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**Imaging, pathology, and diagnosis of solitary fibrous tumor of the pancreas: A case report and review of literature**

Wang WW *et al*. SFT of the pancreas

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**Abstract**

BACKGROUND

A solitary fibrous tumor (SFT) is often located in the pleura, while SFT of the pancreas is extremely rare. Here, we report a case of SFT of the pancreas and discuss imaging, histopathology, and immunohistochemistry for accurate diagnosis and treatment.

CASE SUMMARY

A 54-year-old man presented to our hospital with pancreatic occupancy for over a month. There were no previous complaints of discomfort. His blood pressure was normal. Blood glucose, tumor markers, and enhanced computed tomography (CT) suggested a malignant tumor. Because the CT appearance of pancreatic cancer varies, we could not confirm the diagnosis; therefore, we performed endoscopic ultrasound-guided fine-needle biopsy (EUS-FNB). Pathology and immunohistochemistry were consistent with SFT of the pancreas. The postoperative pathology and immunohistochemistry were consistent with the puncture results. The patient presented for a follow-up examination one month after discharge with no adverse effects.

CONCLUSION

Other diseases must be excluded in patients with a pancreatic mass that cannot be diagnosed. CT and pathological histology have diagnostic value for pancreatic tumors. Endoscopic puncture biopsy under ultrasound can help diagnose pancreatic masses that cannot be diagnosed preoperatively. Surgery is an effective treatment for SFT of the pancreas; however, long-term follow-up is strongly recommended because of the possibility of malignant transformation of the tumor.

**Key Words:** Pancreas; Neoplasm fibrous tumor; Endoscopic ultrasound-guided fine-needle biopsy; Treatment; Case report

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**Core Tip:** We need to be more vigilant for indeterminate pancreatic masses, and then computed tomography and histopathology can play a very important role in clinical diagnosis. Surgery is an effective treatment for solitary fibrous tumor of the pancreas; however, long-term follow-up is strongly recommended because of the possibility of malignant transformation of the tumor.

**INTRODUCTION**

A solitary fibrous tumor (SFT) is histologically characterized as a mesenchymal tumor, probably fibroblastic in origin, located primarily in the pleura; however, it can be found in any other extrapleural region[1-3]. Extrapleural areas include the liver, peritoneum, kidney, and salivary glands[4-7]. SFT of the pancreas is rare, with only about 30 cases reported to date[1-3,6-35]. SFT of the pancreas is usually asymptomatic, and most are detected by physical examination, computed tomography (CT), or ultrasound as pancreatic masses[6,30,32]. The final diagnosis depends on histopathology and immunohistochemistry[7,31].

Here, we report a case of SFT of the pancreas and present the radiological and pathological differential diagnosis.

**CASE PRESENTATION**

***Chief complaints***

A 54-year-old man was admitted to our hospital with a pancreatic space-occupying mass of one month’s duration, identified on a physical exam.

***History of present illness***

A 54-year-old man had been one month before a medical CT finding of pancreas space-occupying lesions, with no adverse reactions, patients for further treatment at our hospital.

***History of past illness***

The patient had no other significant medical history. History of hypertension, diabetes, coronary heart disease, and other chronic disease was denied.

***Personal and family history***

The patient had no significant personal or family history.

***Physical examination***

The patient had no discomfort after the physical examination.

***Laboratory examinations***

There was no abnormal carcinoembryonic antigen [< 0.5 ng/mL (normal 0-5 ng/mL)], carbohydrate antigen 199 3.9 U/mL (average 0-7 U/mL), alpha-fetoprotein 2.4 ng/mL (normal 0-8.8 ng/mL), carbohydrate antigen 125 12.5 (average 0-30.2 U/mL). Fasting glucose was 5. 19 mmol/L (normal 3.89-6. 11 mmol/L).

***Imaging examinations***

A review of an abdominal enhanced CT showed a tumor of about 3 cm × 2 cm in the tail of the pancreatic body, showing uneven enhancement after enhancement, consistent with a malignant tumor (Figure 1).

**FINAL DIAGNOSIS**

A SFT of the pancreas.

