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Silent Gigantic Solitary Fibrous Tumour of the Pleura: 2 Rare Case Reports and Review of the Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Author TTV was a main surgeon, designed the study, wrote the protocol and reviewed the drafted manuscript. Authors LQD and HTB wrote the first draft of the manuscript, managed the analyses of the study, managed the literature searches, followed up the patients during the period. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Solitary fibrous tumours of the pleura (SFTP) are rare primary tumours that arise from the submesothelial tissue. They are mostly benign and asymptomatic, and by growing, they cause chest pain, irritating cough, and dyspnoea due to the pressure created on the surrounding structures. Immunohistochemical analysis has an expanding role in diagnostic. Surgical resection is the main treatment. We reviewed two patients who had undergone complete surgical resection for treatment and followed up periodically for two years at the University Medical Center at Ho Chi Minh City, Ho Chi Minh City, Vietnam.

Keywords: Solitary fibrous tumours (SFTs); pleura.

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1. INTRODUCTION

Primary tumours of the pleura include two major categories: diffuse and localized pattern. The diffuse pattern is pleural mesothelioma which originates from the mesothelial cells of the pleura. This type of tumour is usually associated with asbestos exposure and highly malignant. The other one is the localized tumour, also called solitary fibrous tumour, which arises from the submesothelial mesenchymal layer. After some first cases were reported, there were a lot of arguments about the aetiology of this type of neoplasm. Therefore, there were many terms which applied to this neoplasm such as localized submesothelial fibroma, pleural fibroma, localized fibrous mesothelioma, and pleural mesothelioma.

However, with the development of immunohistochemistry techniques and electronic microscopy, this kind of tumour was finally named as solitary fibrous tumours of the pleural (SFTP) to distinguish it from malignant pleural mesothelioma. We herein report two rare cases of patients with SFTP which underwent successful resections.

2. CASE PRESENTATION 1

A 40-year-old female patient had unremarkable medical history and with no history of exposure to asbestos or any chemical occupational factors, presented symptoms including fatigue, chest pain and a sensation of compression in the left hemithorax which had increased gradually for a year. Examination of the chest revealed dullness, and diminished breath sounds on the left side.

On the chest X-ray figure, a large mass was detected in the left hemithorax. Chest computed tomography with contrast demonstrated a giant mass with a diameter larger than 20 centimetres. It was solid, heterogeneous, well-demarcated and practically invaded over the entire left pleural cavity. The mediastinal structures were shifted mediastinal to the right, with no evidence of chest wall or mediastinal invasion (Fig. 1). Laboratory examination such as haemoglobin and blood counts were in the normal range, and tumour markers had the following results: CEA 1.4ng/ml; beta HCG <1.2; CA19.9 2 U/ml; Cyfra 21-1: 1.4 ng/ml, all items were in the normal range.

On 27th September 2016, a left lateral thoracotomy was performed through the sixth intercostal space with a removal of the seventh rib. We could easily visualize the encapsulated circumscribed gigantic tumor which estimated about 20x25x20 centimetres, growing from the visceral pleura of lingular segment, causing atelectasis of the left inferior and superior lobes. The giant tumour has adhered to the diaphragm and inferior lobe. The tumour which weighed 2.6 kilograms was resected (Fig. 2). The postoperative course was uneventful, and the patient was discharged on a ninth day. A chest X-ray after surgery proved that the right upper lobe had expanded completely. The patient is currently alive without any recurrence two years post-surgery.

