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# Solitary Fibrous Tumor/Hemangiopericytoma of the Ovary: A Rare Case Report and Literature Review

## Tumor fibroso solitario/hemangiopericitoma de ovario: reporte de un caso raro y revisión de la literatura

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**SUMMARY**

*Solitary fibrous tumor/hemangiopericytoma is an uncommon vascular lesion with pericyte differentiation. It can occur anywhere in the body. Most of the lesions are benign but the behavior is unpredictable. This study aimed to describe the presentation, diagnosis, and treatment of solitary fibrous tumors with a review of the literature. The difficulty of making an accurate diagnosis is highlighted. This is a case report of a 30-year-old woman who was admitted to the hospital with abdominal pain. The ultrasonography showed a cystic lesion in the right ovary. The patient finally underwent a hysterectomy with bilateral salpingo-oophorectomy. Histopathologically showed spindle to round cells that were arranged in clusters and around elaborate vasculature. It had slit-like and staghorn vessels. The immunohistochemistry stain showed CD34 positive. Microscopic findings with immunohistochemical staining should allow for a definite diagnosis and to differentiate this tumor from others.*

**Keywords**: *Fibrous tumor, hemangiopericytoma.*

**RESUMEN**

*El tumor fibroso solitario/hemangiopericitoma es una lesión vascular poco frecuente con diferenciación de pericito. Puede ocurrir en cualquier parte del cuerpo. La mayoría de las lesiones son benignas pero el comportamiento es impredecible. Este estudio tuvo como objetivo describir la presentación, diagnóstico y tratamiento del tumor fibroso solitario con una revisión de la literatura. Se destaca la dificultad de hacer un diagnóstico certero. El presente es un reporte de caso de una mujer de 30 años que ingresó al hospital con dolor abdominal. La ecografía mostró una lesión quística en el ovario derecho. La paciente fue finalmente intervenida de histerectomía con salpingooforectomía bilateral. Histopatológicamente mostró células fusiformes a redondas que estaban dispuestas en grupos y alrededor de una vasculatura elaborada. Tenía vasos en forma de hendidura y cuerno de ciervo. La tinción de inmunohistoquímica mostró CD34 positivo. Los hallazgos microscópicos con tinción inmunohistoquímica deben permitir un diagnóstico definitivo y diferenciar este tumor de otros.*

**Palabras clave:** *Tumor fibroso, hemangiopericytoma.*

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## INTRODUCTION

Your Hemangiopericytoma was first documented by Stout and Murray in 1942 for tumors originating in the pericyte and the presence of variably thick-walled, branching staghorn vessels. It may occur anywhere in the body where capillaries are found. Because the descriptions were vagus and included a number of lesions, the term solitary fibrous tumor will be used for this tumor. Solitary fibrous tumors occur in middle-aged adults (1,2). A solitary fibrous pleural tumor is a primary tumor arising from the pleura that can be a benign tumor and has various levels to potentially become malignant (3). Symptoms are mostly related to mass effect, depending on the size and site of the tumor (3). The primary treatment is usually total hysterectomy and bilateral salpingo-oophorectomy (1). However, microscopy findings supplemented by immunohistochemical staining allow a definite diagnosis and help to differentiate it from other tumors of the ovary. A Solitary fibrous tumour (SFT) is a rare mesenchymal neoplasm generally considered to occur in ubiquitous interstitial stem cells situated within soft tissues. Although most appear in the parietal or visceral pleura or peritoneum, they can be present in other extrapleural sites, including the mediastinum, lungs, liver, breasts, retroperitoneum, spine, meninges, and extracranial head and neck regions. Symptoms depend on the location and depth of the tumor (4).