**TREATMENT**

CT revealed a mass with mixed density and inadequate blood supply; these findings were inconsistent with a pancreatic tumor; therefore, we considered a pseudopapillary tumor and a non-functional pancreatic neuroendocrine tumor. We performed an ultrasound endoscopic tissue biopsy. The pathology and immunohistochemistry suggested SFT of the pancreas. After excluding contraindications to surgery and obtaining informed written consent, we performed a laparoscopic distal pancreatectomy with splenectomy. No significant adhesions were seen in the peripancreatic tissue. The pancreatic body was approximately 3 cm × 2 cm (Figure 2). Intraoperative frozen sections showed negative margins. Intraoperative blood loss was 100 mL and no blood transfusion was required.

The patient had no postoperative pancreatic fistula, abdominal infection, or bleeding. Ten days after surgery, he was discharged from the hospital after removing the drainage tube. One month after surgery, the patient returned to the hospital for examination. He did not complain of discomfort. The complete blood count, liver enzymes and renal function were normal.

Histopathological and immunohistochemical results of the postoperative specimen suggested an SFT of the pancreas of 3.0 cm × 2.5 cm × 1.0 cm, negative margins, no tumor involvement in the surrounding lymph nodes, and no tumor involvement in the spleen. Markers were as follows: Signal transducer and activator of transcription 6 (STAT6) (+), cluster of differentiation (CD) 34 (+), B cell CLL/lymphoma-2 (Bc1-2) (+), vimentin (+), CD99 (+), CD117 (-), Ki-67 (+40%), discovered on GIST-1 (+), transducin-like enhancer protein 1 (+), S- 100 (-), cytokeratin pan (pan) (-), somatostatin receptor 2 (-) (Figure 3).

**OUTCOME AND FOLLOW-UP**

No specific treatment was given after the patient was discharged from the hospital, and he had no complaints for three months after the procedure. He returned for regular follow-up. No abnormalities were found on complete blood counts, blood glucose, tumor markers, or CT.

**DISCUSSION**

SFT is a mesenchymal tumor comprising less than 2% of soft tissue tumors[36]. About 65% of SFTs originate from the pleura[3]; however, they can also be found in extrapleural areas[6], with only 34 cases reported to date, including the present case (Tables 1 and 2). SFT of the pancreas is extremely rare. We searched PubMed and Google Scholar for pancreatic tumors and SFT and found 34 cases. Of these, 14 (41. 1%) were male, and 20 (58.9%) were female. The mean age was 54. 17 ± 15.4, and the median age was 54; 17 patients had lesions in the pancreatic tumor head [three (17.6%) male and 13 (76.4%) female]. Seventeen had tumors in the tail of the pancreatic body [ten (58.8%) male and seven (41.2%) female]. The mean tumor diameter was 5.2 cm ± 3.8 cm. Of the 34 patients, 12 presented with pain (12/34), 12 were discovered on physical examination (12/34), four presented with jaundice (4/34), one presented with an abdominal mass (1/34), and five were detected by other means (5/34) (Table 1).

Most SFTs of the pancreas are detected by physical examination; clinical signs and symptoms include abdominal pain and jaundice. Because these are not typical symptoms, it is challenging to differentiate SFT from other pancreatic diseases. Histopathology and immunohistochemistry are the gold standards for diagnosis. We recommend ultrasound endoscopic aspiration biopsy for space-occupying pancreatic lesions that cannot be diagnosed on imaging.

Our preoperative diagnosis relied on ultrasound endoscopic puncture biopsy in the present case. The preoperative and postoperative pathological histological examination and immunohistochemistry were consistent with SFT of the pancreas with no tumor involvement in the peripheral lymph nodes, no tumor involvement in the incised margin of the pancreas, and no tumor involvement in the spleen.