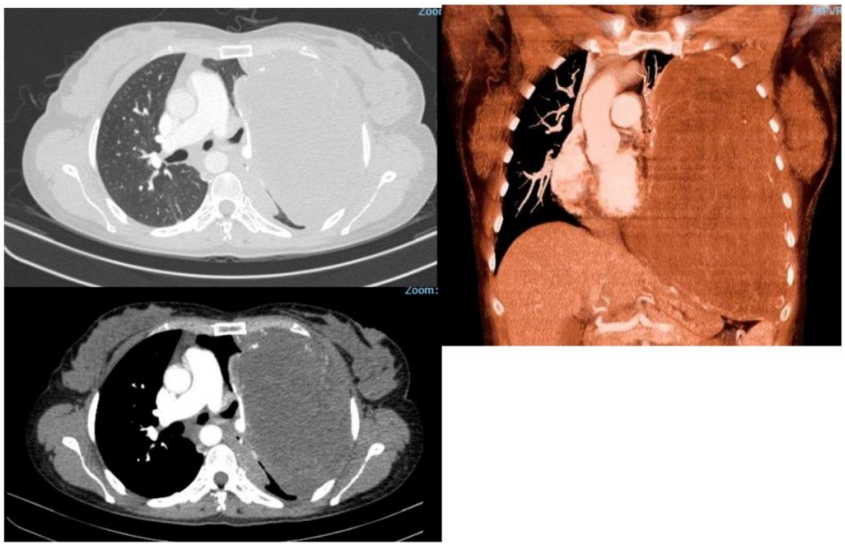

Fig. 1. Computed tomography (CT) scan images

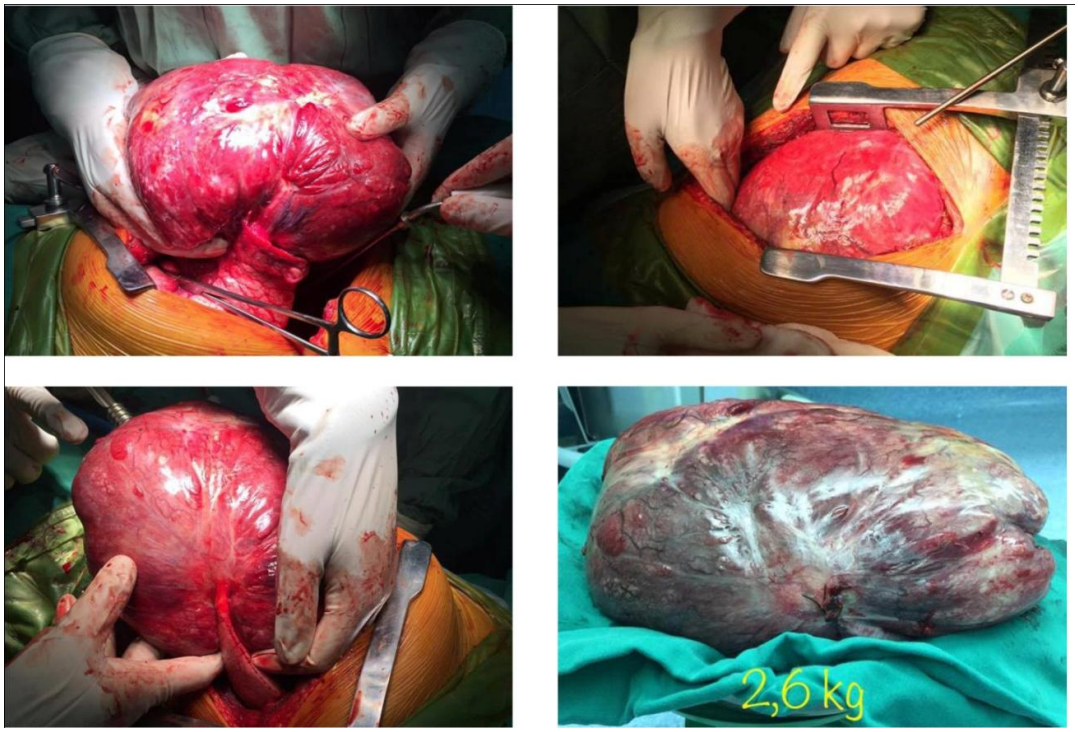

Fig. 2. Giant tumour specimens after resection

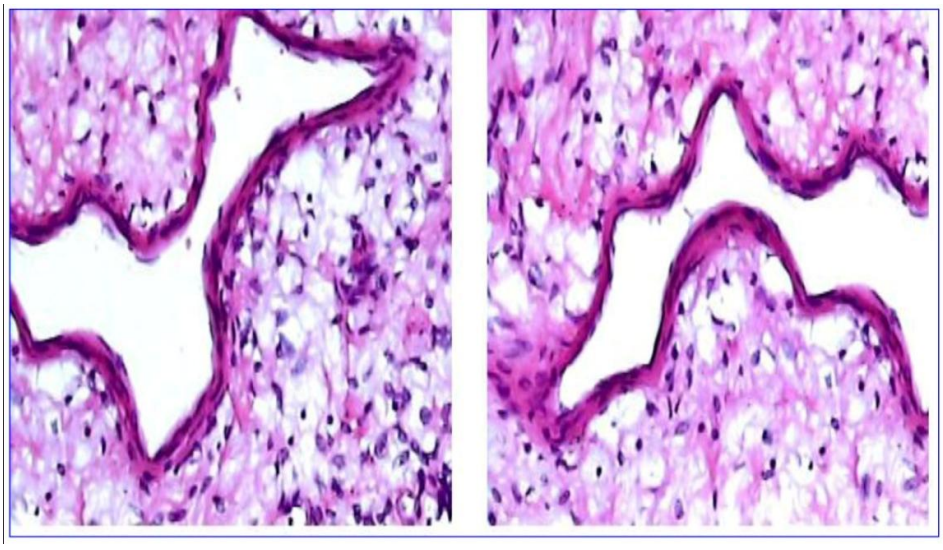

Fig. 3. Microscopic (histologic) images

The surface was lined with a shiny, smooth and thin capsule with clear boundaries. The sectioned surface was grey-white and homogeneous. Some small calcifications were present. A microscopic specimen of the tumour showed patternless spindle-shaped cells mixed with amounts of collagen and hyalinization stroma. The spindle cells characterized by smooth cytoplasm and oval nuclei concentrated around the staghorn blood vessels. Mitosis was rarely present (Fig. 3).

Basing on the morphology and histopathological examination, the diagnosis of the benign giant

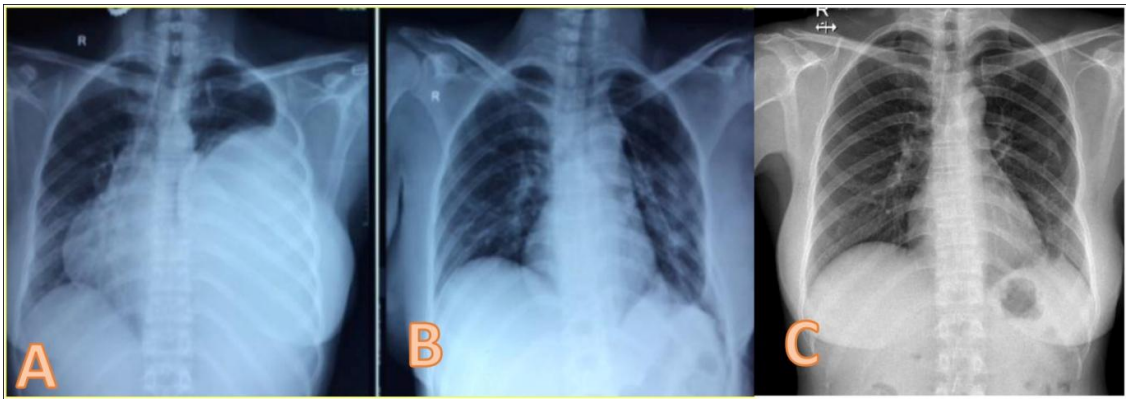

Fig. 4. Chest radiography at the preoperative (A) and postoperative period (B) and after one year (C)

solitary fibrous tumour of the pleura was confirmed. The postoperative course was uneventful, and the patient was discharged on a ninth day. A chest X-ray after surgery proved that the right upper lobe had expanded completely (Fig. 4). The patient is currently alive without any recurrence 2 years post surgery.

3. CASE PRESENTATION 2

A 69-year-old female patient, who was asymptomatic, visited the University Medical Central after an abnormal shadow on a plain chest X-ray (Fig. 5). She also had no history of smoking and exposure to asbestos or any chemical occupational factors. A chest computed tomography revealed a mass about 10x8x3 centimetres in size (Fig. 5). It was also solid, heterogeneous, well-demarcated and no sign of chest wall or mediastinal invasion. All laboratory examination results were within normal range.

