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Case report

Thymic carcinoma and superior vena cava syndrome: Case report

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ABSTRACT

Tumor thrombus into the superior vena cava have been rarely reported in cases with mediastinal tumors. These tumors are frequently invasive and continuous from the main tumor that normally shows direct vessel wall invasion, but not in this case. In some cases, the tumor thrombus can be removed through a simple venotomy. To prevent superior vena cava and the left innominate vein stenosis, we used a pericardial patch to close the venotomy site. At this case we report a thymic carcinoma with superior vena cava syndrome, which was caused by a tumor thrombus in the superior vena cava without vessel wall invasion, an even more rare condition.

1. Introduction

Thymic carcinoma is a rare, aggressive neoplasm of the anterior mediastinum that has clinical and biological behavior distinct from thymoma [1]. Patients with thymic carcinoma can be asymptomatic or present with local symptoms as chest pain, cough, and shortness of breath. Once the tumor invades surrounding tissues, more severe symptoms can occur, such as superior vena cava syndrome (SVCS) which is associated with malignancy and therefore presents poor prognosis [2].

Complete resection is the mainstem therapeutic modality for early stage thymic carcinoma. Because of the involvement of surrounding vital structures, the treatment and prognosis of stage III thymic carcinoma is different from stage I and II patients [3].

Among the etiological possibilities of SVCS, it includes intrathoracic malignant tumor, which is responsible for 60%–85% of cases (lung cancer, lymphomas and thymomas). Non-tumor causes represent 15%–40% of cases and SVC thrombosis is on the rise due to the increasing use of intravascular devices. When secondary to thymic carcinoma - due to intraluminal invasion- SVCS is uncommon, and tumor thrombosis without direct invasion is even more rare, as we describe in the present case [4].

2. Case report

A 62-year-old female patient referred to emergency care with a history of progressive dyspnea and increased cervical volume for the last two weeks. Upon admission, physical examination was consistent with SVCS.

Chest CT scan revealed a mediastinal mass with extensive invasion to the SVC (Fig. 1A and B), and positron emission tomography (PET-CT) showed significant mediastinal mass and SVC uptake, but no signs of distance disease (Fig. 1C). Tumor-guided biopsy revealed a squamous cell carcinoma.

The patient underwent a median sternotomy, and the main mediastinal tumor did not show left innominate vein (LIV) invasion, and the superior vena cava syndrome was a result of separate tumor thrombus that was free of vessel wall invasion. The tumor thrombus was removed through LIV and SVC venotomy without cardiopulmonary bypass (Fig. 2A & B). Reconstruction of SVC and LIV to prevent stenosis with autologous pericardium patch was performed through a prolene 4-0 continuous suture (Fig. 2C). Total veins clamping was 40 minutes.

The tumor thrombus was inside left innominate vein and superior vena cava, but there was no vessel wall invasion. The tumor itself did not invaded these structures. The thrombus growth inside the vessels originate from de thymic vein, as shown on final pathology description.

Gross examination of surgical specimen revealed a whitish, poorly delimited tumor mass, 7,0 cm in size, with some small cystic cavities. Histologically, the neoplasia consisted of solid anastomosed blocks of cells with large and vesiculous nuclei, huge eosinophilic nucleoli and relatively scarce basophilic cytoplasm (Fig. 3 A, B, C and D). Mitotic figures and apoptotic bodies were numerous, as were foci of necrosis.