The immunohistochemical differential diagnosis of SFT of the pancreas should include spindle cell tumors such as gastrointestinal stromal tumor (GIST), smooth muscle sarcoma, nerve sheath tumor, fibrous mucinous sarcoma, perivascular epithelioid cell tumor, and vascular tumors[3,16,20,37]. The immunomarkers of SFT of the pancreas include STAT6, CD34, bc1-2, vimentin, and CD99[34]. These features help to distinguish SFT from other mesenchymal tumors[34,37]. SFT expresses CD34 and vimentin in 80%-90% of cases and CD99 and bcl-2 in 70%. SFTs are usually negative for c-kit (CD117), smooth muscle actin, junctional protein, S-100 protein, and cytokeratin (markers for GIST, smooth muscle sarcoma, nerve sheath tumor, and fibrous mucinous sarcoma, respectively) are negative[3]. NAB2-STAT6 fusion is a driver mutation in SFT, where transcriptional repressors of the cytokinesis pathway are converted into transcriptional activators[31,38,39]. STAT6 has a sensitivity of 98% and a specificity of 85% for SFT and is therefore considered the most characteristic SFT marker[40,41]. In our case, the tumor was positive for STAT6, while CD34, bc1-2, vimentin, and CD99 were positive.

In this case, CT showed no enhancement in the arterial phase and heterogeneous enhancement in the venous area. We believe that it should be distinguished from neuroendocrine tumors, which show enhanced CT from the arterial phase to the portal venous phase[13,37], which makes it difficult for us to distinguish the disease, so many scholars before us also misdiagnosed it before surgery[1,10,11,13,26]. At the same time, we believe that it should also be differentiated from pancreatic cancer and solid pseudopapillary tumors of the pancreas. The imaging features of this tumor have been described in detail in our previous work on pancreatic tumors[42].

Most SFTs are benign[43], and malignant SFTs account for 10%-15%[30,39,44,45]. The histopathological features of malignant SFT: (1) Hypercellularity; (2) more than four mitotic figures per ten high-power fields; (3) nuclear pleomorphism; (4) hemorrhage and necrosis; (5) tumor diameter ≥10 cm; and (6) positive margins[15,21,46]. Ki-67 can also differentiate benign from malignant tumors, with a cutoff value of 0%-5% (indeterminate in 5%-10%) for benign tumors and > 10% for malignant SFTs[40,47]. In our case, our patient had a Ki-67 proliferation index of 40%; therefore, the tumor was possibly malignant. Because SFT of the pancreas is rare, there are no uniform treatment criteria; nevertheless, complete resection is the treatment of choice for intra-abdominal SFTs[1,7,10-12,15], and post-surgical follow-up is critical because SFTs have a high recurrence rate. Due to the increasing number of reported cases of SFT, we believe there will be a complete system of treatment.

**CONCLUSION**

Because of the non-specific clinical symptoms and radiological features of SFT of the pancreas, the diagnosis is challenging with preoperative radiological and laboratory examinations alone. A definitive diagnosis relies on histopathology and immunohistochemistry. In cases where the tumor is found in the pancreas, and the diagnosis cannot be confirmed, it is recommended to obtain histopathology with ultrasound aspiration. As this presentation is rarely reported, there is a lack of uniform treatment criteria, and surgery is effective. However, the tumor may lead to potential recurrence or metastasis; therefore, long-term follow-up is recommended.