Our surgical team decided that the patient was a candidate for surgery, in order to confirm the diagnosis and to provide adequate treatment. On 12th April 2016, a left lateral thoracotomy was performed through the seventh intercostal space. A large tumour rose from the visceral pleural of left inferior lobe. It was oval- shaped, well-demarcated, not adhered to other organs. It was easy to remove completely (Fig. 6).

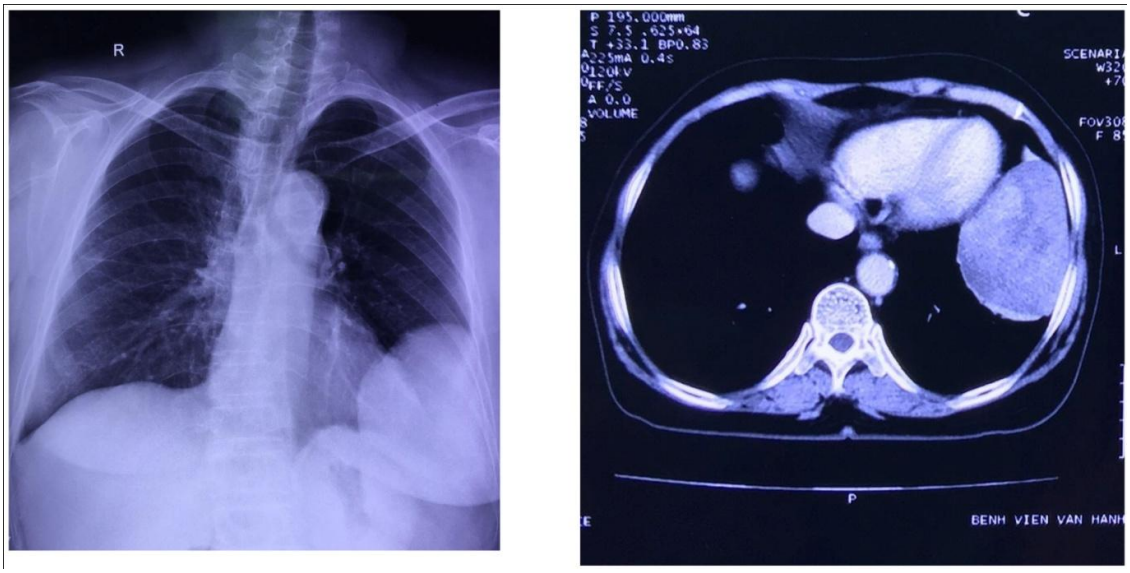

Fig. 5. Chest radiography and computed tomography (CT) scan images

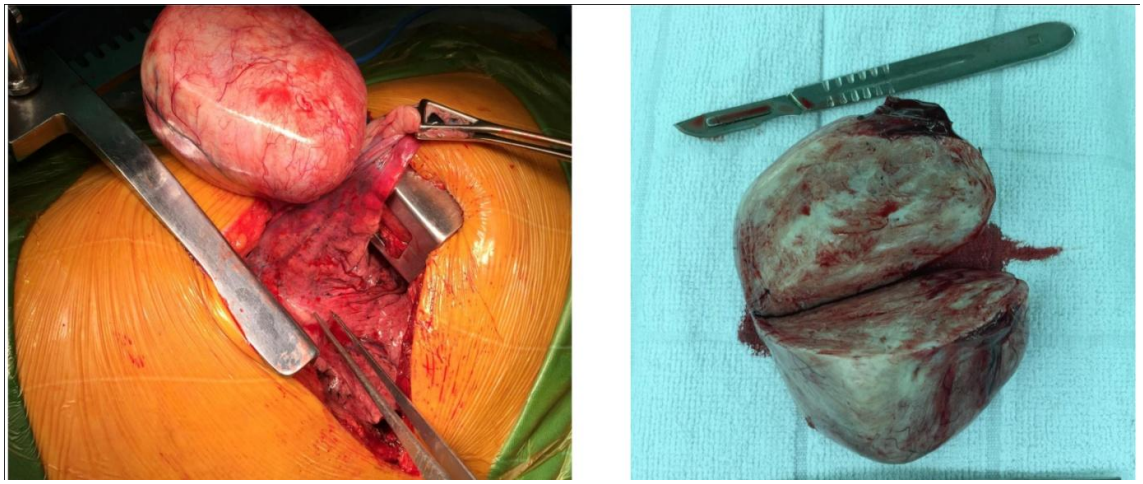

Fig. 6. The tumour specimen after resection

The pathohistological images were similar to the Case Presentation 1. It was characterized by spindle-shaped cells and staghorn blood vessels around. Immuno-histochemical features were as follows: CD 34 (+), actin (-), Desmin (-), Ki67 (+) < 5%, S100 (-). The final diagnosis was SFTP. The postoperative course was uneventful, and the patient was discharged on the fifth day. She was still alive without recurrence at the two year follow-up.

4. LITERATURE REVIEW

Solitary fibrous tumours of pleura (SFTP) are rare primary neoplasm that arises from the submesothelial tissue. They have an incidence of approximately 5% of all neoplasms involving the pleura [1]. Although these neoplasms were reported firstly in pleura, some reports showed that they also originated primarily in the other sites such as the lung, pharynx, epiglottis, nose, salivary gland, thyroid, breast, kidney, bladder, spinal cord and meninges [2]. Wagner E. Das was the first to describe a localized primary pleural tumour in 1870. In 1931, Klemperer and Rabin set a hypothesis that this kind of tumour was a localized mesothelioma, covering the intact layer of mesothelial cells, stemming from structures under the mesothelial layer [3]. Due to the development of electron microscopic and immunohistochemical techniques, almost experts finally confirmed that SFTP arise from deeper-lying mesenchymal cells of the pleura [4]. The term “localized mesothelioma” was altered by the term "solitary fibrous tumors of the pleural" (SFTP).

5. EPIDEMIOLOGY

There are approximately 900 cases which have been reported in the global literature. It is an uncommon tumour, occur equally in male and female patients and the incidence of about 2 cases per 1.000.000 people [5,6]. All age groups (5 to 87 years) were reported and the highest incidence referred between 60 and 70 years of age. Generally, there is not any evidence of correlation with genetic factors, and in contrast with malignant mesothelioma, there is no relation to the exposure of asbestos or any other environmental factors [7].

