ABSTRACT

Hemangiopericytoma is a rare disease. We report the first case presented with such disease at our hospital. The patient was first diagnosed with is vascular tumour 10 years ago and was discharged without chemotherapy. The patient was recently hospitalised due to tumour recurrence.

The patient was Mrs. Tran Thi Kim H, born in 1970, married, had 4 children, and was a farmer. The patient lived at An thoi Dong, Can Gio district, and was admitted on 26/9/2019 due to a recurrent left lung tumour. The patient suffered from headache and vertigo in the past 2 years and she underwent surgical removal of a lung tumour 10 years ago at Binh Dan Hospital. Physical examination was normal. Laboratory results were within normal range, except a mild anaemia.

We had planned to use thoracoscopy but after general anaesthesia, the left lung could not be deflated and therefore the surgery was postponed. A week after, we attempted to use thoracotomy. We observed a tumour that adhered to the thoracic wall and to the left lung. We dissected the adhesion and removed the tumour, haemostasis was achieved by suturing with a Vicryl 3.0 stitch. The surgical incision was closed and a chest drainage was placed. Post-operative follow-up was uneventful and the patient stayed in the hospital for 15 days. Pathological studies:

Biopsy with MSCT guidance revealed a diagnosis of undifferentiated non-small cell lung cancer; surgical biopsy showed that this was a lymphocyte with big cell tumour. Finally, the diagnosis of lung hemangiopericytoma was confirmed by immunohistochemistry. Whether chemotherapy should be indicated for this patient is being considered.

Key word: Hemangiopericytoma

I. CLINICAL CASE

Hemangiopericytoma is a rare disease. We report the first case presented with such disease at our hospital. The patient was first diagnosed with is vascular tumour 10 years ago and was discharged without chemotherapy. The patient was recently hospitalised due to tumour recurrence.

The patient was Mrs. Tran Thi Kim H, born in 1970, medical record number 2019/24341. The patient lived at An Thoi Dong, Can Gio, Ho Chi Minh City and was hospitalised on 26/9/2019 due to a recurrent left lung tumour. She had undergone surgical removal of a left lung tumour in 2009 at Binh Dan Hospital. During this episode, she had been treated at Ho Chi Minh Oncology Hospital for 20 days and MSCT showed a vascular tumour that led to her transfer to Binh Dan Hospital.

History of present illness: In the past two years, the patient suffered from vertigo, headache, weight loss, and back pain. Chest CT scan at Hoa Hao showed a vascular-type lung tumour. She had four children and underwent surgical sterilisation at 30 years of age.
In the first surgery, left thoracotomy was performed and the tumour was removed. The patient stayed in the hospital for 14 days. Pathological studies showed that it was a benign tumour and the patient was discharged without chemotherapy. Since then, the patient was asymptomatic until 2 years ago when patients started to experience hot and cold feeling. In the past 2 months, the patient usually complained of fever with no chest pain. At admission, besides the tumour, the patient had no pain with mild left-sided chest discomfort. She was referred to Binh Dan Hospital for surgical removal of the recurrent tumour.

The patient was an average-sized woman, there was a surgical scar on the left thorax at the 5th intercostal space that extended from the anterior to posterior chest line and a broken 6th rib (due to surgical retractor). During the first thoracotomy, the tumour was cut in piece and removed completely, the pleural space was cleaned, the incision was closed and the drainage was placed. Three days after surgery, the surgical wound was healing well and the lungs were deflated. A week later, the pathology results showed that it was a benign tumour so no chemotherapy was indicated. After surgery, the patient was asymptomatic and therefore did not have follow-up visits. In the past two years, she experienced headache, vertigo and one week before hospitalisation, she suffered from left-sided chest pain. During examination, chest x-ray showed a tumour at the base of the left lung and the patient was referred to Binh Dan Hospital. 32-slide Contrast-enhanced CT scan showed a 17x22 mm mass at the base of the left lung with enhanced contrast. No other lesions were found. The initial diagnosis was pleural tumour at the base of the left lung.

Figure 1: Chest x-ray in 2008
Figure 2: A: MSCT for the first surgery: Big left lung tumour that occupied all the left thoracic cavity
B: The mediastinum is pushed to the right by the tumour. A mass with uneven density with adjacent big vessel, suggesting that this is a haemangioma.
C: Mrs. Tran Thi Kim H., 1970 ; D: The old surgical scar

The thoracic department ordered laboratory tests and found no other lesions except for the left lung tumour. Blood tests results were as follows: B-type blood, Hct 37.2%, HGB 11.1 g/dL, Leukocyte count 12.24K/uL, PLT 265 K/uL (9/10/2019). After surgery the leukocyte count was 18.32 K/ul, PLT 313 K/ul (10/10/019). Electrolytes levels were as follows: Na+ 134, K+ 4.1, Cl− 100, Ca ++ 106 mmol/L. Urinalysis results were within normal limits. Sputum test showed no alcool acid resistant bacilli (27/9/2019). Echocardiography showed normal heart function (EF 74%). ECG showed sinus rhythm. Pulmonary functioning test was normal, chest X-ray showed a broken 6th rib bone and a suspected tumour at the left lung base (7/10/2019). Chest biopsy with MSCT guidance showed that this was an undifferentiated adeno-carcinoma.