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**Footnotes**

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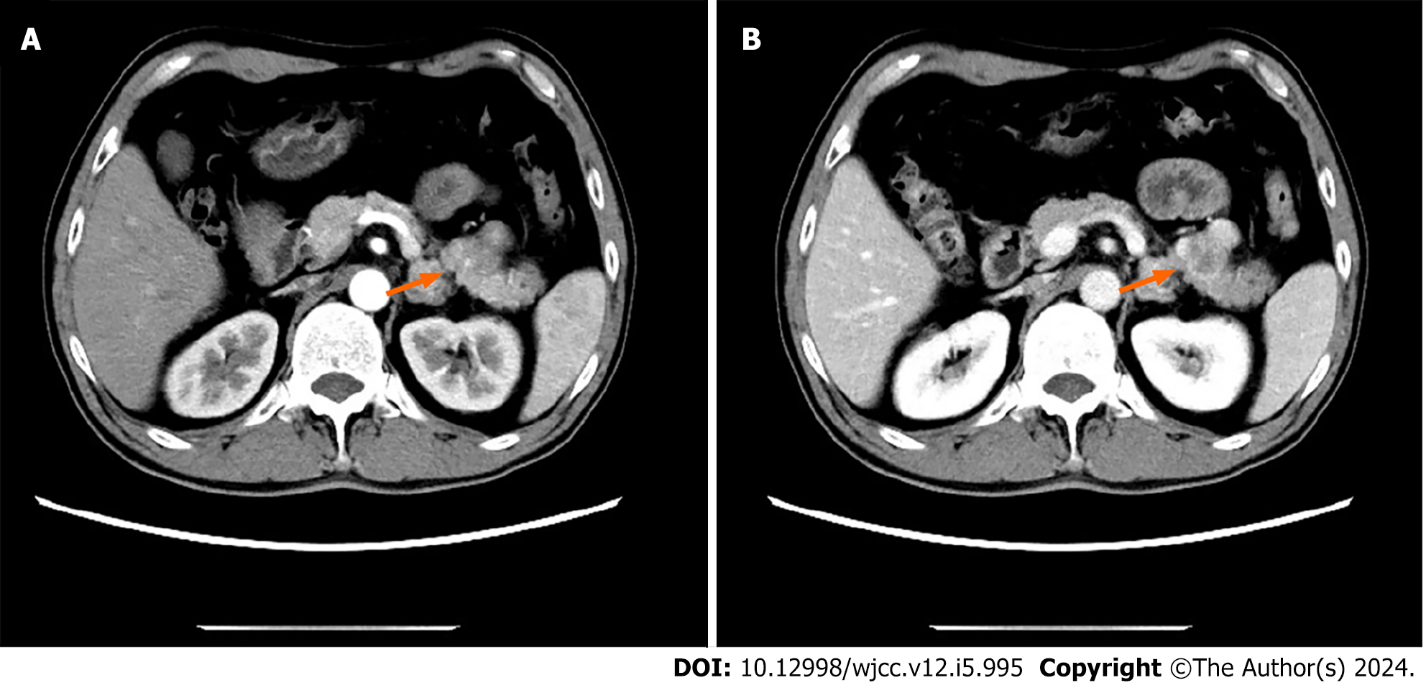
Grade C (Good): 0

Grade D (Fair): 0

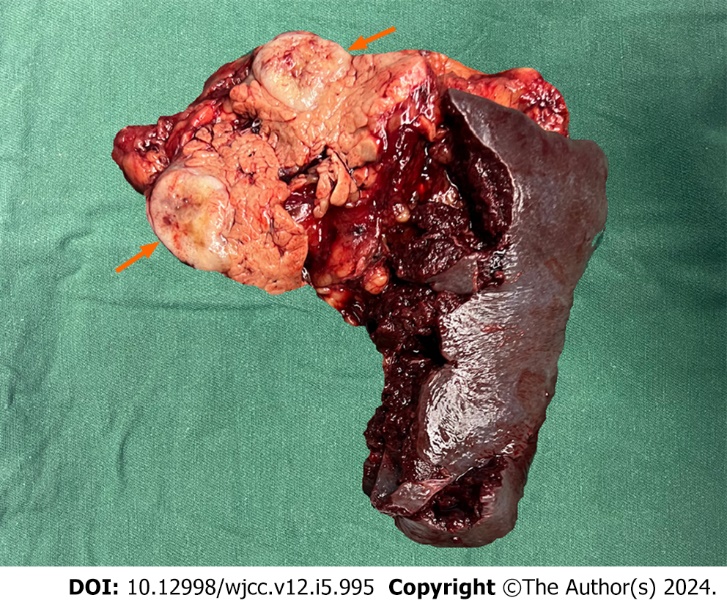
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**Figure Legends**



**Figure 1 Abdominal computed tomography scan showing a 5.52 cm × 2.82 cm × 2 cm mass in the pancreas (orange arrows).** A: No enhancement in the arterial region. B: Heterogeneous enhancement in the venous area.



**Figure 2 Postoperative surgical specimen: Pancreatic tail and spleen (tumor cut open chart) (orange arrows).**

地图

中度可信度描述已自动生成

**Figure 3 Representative results of hematoxylin and eosin and immunohistochemical staining of surgical specimens of solitary fibrous tumor of the pancreas.** A: Hematoxylin and Eosin staining (hematoxylin and Shuhong); B: Immunohistochemistry (original magnifcation of × 400) signal transducer and activator of transcription 6; C: CD34; D: CD99; E: Vimentin; F: Vimentin; G: Ki-67.