6. HISTOPATHOLOGICAL CHARACTERISTICS

SFTPs are often large in size with some reported cases of giant tumours measuring over 20 cm in diameter [4]. In macroscopic images, the majority of them arise originally from the visceral pleura (80%), they are usually well-circumscribed masses with lobular or smooth external surfaces and are encapsulated within a thin, glistening translucent serosa through which a network of prominent blood vessels may be seen. The cut surface appears grey-white to tan with a whorled pattern and may show areas of haemorrhage and necrosis. Benign tumours may show hemorrhagic and necrotic areas, but these features usually predominate in the malignant forms [7]. In microscopic images, SFTPs are characterised by the appearance of spindle-shaped cells and staghorn- shaped blood vessel branches around [8]. In histopathological classification, there are four types of variants:

classic, giant cell angiofibroma, cellular and fat-forming modification.

Histologic signs of malignancy include: (1) high mitotic counts, defined as more than 4 mitoses per 10 high-power fields; (2) mild to marked pleomorphism based on nuclear size, irregularity, and nuclear prominence; (3) bundles of high cellularity with crowding and overlapping of nuclei; (4) presence of necrotic or hemorrhagic zones; and (5) stromal or vascular invasion [5].

Immunohistochemistry may be quite useful in diagnosing and differentiating the SFTP from mesotheliomas and sarcomas. CD34 and BCI2 are useful with the positive rate of 88.7% and 96.2% respectively. Some recent reports show the role of maker STAT6 in diagnosing SFTP [9].

7. CLINICAL FEATURES

Small SFTP is usually asymptomatic. They are only detected incidentally on chest X-ray images. When the tumours grow and press on the surrounding structures, they may appear some symptoms such as coughing, dyspnoea, and chest pain. Rarely, there are signs of hemoptysis, obstructive pneumonitis or atelectasis [2]. In large tumours, digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre Marie Bamberg syndrome) may be present or signs of hypoglycemia on account of insulin-like growth hormone release (Doege Potter syndrome) [4]. Lary A. Robinson reviewed the table of incidence of symptoms in patients with SFTP (Shown in Table 1 [10,11,12]).

