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Case Report

De Novo Angiosarcoma of the Thoracic Outlet: A Rare Entity

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ABSTRACT

Angiosarcoma is a rare soft tissue highly malignant tumor of vascular origin, accounting for only 1% to 2% of these tumors. We present a rare case of De novo (unrelated to irradiation or pyothorax) angiosarcoma of the thoracic outlet with a 10-year disease free survival. A 49-year-old male was admitted to our department due to a tumor of the thoracic outlet referred by the neurologists to whom he addressed complaining for vertigo and instability. After a thorough examination including chest X-ray, CT scan and MRI, a well circumscribed vascularized lesion of a diameter of 5cm was detected in the thoracic outlet. A head and neck angiography was performed along with a full staging in order to exclude metastatic disease. The patient was submitted to high axillary thoracotomy. The adjacent structures were dissected, small arterioles arising from the subclavian artery, neo-vessels were ligated or cauterized and the soft largely encapsulated tumor was excised. Gross observation of the resected specimen demonstrated a regular-shaped neoplasm. Histology revealed a well differentiated angiosarcoma comprised of multiple anastomosing blood vessels lined by endothelial cells showing malignant features but with little nuclear pleomorphism. The recovery was uneventful. The patient did not receive postoperative radiation or chemotherapy and 10 years postoperatively is free of disease. In conclusion, de novo primary pleural angiosarcoma are rare entities that should not be misdiagnosed. The immunohistochemical examination is the key for diagnosis and will offer the definite histotype. Although the prognosis is bad, early detection will give the patient the best chance for successful surgical treatment. Our patient being well and asymptomatic after 10 years follow-up represents a rare case and the first mentioned in the literature with a location of the lesion in the thoracic outlet.

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Introduction

Angiosarcoma is a rare soft tissue highly malignant tumor of vascular origin, accounting for only 1% to 2% of these tumors [1]. It may affect every organ but most commonly occurs in the skin and soft tissues, liver, spleen, heart and breast [1, 2]. Primary angiosarcomas of the chest wall and pleura are extremely rare and usually have a dismal prognosis. The etiologic factors include trauma, radiation, foreign bodies, thorium

dioxide, viral infections. We present a rare case of De novo (unrelated to irradiation or pyothorax) angiosarcoma of the thoracic outlet with a 10-year disease free survival.

Case Report

A 49-year-old male was admitted to our department due to a tumor of the thoracic outlet referred by the neurologists to whom he addressed complaining for vertigo and instability. After a thorough examination

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including chest X-ray, CT scan and MRI, a well heterogeneous contrast-enhanced lobulated vascularized mass of a diameter of 5cm was detected in the thoracic outlet (Figures 1 & 2). A head and neck angiography along with a full staging took place to exclude metastatic disease. Positron emission tomography (PET) only showed a large lesion occupying the thoracic outlet of the left pleural cavity, with no 18F-fluorodeoxyglucose (FDG) uptake. No abnormalities were observed during bronchoscopic examination. Fine-needle aspiration biopsy (FNAB) was not performed due to the location of our tumor and the risk of possible dissemination of cancer cells.

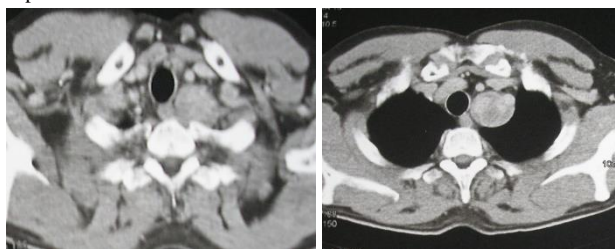


Figure 1: Chest CT scan revealing a paratracheal tumor in the thoracic outlet.

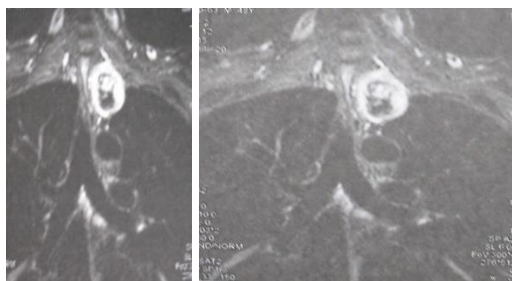


Figure 2: Magnetic Resonance Imaging shows the relation of different structures to the lesion.



Figure 3: Specimen showing a well circumscribed, encapsulated lesion.

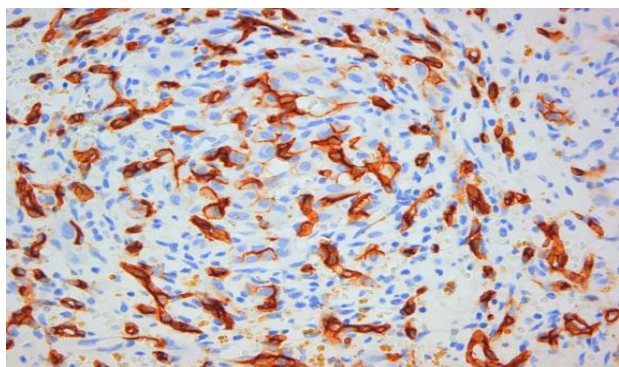


Figure 4: Histology revealed a well differentiated angiosarcoma comprised of multiple anastomosing blood vessels lined by endothelial cells showing malignant features but with little nuclear pleomorphism. CD34 immunopositivity in epithelioid cells.