**Table 1 Characteristics of pancreatic solitary fibrous tumors**

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **No** | **Ref.** | **Age** | **Sex** | **Pancreatic site** | **Symptoms** | **Size (cm)** | **Pancreatic surgery** |
| 1 | Lüttges *et al[*1] | 50 | F | Body | Incidental | 55 | DP |
| 2 | Chatti *et al*[8] | 41 | M | Body | Abdominal pain | 13 | DP |
| 3 | Gardini *et al*[9] | 62 | F | Head | Abdominal pain | 3 | PD |
| 4 | Miyamoto *et al*[10] | 41 | F | Head | Abdominal pain | 18 × 15 | Enucleation |
| 5 | Kwon *et al*[11] | 54 | M | Body | Incidental | 76 × 6 | MS |
| 6 | Srinivasan *et al*[12] | 78 | F | Body | Back pain, weight loss | 5 | DP |
| 7 | Chetty *et al*[13] | 67 | F | Head | Incidental | 26 | PD |
| 8 | Ishiwatari *et al*[14] | 58 | F | Head | Incidental | 3 | PD |
| 9 | Sugawara *et al*[15] | 55 | F | Head | Incidental | 6 × 4 | PD |
| 10 | Santos *et al*[16] | 40 | M | Body | Incidental | 3 | Enucleation |
| 11 | Tasdemir *et al*[17] | 24 | F | Body | Epigastric pain | 11 | Enucleation |
| 12 | van der *et al*[18] | 67 | F | Head | Abdominal pain | 28 × 16 | Enucleation |
| 13 | Chen *et al*[19] | 49 | F | Head | Abdominal pain | 13 | PD |
| 14 | Hwang *et al*[20] | 53 | F | Head | Incidental | 52 × 45 × 40 | PHR |
| 15 | Baxter *et al*[21] | 58 | F | Head | Abdominal pain | 35 × 3 | LPD |
| 16 | Estrella *et al*[22] | 52 | F | Head | Jaundice | 15 × 10 × 10 | LPD |
| 17 | Han *et al*[23] | 77 | F | Head | Jaundice | 15 × 14 | Biopsy |
| 18 | Murakami *et al*[24] | 82 | M | Body | Hypokalemia hypertension, edema | 6 | DP |
| 19 | Spasevska *et al*[3] | 47 | M | Head | jaundice | 35 × 2 × 18 | LPD |
| 20 | Paramythiotis *et al*[7] | 55 | M | Body | Abdominal pain | 31 × 28 | DP |
| 21 | D'Amico FE *et al*[25] | 52 | M | Body | Incidental | 12 | DP |
| 22 | Oana *et al*[26] | 73 | M | Head | Abdominal discomfort | 65 × 55 | Enucleation |
| 23 | Sheng *et al*[27] | 1 | M | Head | Jaundice | 20 | DP |
| 24 | Geng *et al*[28] | 48 | M | Body | Hypoglycemia | 65 × 5 | DP |
| 25 | Qian *et al*[29] | 46 | M | Body | Hypoglycemia | 70 × 61 | DP |
| 26 | Rogers *et al*[30] | 37 | F | Head | Abdominal pain | 23 | PD |
| 27 | Taguchi *et al*[31] | 60 | M | Head | Palpable mass | 9 × 7 × 7 | PD |
| 28 | Jariwalla *et al*[32] | 64 | F | Body | Abdominal pain | 19 | DP |
| 29 | Marotti *et al*[33] | 75 | F | Body | Incidental | 13 | Enucleation |
| 30 | Addeo *et al*[6] | 59 | M | Body | Incidental | 4 | DP |
| 31 | Rodriguez *et al*[2] | 48 | F | Body | Abdominal pain | 13 × 10 × 95 | TP |
| 32 | Jones *et al*[34] | 61 | F | Body | NA | 27 | DP |
| 33 | Liu *et al*[35] | 54 | F | Head | Incidental | 31 × 23 | LDPPHRt |
| 34 | Present case | 54 | M | Body | Incidental | 3 × 2 | DP |

LDPPHRt: Laparoscopic duodenum-preserving pancreatic head resection; Ms: median segmentectomy; PHR: Pancreatic head resection; TP: Total pancreatectomy; PD: Pancreaticoduodenectomy; DP: Distal pancreatectomy.