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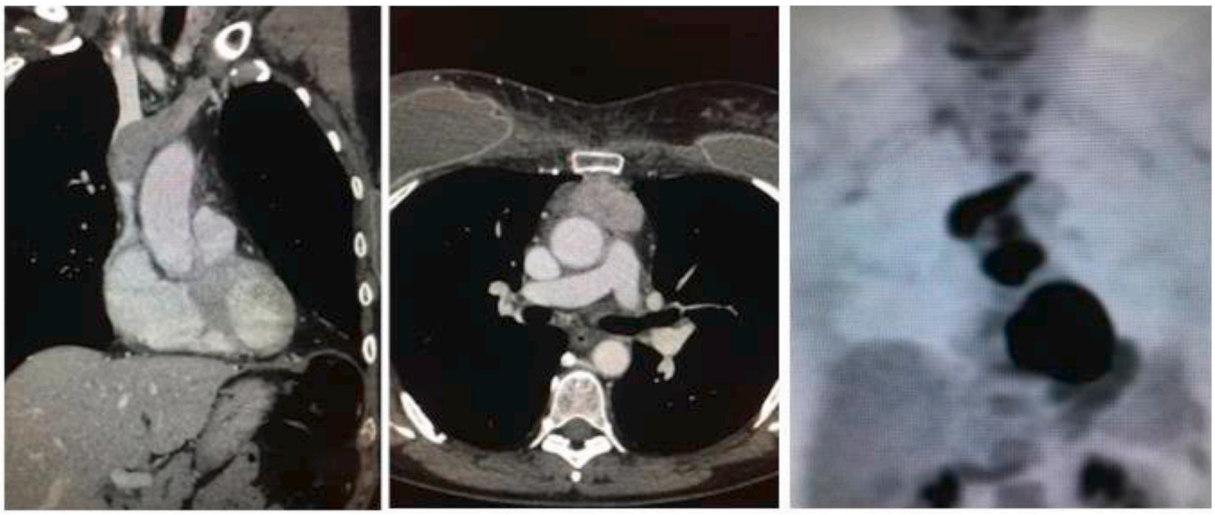

Fig. 1. 1A – tumor thrombus inside SVC; 1B – anterior mediastinal tumor; 1C –mediastinal mass and tumor thrombus uptake.

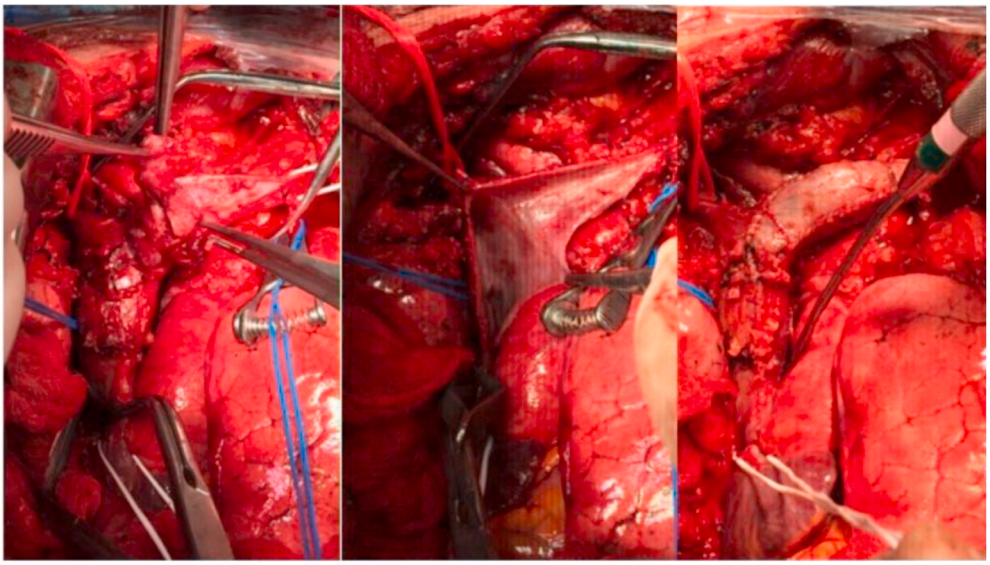

Fig. 2. 2A. tumor thrombus inside SVC and LIV; 2B: SVC without tumor thrombus; SVC and LIV reconstruction.

The neoplasia infiltrated the perithymic fat and we could rapidly identify invasion of thymic veins. Residual thymic parenchyma could be easily demonstrated. Surgical margins were close (less than 1 mm on various sites) but not directly involved.

After 4 weeks, the patient was referred to adjuvant 54 Gy radiotherapy, with low side effects. After five years of follow up, there is no evidence of disease recurrence.

3. Discussion

Thymic tumors as the cause of SVCS is unusual and external compression is the cause, but direct tumor invasion into the SVC is far uncommon [5]. Thymic tumor thrombus normally invades SVC through thymic veins.