## CASE DESCRIPTION

A 30-year-old woman was admitted to our hospital with lower right abdominal pain. The pain had been felt for 7 months and became burdensome 2 days before admission to the hospital. There were complaints of abdominal pain accompanied by nausea. The patient had a history of chronic gastritis and had undergone surgery on the right thigh 2 months before with a pathological diagnosis of lymphangioma. An acute abdominal sign was obtained on physical examination. Ultrasound examination showed a cystic mass in the right ovary. The patient underwent laparoscopy with an indication of an endometriosis cyst. The specimen was sent for an anatomical pathology examination. Gross examination showed a piece of brownish-white tissue, measuring 3cmx2cmx1cm; on the cut section, it was whitish and firm. Histologically, the tumor was composed of the spindle to round cells. The nuclei were vesicular with fine chromatin, some with clear nucleoli. The cells were arranged in clusters and around elaborate vasculature. Some vessels were thick-walled, and some formed a slit-like and staghorn pattern. Mitoses were not seen (Figure 1 A - D). Immunohistochemical stain showed positive for CD34. It was appropriate for hemangiopericytoma (Figures 1 E and F). Over the next 2 months, the patient underwent a hysterectomy and bilateral salpingo-oophorectomy. The final pathology report showed hemangiopericytoma.

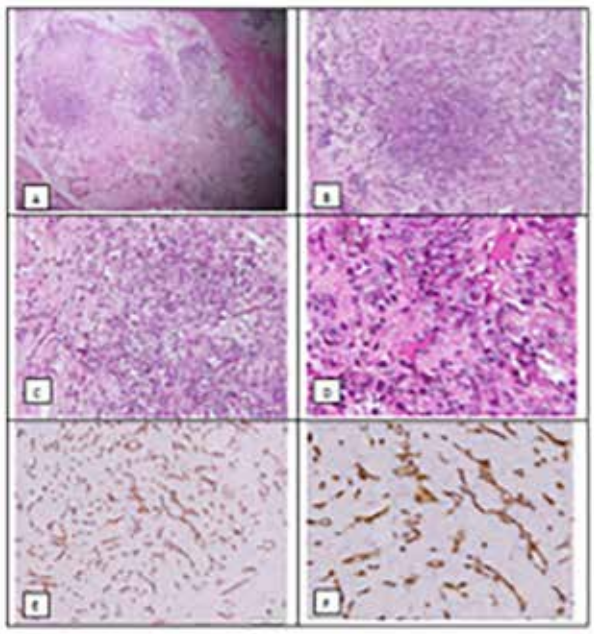

Figure 1. E and F. Immunohistochemical stain showed positive for CD34 (x100) (E), (x200) (F).

Solitary fibrous tumor with hypercellular and hypocellular fibrous area (HE, x40) (A). Tumor cells are arranged around elaborate vasculature (HE, x100) (B). The tumor consists of small and large vessels, slit-like and staghorn vessels (HE, x200) (C). Perivascular hyalinized (HE, x400)

(D). Immunohistochemical stain showed positive for CD34 (x100) (E), (x200) (F).

## DISCUSSION

The solitary fibrous tumor is a ubiquitous mesenchymal tumor of a fibroblastic type, which in the past was termed haemangiopericytoma (2). Hemangiopericytoma was first documented by Stout and Murray in 1942 for tumors originating in the pericyte and the presence of variably thick-walled, branching staghorn vessels (1,2). The solitary fibrous tumor was originally described in the pleura but is now more frequently found at extrapleural sites. It is almost exclusively found in deep soft tissue, particularly the thigh, pelvic fossa (5), retroperitoneum, and serosal surfaces (1).

We reported a solitary fibrous tumor in a 30-year-old female which was located in the ovary. The solitary fibrous tumor most often occurs in middle-aged adults (20-70 years), and both sexes are affected equally (2). It may occur anywhere in the body where capillaries are found (1,2,6). An ovary solitary fibrous tumor (hemangiopericytoma) is a very rare entity. Hasan et al. have reported a rare case of hemangiopericytoma of the uterus (4). Many benign tumors are small and located in subcutaneous tissue. The malignant tumors are more likely to be located deeper (7).