The patient was led to surgery in order to get both, diagnosis and definite treatment. He was submitted to high axillary thoracotomy under general anaesthesia with a double lumen tube. The mass adhered to the mediastinal pleura and to the parietal pleura of the thoracic outlet close to the vessels. The lung was not found to be invaded. The adjacent structures were dissected with difficulty, small arterioles arising from the subclavian artery and neo-vessels were ligated or coagulated by using a bipolar diathermy and the soft largely encapsulated tumor was excised. No axillary or mediastinal lymphadenopathy was identified. Gross observation of the resected specimen demonstrated a regular-shaped neoplasm (Figure 3). Histology revealed a well differentiated angiosarcoma comprised of multiple anastomosing blood vessels lined by endothelial cells showing malignant features but with little nuclear pleomorphism (Figure 4). The recovery was uneventful. The patient did not receive postoperative radiation or chemotherapy and 10 years postoperatively is free of disease.

Discussion

Angiosarcomas are soft tissue tumors arising from endothelial cells of the small vessels [1]. The term angiosarcoma is now used for malignant tumors, and the term hemangioendothelioma is applied for borderline tumors [2, 3]. Although the lung is a common site of metastasis in the natural history of angiosarcoma, primary pleuropulmonary angiosarcoma is a rare entity, with only few cases reported in the medical literature [2, 4]. The etiology is uncertain, although there are reports associating the lesion with a history of chronic tuberculous pyothorax, and exposure to radiation and asbestos [2, 4-6]. When unrelated to association factors, the case is described as *de novo* tumor [2, 4].

Usually the symptoms are nonspecific and include chest pain, dyspnea, hemoptysis, pericarditis, pleural effusion and massive recurrent hemothorax with anemia [2, 4, 7]. Diagnosis may be difficult because chest radiographs computed tomographic scans are nonspecific, showing masses, thickening of the pleura and diffuse opacity in advanced lesions being similar to those of mesothelioma or advanced pulmonary carcinoma [8, 9]. A positron emission tomography scan can only help in determining the extent of the disease, because the diagnosis is established only by biopsy [5]. Fine-needle biopsy (FNB) might be helpful, especially in non-surgical cases [10]. Although the histopathological features of angiosarcoma have been described by Stout in the 1940s, the definitive diagnosis and definition of its primary or secondary origin may be difficult [11]. Ancillary methods are needed because of the diverse cytomorphic features that can overlap with those of other spindle cell and mesenchymal neoplasms, as well as those of carcinomas and post-irradiation atypia [5, 6].

Angiosarcoma primarily comprised of large and round “epithelioid” endothelial cells with abundant cytoplasm and large vacuolated nuclei. Positive expression of at least one endothelial marker (including CD31, CD34, and factor VIII) is required to confirm the diagnosis of angiosarcoma, though CD31 is considered the most specific and most sensitive marker [2, 4, 12-15]. Folpe *et al.* reported that FLI-1 was expressed by 50 of 53 vascular tumors (94%), including 20 of 22 angiosarcoma cases [16]. These results suggest a role for FLI-1 as a novel nuclear marker of vascular tumors. Angiosarcomas may also express epithelial markers, such as CK, especially epithelioid variants and can be easily misdiagnosed as malignant mesotheliomas [9].

Metastatic patterns of primary pleural angiosarcomas are not well documented. Pulmonary metastasis from cutaneous and cardiac angiosarcomas occurs in 60%-80% of cases [17]. Primary pleural angiosarcomas have been shown to present in most segments of the pleura with invasion commonly to the lung parenchyma, blood vessels, pericardium, diaphragm, and bronchial wall [2, 17-20]. Prior reports of patients with primary pleural angiosarcomas showed limited established patterns of metastasis, but there are some authors that presented cases with metastatic locations, including brain, skin, oral mucosa, and lung [6, 17].

Management modalities of angiosarcoma include surgery, radiotherapy, and chemotherapy [8, 21-23]. Surgery must be performed, whenever possible, and is indicated in cases where a radical excision is possible. Different surgical treatments are mentioned in the literature including complete surgical resection, debulking with pneumonectomy and rib excision, as well as cauterization of bleeding points [2]. Radiotherapy may have a beneficial role in postoperative settings in the absence of diffuse or metastatic disease [4]. Chemotherapy generally, has minimal effect and is merely utilized for purposes of symptom palliation, and is rarely used in patients with poor performance status [2]. Preoperative vascular embolization can be used to reduce tumor vascularity and accordingly intraoperative pleural bleeding and might also decrease the size of the tumor [2, 8]. Chemotherapy has little effect and is expected only to palliate the symptoms, whereas radiotherapy may play a role as an adjuvant therapy, except for cases of diffuse or metastatic disease.

Prognosis is generally extremely poor and death often occurs soon within 24 months from the time of diagnosis [9]. In conclusion, De-novo primary pleural angiosarcomas are very rare tumors and should not be misdiagnosed based on their rarity. The immunohistochemical examination is the key for definite diagnosis and will offer the definite histotype. Although the prognosis is bad, early detection will give the patient the best chance for successful therapy.

Our patient being well and asymptomatic after 10 years follow-up represents a rare case and the first mentioned in the literature being located in the thoracic outlet.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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