**Table 2 Histological features and outcomes of pancreatic solitary fibrous tumors**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **No** | **Ref.** | **Immunohistochemistry** | **Outcome** | **Follow-up** |
| 1 | Lüttges *et al[*1] | CD34, CD99, Bcl-2, vimentin | Alive | 20 months |
| 2 | Chatti *et al*[8] | CD34, CD99, Bcl-2, vimentin | Death | 3 d |
| 3 | Gardini *et al*[9] | CD34, CD99, Bcl-2, vimentin, SMA | Alive | 16 months |
| 4 | Miyamoto *et al*[10] | CD34, Bcl-2 | Alive | 7 months |
| 5 | Kwon *et al*[11] | CD34, CD99, vimentin | NA | NA |
| 6 | Srinivasan *et al*[12] | CD34, Bcl-2 | Alive | 7 months |
| 7 | Chetty *et al*[13] | CD34, CD99, Bcl-2 | 42 mo | 6 v |
| 8 | Ishiwatari *et al*[14] | CD34, Bcl-2 | Alive | 42 months |
| 9 | Sugawara *et al*[15] | CD34 | NA | NA |
| 10 | Santos *et al*[16] | CD34, betacatenin | NA | NA |
| 11 | Tasdemir *et al*[17] | CD34, Bcl-2, beta-catenin, vimentin, Ki67 < 2% | Alive | 3 months |
| 12 | van der *et al*[18] | CD34, CD99, Bcl-2 | NA | NA |
| 13 | Chen *et al*[19] | CD34, Bcl-2, vimentin, CD68, muscle-specific actin | Alive | 30 months |
| 14 | Hwang *et al*[20] | CD34, Bcl-2, muscle-specific actin, CD10, ER, PR | Alive | 30 months |
| 15 | Baxter *et al*[21] | CD34, Bcl-2 | NA | NA |
| 16 | Estrella *et al*[22] | CD34, Bcl-2, keratin (rare), p16, p53 | Alive | 40 months |
| 17 | Han *et al*[23] | CD34, CD99 | No progression | 10 months |
| 18 | Murakami *et al*[24] | STAT6, CD34, Bcl-2, ACTH, POMC, NSE | Death | 4 months |
| 19 | Spasevska *et al*[3] | CD34, vimentin, CD99, Bcl-2, nuclear betacatenin | Death | 1 wk |
| 20 | Paramythiotis *et al*[7] | CD34, CD99, Bcl-2 vimentin, S-100 | Alive | 40 months |
| 21 | D'Amico FE *et al*[25] | STAT6, CD34 | Alive | 24 months |
| 22 | Oana *et al*[26] | CD34, Bcl-2 | Alive | 36 months |
| 23 | Sheng *et al*[27] | CD34, vimentin, SMA, Ki67 < 3% | Alive | 12 months |
| 24 | Geng *et al*[28] | STAT6, CD34, Bcl-2, CD31, PHH-3, D2-40, Ki67 > 10% | Alive | 6 months |
| 25 | Qian *et al*[29] | STAT6, CD34, Bcl-2, Ki67 10% | Alive | 10 months |
| 26 | Rogers *et al*[30] | STAT6, CD34, Bcl-2, CD99 | Alive | 4 months |
| 27 | Taguchi *et al*[31] | STAT6, CD34, Bcl-2, vimentin, cytokeratin AE1/AE3 | Alive | 12 months |
| 28 | Jariwalla *et al*[32] | STAT6, CD34 | NA | NA |
| 29 | Marotti *et al*[33] | STAT6, CD34 | Alive | 6 months |
| 30 | Addeo *et al*[6] | STAT6, CD34, Bcl-2, Ki67 7% | NA | NA |
| 31 | Rodriguez *et al*[2] | STAT6 | Alive | 12 months |
| 32 | Jones *et al*[34] | STAT6, CD34 | Alive | 1 months |
| 33 | Liu *et al*[35] | CD34, STAT6, CD99 | Alive | 6 months |
| 34 | Present case | TAT6, CD34, Bc1-2, Vimentin, CD99, Ki67 40% | Alive | 3 months |

STAT6: Signal transducer and activator of transcription 6; ER: Estrogen receptor; PR: Progesterone receptor; SMA: Smooth muscle actin; NA: Not applicable.



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