100 case reports of this rare tumour. So far, all reports of sclerosing stromal tumour have involved only a single ovary except for two unusual cases of bilateral SST of the ovary in a pregnant woman with Gorlin's syndrome and an 11-year old premenarchal girl.<sup>4,5</sup> Most patients present with non-specific symptoms related to an adnexal mass. The tumour with rare exceptions is hormonally inactive.<sup>4</sup> If it is hormonally active, it is usually androgenic and most frequently occurs during pregnancy. Diagnosis of SST is often made by post-operative pathologic examination. The important differential diagnoses are other sex cord tumours including fibroma, thecoma and lipoid cell tumours.<sup>5,6</sup> We report two cases of unilateral sclerosing stromal tumour of the ovary.

## CASE REPORTS

### CASE 1

A 29-year old nulliparous woman presented with recurrent lower abdominal pain of eleven months duration. She had

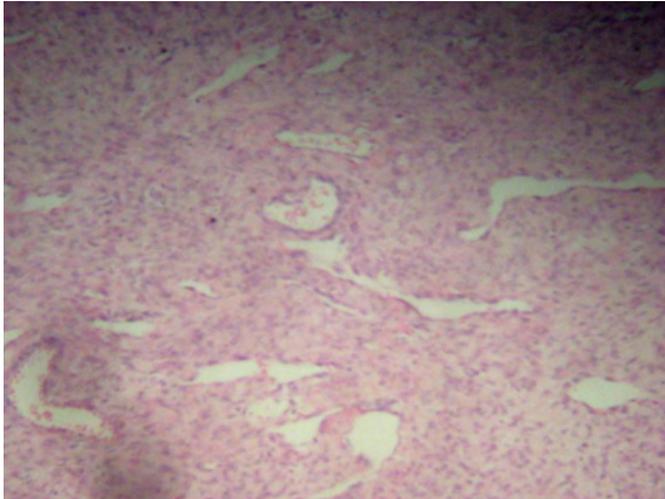
abdominal swelling three months before presentation.

On clinical examination, a 14-week size abdomino-pelvic was palpable. Ultrasonography showed a huge heterogeneous predominantly solid pelvic mass with some cystic foci. All laboratory tests including Full blood count, Urea & Electrolyte, Creatinine and urinalysis were normal.

The patient underwent Left salphingo-oophorectomy. Gross examination showed an encapsulated 10x9.5x7.5cm mass weighing 200g was received. Cut section showed solid grey and yellow areas with degenerated cystic foci. Histology showed a cellular tumour arranged in pseudo-lobular pattern composed of spindle-shaped fibroblastic cells and haemangiopericytoma-like vascular areas. (Fig 1)

**Figure 1**

Figure 1: Sclerosing stromal tumour with haemangiopericytoma-like area.



**CASE 2**

A 34-year-old nulliparous woman presented with 6 years history of inability to conceive and a year history of recurrent abdominal pain. On clinical examination, a 20 weeks size, non-tender, firm and mobile abdomino-pelvic mass was palpable. Ultrasonography revealed a lobulated right adnexa mass with some cystic foci. All laboratory investigations, Full blood count, Urea and Electrolytes, Creatinine and urinalysis were all within normal limits.

The patient underwent right salpingo-oophorectomy. Tissue measuring 12x10x8cm and weighing 120g attached to the omentum was sent for histology. Cut sections show solid grey and yellow areas with degenerated cystic foci. Histology revealed a cellular tumour arranged in pseudo-lobular pattern, composed of spindle-shaped fibroblastic cells and haemangiopericytoma-like vascular areas, like Fig 1.

**DISCUSSION**

Sclerosing stromal tumour (SST) of the ovary is a distinct subtype of sex cord-stromal tumours, which are derived from the ovarian stroma and, in turn, from sex cords of the embryonic gonad.<sup>4</sup> This tumour was first described by Chalardjian and Scully<sup>3</sup> in 1973, and occurs frequently in the second and third decades of life, with an average age of occurrence of 27.5 years. The ages of our patients in this report are 29 and 34 years respectively, these falls within the age of presentation of this tumour. Reports of SST, to date, have been benign, unilateral in nature and mostly manifesting in menstruating females.<sup>7</sup> However, two cases of

unusual bilateral SST of the ovary have been reported so far, one in a pregnant woman with Gorlin's syndrome who had undergone clomiphene therapy, and an 11-year old premenarchal girl.<sup>4,5</sup>

Most SSTs are hormonally inactive, Chalvardjian and Scully<sup>3</sup> originally considered ovarian SST to be a non-functioning tumour, but some investigators have described endocrine alterations caused by secretion of oestrogen, progesterone or testosterone as well as induction of precocious puberty.<sup>8,9,10</sup> Other symptoms that are usually present include pelvic pain, a palpable pelvic mass, metrorrhagia and menstrual irregularity including postmenopausal bleeding in elderly patients. Though, we could not demonstrate the functionality of this tumour by assaying for the hormones that is been secreted by the tumour, been in a resource limited environment. However, our patients had palpable pelvic masses, abdomino-pelvic pain, inability to conceive and irregular menstruation.

Both excised tumour in our patients were solid, well circumscribed with degenerated cystic areas containing straw-coloured fluid, this is compared to other reported ovarian SSTs.<sup>4,11</sup>

Histologically, SST of the ovary is distinguished from other ovarian stromal tumours by its pseudolobular pattern of cellular areas, with hypocellular, oedematous and collagenous areas. Other aspect of the tumour show prominent vasculature having haemangiopericytoma-like areas, sclerosis around clusters of individual cells and cellular heterogeneity of vacuolated lutenized theca-like cells and spindle-shaped fibroblast-like cells in the cellular areas.<sup>3,12</sup> Pseudo-lobulation and prominent vascularity are extremely rare in lutenized thecomas, fibromas and Krukenberg tumours.<sup>4,12</sup>

A comprehensive medical history, thorough physical examination, imaging studies, high index of suspicion on the part of the Pathologist, and correct tissue histology interpretation showing pseudolobular pattern of cellular with prominent haemangiopericytoma-like areas, and collagenized hypocellular areas; is significant in the diagnosis of this tumour.

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