Symptoms are mostly related to mass effect, depending on the size and site of the tumor (8). Most tumors present as a painless and slowly growing mass. Large tumors may cause symptoms due to compression (2). In this case, the woman had pain because of the adhesiveness of the tumor with adjacent tissue. About 5 % of solitary fibrous tumors may cause hypoglycemia due to the production of insulin-like growth factor by the tumor (1,2,8). In this case, the laboratory findings were normal.

Grossly, the soft tissue tumor is usually well circumscribed. Tumor size ranges from 1 to 25 cm (2,8). It has a multinodular, whitish, and firm appearance. Myxoid and haemorrhagic changes are rare in benign tumors. Tumor necrosis factor-$\alpha$ (TNF-$\alpha$) (9) and infiltrative are observed in malignant tumors (2,10).

The solitary fibrous tumor has a wide range of appearances. This depends on the relative proportion of cells and fibrous stroma. It may be fibrous in form or cellular in form, but in many cases may have both features. The fibrous form is characterized by a combination of hypercellular (patternless) and hypocellular fibrous areas. There may also be perivascular hyalinization. The fibrous form is also characterized by being rounded, medium-sized, and thickened and hyalinized walls of the vessels. The hypercellular area consists of round to spindle cells, with vesicular chromatin (11) and pseudo inclusions of the nuclei. The cellular form resembles classical hemangiopericytoma. It consists of moderate to high cellularity, and a monotonous appearance of round to fusiform cells with indistinct cytoplasmic borders. The cells are arranged around elaborate vasculature, thin-walled staghorn vessels. Hemorrhage and cystic degeneration may be seen, mitoses are usually sparse, and necrosis is rare in this tumor (2,8,12). These features were found in this case.

Most cases of solitary fibrous tumors are benign. However, the behavior may be unpredictable. The benign solitary fibrous tumor tends to be non-recurring and non-metastasizing (2). The malignant tumor tends to metastasize. It can metastasize to the lungs, liver, and bone. The criteria of malignancy include tumor measuring >5cm, >4 mitoses per 10 hpf, and necrosis. In some of the literature, it is mentioned that high mitoses are the most reliable criterion for malignancy (8,13).

Because of the histological findings resembling hemangiopericytoma and the behavior is unpredictable, we confirmed the diagnosis with an immunohistochemical stain. Tumor cells express CD34+ in 90 % of cases. Some of the tumors may express EMA (30 %), SMA (20 %), and bcl-2 (30 %). Desmin, cytokeratin, and S-100 protein are usually absent (1,2,7,8). In this case, we examined the CD34+ expression because it has high sensitivity and a consistent diagnosis of the entity.

Some tumors may resemble solitary fibrous tumors. Sometimes the solitary fibrous tumor is difficult to distinguish from deep fibrous histiocytoma. Both of them have hemangiopericytoma-like vessels and may

express CD34+. However, solitary fibrous tumors rarely show storiform growth patterns. Malignant peripheral nerve sheath tumour (MPNST) also may be confused with solitary fibrous tumor, but MPNST has more fascicular architecture and focal expression of S100. The solitary fibrous tumor also should be distinguished from low-grade dedifferentiated liposarcoma. Both can express CD34+, but the dedifferentiated liposarcoma is usually less well-circumscribed (8,14).

It is stated that complete surgical resection is the only effective therapy for this tumor (15). Because of the unpredictable behavior of this tumor, the surgeon did a hysterectomy and bilateral salpingo-oophorectomy 2 months after the first laparoscopy. Long-term follow-up required an understanding of the behavior of the tumor.

## CONCLUSION

Hemangiopericytoma is a vascular tumor with pericytic differentiation. Soft tissue tumor is preferred as a better term than hemangiopericytoma. Most of the tumors are painless and have unpredictable behavior. They can be benign or malignant based on mitotic activity. Microscopic findings with immunohistochemical staining should allow for a definite diagnosis and to differentiate this tumor from others.

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