According to the 2015 World Health Organization classification of tumors, the term "thymic carcinoma" can be classified into 13 distinct histological subtypes [6]. Squamous cell carcinoma is the most common subtype comprising more than 70% of all cases. There are two hallmarks for the diagnosis of thymic squamous cell carcinoma: the clear-cut cytological atypia in the large epithelial cells that are arranged in nests and cords, and the broad zone of fibrohyaline-stroma separating the tumor cell nests. The squamous cell carcinoma has an aggressive clinical course, tends to invade surrounding tissues, and metastasizes at an early stage, but they have a better prognosis than other types (with the exception of basaloid carcinoma) [6,7].

The patient is classified as T3N0M0 and prognostic stage group IIIA according to the American Joint Committee On Cancer (AJCC), International Thymic Malignancies Interest Group (ITMIG) and the International Association for the Study of Lung Cancer (IASLC) [8,9].

Complete resection is the mainstem therapeutic modality for early stage thymic carcinoma patients. With an R0 resection, surgery has a 10-year survival of 80%, 78%, 75%, and 42% for stages I, II, III and IV, respectively [10]. Generally, the disease-free survival (DFS) is greater in series of cases in which the majority of the patients had well differentiated and localized tumors than in differentiated tumors which are related to lower survival even after radical treatment [4,5]. Even though studies have showed that invasion of SVC it's a negative prognosticator in T3N0M0 undergoing complete resection, the patient had no evidence of recurrency of thymic carcinoma in the past 5 years [11]. Nevertheless, since the thrombus is intraluminal and not in the wall, this thymoma patient has a risk of its thrombus shedding off during operation and cause pulmonary embolism, which is an event frequently described in

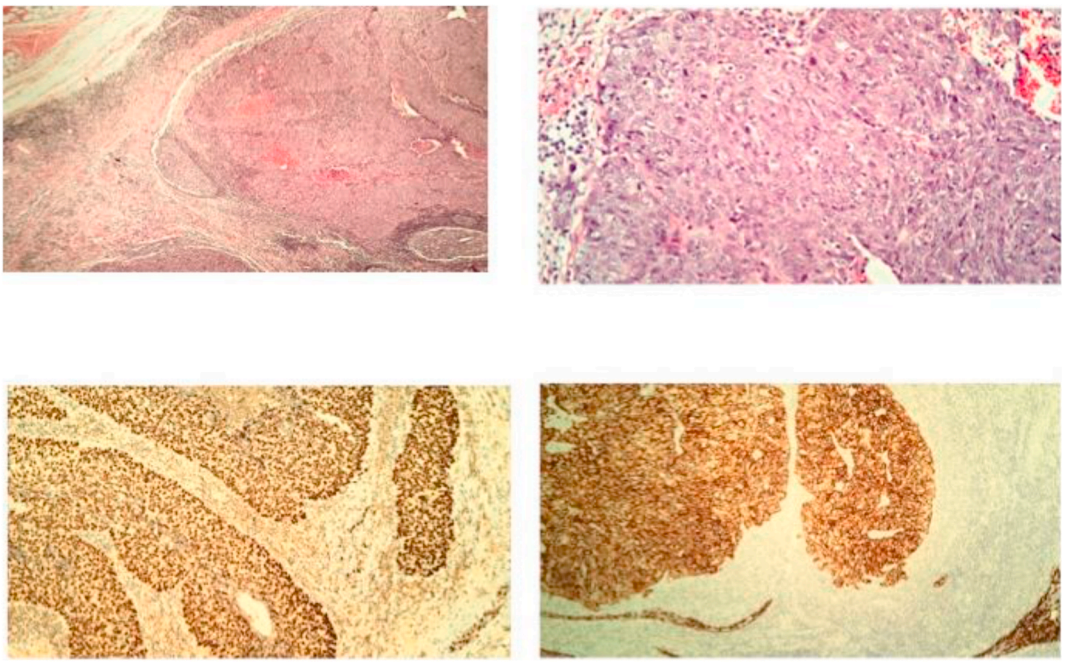

Fig. 3. Well differentiated squamous cell carcinoma of the Thymus, showing eosinophilic cytoplasm, atypical mitoses and focal keratinization.

literature. In this case a pulmonary artery exploration would be necessary or even a pulmonary angiography during operation should be performed [12].

4. Conclusion

The case reported suggests a more aggressive approach seeking complete resection. After performed vein clamping, without cardiopulmonary bypass, further treatment with radiation therapy was done to recurrence risk reduction. The case is considered successful because, pathologically, complete resection was achieved and there was no evidence of disease recurrence in 5 years.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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