8. RADIOGRAPHIC FEATURES

Plain chest X-ray plays a role in detecting this kind of tumour. A CT with contrast provides valuable data on the exact location of SFTP, its relation to surrounding structures, tumour homogeneity or potential bleeding areas or necrosis, chest wall destruction and the presence of pleural effusion. However, a CT scan cannot differentiate between benign and malignant SFTP cases. Large tumours are more likely to be malignant, with distinct heterogeneous structure, not clearly separated from the surrounding environment and in potential presence of pleural effusion, a PET – CT or MRI can be performed to distinguish [4].

9. TREATMENT

In most cases, the treatment method for this kind of tumour is surgical resection via thoracotomy or thoracoscopy approach. Because SFTP is not a primary lung, it arises from the visceral pleura, saving lung in safe restriction which is about 1-2 cm from the tumour's margin is considerate. Some sessile tumours which develop from the upper pleura,diaphragm, or mediastinum tend to recurrence and require a wide local extrapleural excision.

When there is surely evidence that the tumour is malignant, resection of a portion of the chest wall may be necessary. Malignant SFTP tends to be larger in size and more invasive, and they are almost never pedunculated. Adhesions may be seen in the larger tumours, which often complicate the resection [2]. In cases of larger SFTP, the continuation of surgical treatment with adjuvant chemotherapy is indicated Park et al. have found that the combination of temozolomide and bevacizumab had high rates of overall response and long-term disease control [13].

Table 1. A summary of common symptoms

Symtoms	Okike et al. 1978, N=52	England et al. 1989, N=138	Sung et al. 2005, N=63
Asymptomatic	54%	67%	43%
Cough	33%	12%	43%
Chest pain	23%	19%	17%
Dyspnea	19%	11%	17%
Fever	17%	1%	1%
HPO	19%	-	2%
Weight loss	6%	2%	3%
Hemoptysis	2%	-	-
Pneumonia	2%	-	-

HPO: Hypertrophic pulmonary osteoarthropathy

10. PROGNOSIS

SFTPs are benign in 78-88% of cases. Surgical resection is always useful with an overall long-term cure rate for all patients of 88% to 92% [2]. However, a case report of recurrences has occurred as long as 17 years after the complete resection [14]. According to review of some case series of recurrence which relates to histologic and morphologic data, Perrot et al announced a classification of SFTP according to tumour features, and prognosis: (1) benign pedunculated tumours had a 2% recurrence rate, (2) benign sessile tumors had an 8% recurrence rate, (3) malignant pedunculated tumors had a 14% recurrence rate, and (4) malignant sessile tumours had a 63% recurrence rate and a 30% mortality, with most deaths occurring within 24 months [5].

11. DISCUSSION

In the two clinical cases we have described above, one patient was completely asymptomatic although the size of the tumour was quite large (9x8x3 cm), and the other patient had a giant tumour (20x25x20 cm), weighing 2.6 kg, presenting symptoms such as fatigue, chest pain and a sensation of compression that increased gradually over a period of one year. Thus, these cases are similar to those found in the literature. Both cases were eradicated by the thoracotomy, with no recorded complications.

Histopathological results of the two cases showed that the two tumours were characterized by smooth masses, the appearance of spindle-shaped cells, and staghorn blood vessels around. All of these characteristics are the same as those mentioned in the literature.

12. CONCLUSIONS

Solitary fibrous tumours of the pleura are a rare neoplasm, characterized in microscopies by spindle cells and staghorn blood vessels. The patients are usually asymptomatic in the early stages, the symptoms only appear when the tumour has compressed the surrounding organs. Most of them are benign, and surgical removal of the entire tumour is the primary method of treatment. Definitive diagnosis and differential diagnosis are often based on histopathology and immunohistochemistry. Prognosis is usually good. However, it can be recurrent or malignant. Therefore, long-term follow-up after surgery is necessary.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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