The patient underwent the second surgery on 17/10/2019 under general anaesthesia. (Surgical team included Prof. Van Tan, Dr. Nguyen Van Viet Thanh, Dr. Doan Hung Dung, anaesthetist Nguyen Cao Thuy Hang, Ton That Hung).
**Indication for surgery:** Removal of the tumour and a part of the left lung

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**Figure 3:** Anterior-posterior chest X-ray and MSCT of the recurrent tumour

Positioning the patient: 90 degree to the right. Skin incision was performed on the previous surgical scar. We opened the 5-6th intercostal space to approach the thoracic cavity. The tumour was very adhesive to the lung and the chest wall. The dissection of the tumour was difficult and the mass was broken. The whole tumour and a part of the left lung base were removed. Haemostasis was carefully achieved by suturing with a Vicryl 3.0 stitch. A chest drainage was placed. The duration of the surgery was 100 minutes and the volume of blood loss was 200 ml.

The attempt of thoracoscopy was not successful because the left lung could not be deflated and the surgery was postponed. A week later, the patient underwent open surgery as this was re-operation and the pleural would be very adhesive.

Pathology studies of the tumour: B-type lymphoma. Immunohistochemistry: lung hemangiopericytoma.

**II. DISCUSSIONS:**

Lung hemangiopericytoma is a rarely seen tumour and it could be either benign or malignant [1, 2, 3]. The first removal, chemotherapy was not indicated since pathological results were negative. After 10 years, the tumour recurred and the patient was re-operated at Binh Dan Hospital to remove the tumour and a part of the left lung. The first pathological result was cancer (CT-guided needle biopsy), the second was B-type lymphoma, and finally with immunohistochemistry, hemangiopericytoma. The problem in question is whether chemotherapy should be indicated.

This tumour could be seen from new-borns to 60 years of age and in theory, the tumour rarely develops in places with micro vessels [7, 11,13,14]. The tumour originates from the tissue surrounding the blood vessels. Tumour cells are smooth muscle cells that can change and have elastic features that could expand the micro vessels. When reviewing 197 patients, Stout showed that this tumour usually first develops from peripheral tissue [6,17]. This type of tumour originating from the lung is rarely seen. Many patients have no symptoms when the tumour is smaller than 5 cm in diameter and could only be diagnosed based on chest imaging.
Stout AF and Murray MR, in 1942, were the first to report the surgical removal of pathologically confirmed hemangiopericytoma, with the original tissue was cells surrounding blood vessels Zimmerman. They described 9 cases with hemangiopericytoma [17] from 1923.

Enzinger FM, Smith BH [7] divide the tumour into two groups, young and old patients. In the young group, the tumour is usually seen in the neck and head, limbs and trunk, is often benign. In the old group, the tumour is usually found in the nasal sinuses and other places. Malignant disease is seen in 1% of haemangioma and 5% of sarcoma [15]. Lung hemangiopericytoma is extremely rare, only 145 cases were reported in the literature from 1954 to 2007 [2,5,7,13,18]. Lung tumours usually present at the age of 50-60, equally in men and women. The most common symptoms are chest pain, dyspnoea, coughing and haemoptysis [5]. Chest X-ray will show the tumour with small pleural effusion. The tumour is a group of cells surrounding the vessels. The cells are small with round nuclear and little cytoplasm. Silver pigmentation showed that the cells are outside of the vessels [12]. According to the pathological studies, the authors classified this type of tumour into group I, II and III based on WHO classification in 2016. Group I can be considered benign as the cells are not changed; Group III includes highly-developed cells (>5 per hpf); Group II has HPC anaplastic criterion that is almost similar to Group III. The mean age of 60 patients was 42.5 years old [5].

According to WHO classification, Sung et al [18] observed that in group I with 42.9 months of follow-up, there was no recurrence as there was no new cells, not many cells, and no abnormal nuclear. Group II had 8 recurrences during treatment (29.6%) and group III also had 6 cases (61.5%) of recurrences but the difference did not reach statistical significance. According to the authors, there were 1 case of death due to distant metastases, 4 cases of brain metastases, 1 intracranial bleeding, 1 tissue bleeding from residual tissue. Therefore, there were 5 cases of distant metastases (other than the brain – 12.5%). Guthrie et al reported that distant metastases are associated with increased mortality rate [4,9]. Moreover, among patients with distant metastases, 3 out of 6 patients died [9]. The earliest metastasis in these studies was from 7 to 20 years. The longest duration until the first metastasis was 182.3 months. Accordingly, group I is considered not metastatic, group III has very rapid metastasis and group II has average risk of metastasis [17].

The precise origin of this type of tumour is unknown, maybe from the undifferentiated mesenchyme cells and is a controversial topic [5,7,11]. Most authors classified hemangiopericytoma in the sarcoma group. But we believed that our patient would be classified as group II as we found the metastasis at the left lung base at approximately 8 years after the first surgery. Regarding chemotherapy, use Panzopanib according to Lee et al (possibly due to its tyrosine, kinase inhibitory properties) [14]. Other authors use Doxorubicin in insolation or combination with Cisplatin, Etoposide, and Gemcitabin [5, 20].
III. CONCLUSIONS

Hemangiopericytoma is difficult to diagnose. We estimated that this patient could be classified as group II. Because group II has slow metastases, the tumour recurred after 10 years. Actually, the patient had suffered from headache, vertigo approximately 2 years ago so we thought the recurrence may have occurred at 8 years after surgery and the metastasis started at the same time. At that time, the tumour was small and diagnosis was difficult due to the lack of suggestive symptoms and lung MSCT. Surgical removal should be performed if possible.

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