A Giant Cardiac Malignant Peripheral Nerve Sheath Tumor Presenting With Total Obstruction of the Superior Vena Cava

Edvin Prifti, MD, PhD, Arben Baboci, MD, and Majlinda Ikonomi, MD

Division of Cardiac Surgery, University Hospital Center of Tirana, Mother Teresa, Tirana, Albania

A 16-year-old boy presenting with dyspnea, facial swelling, cyanosis, and fatigue was found to have a tumor involving the heart, causing superior vena cava and brachiocephalic venous trunk total obstruction. This was diagnosed as malignant peripheral nerve sheath tumor, a rare sarcoma of the heart. The patient underwent successful resection of the tumor, and reconstruction of the superior vena cava and right atrium. Immunohistochemistry was utilized to establish the diagnosis. The details of the patient’s clinical course and imaging findings with morphologic and immunohistochemistry data are reported.

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Primary neurogenic neoplasms of the heart are very rare. Neurogenic malignant tumors are composed of cells that constitute the peripheral nerve sheath (Schwann cells, perineural cells, and so forth) and account for 5% to 10% of all malignant soft tissue tumors. Neurogenic malignant tumors are classified into malignant schwannoma, neurofibrosarcoma, neurogenic sarcoma, and so forth, and are included under the entity of malignant peripheral nerve sheath tumor (MPNST). Here we present a patient with superior vena cava (SVC) and brachiocephalic venous trunk (BVT) obstruction due to a giant MPNST, who underwent a successful complete surgical resection and SVC and right atrium (RA) reconstruction.

A 16-year-old boy, with a 1-month history of dyspnea, facial swelling, cyanosis, and fatigue was admitted to our hospital. On clinical examination, he had a respiratory rate of 24 breaths per minute, oxygen saturation of 92% while breathing room air, and engorged jugular venous system. His face, arms, and upper chest were edematous and cyanosed. Transthoracic echocardiography revealed a giant mass on the RA. Contrast-enhanced computed tomography revealed a large atrial mass migrating into the right ventricle through the tricuspid valve, extending and obstructing totally the SVC and BVT (Fig 1A).

The patient was referred for surgical treatment. The ascending aorta and inferior vena cava were cannulated. Cardiopulmonary bypass was instituted, and the patient was cooled to 24°C. The RA was opened and a giant mass was identified (Fig 2A). The tumor was resected on block from the atrial wall. After short periods of intermittent circulatory arrest, the SVC and BVT were opened and the tumor was removed totally. The patient had an uneventful postoperative course. He was referred to the oncology service, where he was treated with seven cycles of adjuvant chemotherapy with ifosfamide and doxorubicin and local radiotherapy [1, 2].

The preliminary histologic evaluation revealed clear margins of a lobulated mass with brown areas and soft zones with a myxoid-like consistency. Histologically, the tumor presented a fascicular spindle cell architecture, in some parts organized in a herringbone pattern and in others in a more storiform one with myxoid and necrosis areas and mitotic activity (Fig 2B). Another feature was the presence of numerous vascular channels in the myxoid stroma that were surrounded by hypercellular tumor cell aggregates that blended with the vascular structures. The tissue was examined immunohistochemically with the automated BenchMark ULTRA system (Ventana Medical Systems, Tucson, AZ). The immunohistochemical examination showed intense expression for Vim (Fig 2D) and a Ki67 proliferative index of 30%. The SMA, CD34, CD68, Bcl2 were negative whereas there was a focal positivity for S100, a more expressed positivity for CD56 (Fig 2C), and focal positivity for CD57—making possible the diagnosis of MPNST.

Three months later the patient underwent contrast-enhanced computed tomography angiography, which did not reveal the presence of any intracardiac tumoral mass (Fig 1B). At 1 year after the surgical correction, the patient is alive and without tumoral recurrence.

Comment

Malignant peripheral nerve sheath tumor is defined as malignant tumor arising from peripheral nerves or continuous to nerves, or as benign neurogenic tumors converted into malignant tumors, or malignant tumors developing in neurofibromatosis [1, 2]. Although MPNST primarily affects extremities, it can develop in various other sites including, rarely, the thoracic cavity. The majority of MPNST occur on the right side of the heart, in close proximity to the interatrial septum, probably because of the origin of MPNST from cardiac branches of the vagus nerve and cardiac plexus [1, 2]. However, the presence of MPNST on the left side has been reported [3]. Primary sarcomas arising in veins are extremely rare [4]. Rytina and coworkers [5] have reported the unique case of a sarcoma originating from the BVT. Other reports demonstrated that an intracardiac primary sarcoma might extend retrogradely into the SVC, causing obstruction [6]. It is possible that the MPNST in our patient originated from the BVT, extending to the SVC and RA until causing total obstruction. The other option is that the tumor has been extended retrogradely to the SVC. This remains the first case with MPNST causing...
SVC obstruction undergoing successful surgical correction.

The most common neoplastic causes of SVC obstruction are non-small cell lung cancer, small cell lung cancer, lymphoma, and metastasis. Primary cardiac tumors are an extremely rare cause of SVC obstruction [4-6]. Symptoms do not develop until the tumor interferes with cardiac function. Dyspnea, fatigue, cough, chest pain, anorexia, weight loss, neck edema, arrhythmia, and peripheral embolism have been reported [1, 2].

Histologically, MPNST has a variety of appearances; the majority show fascicular spindle cell architecture, associated with myxoid areas and perivascular whirling. Necrosis and mitotic activity are common, reflecting the high-grade nature of these tumors. The MPNST may be indistinguishable from other sarcomas on routine staining, and immunocytochemistry is important for diagnosis. In our case based only on hematoxylin and eosin staining, it was given a preliminary diagnosis of fibromyxosarcoma. Approximately 50% to 90% of MPNST are positive for S100. In high-grade MPNST, only scattered tumor cells are S-100 positive. This is the same feature as in our case. Other markers such as CD56, CD57, protein gene product, and glial fibrillary acid protein are useful as second-line markers [7]. In our case, the immunohistochemical examination showed intense expression for Vim, focal positivity for S100, a more expressed positivity for CD56, and focal positivity for CD57.

Thus far, the best modality for MPNST treatment is still complete surgical resection especially when the SVC obstruction occurs secondary to a primary cardiac tumor. Surgery may also be offered for tissue diagnosis and palliation of symptoms. In addition to a complete surgical resection, adjuvant therapy is usually advocated, as in our case [1, 2].
In conclusion, this is the first reported case of a patient with giant MPNST causing total obstruction of the SVC and BVT who underwent successful surgical correction. Full histologic examination is required to confirm diagnosis. Complete resection associated with adjuvant chemotherapy and radiotherapy seems to offer an excellent midterm outcome.

References


