



PALLIATIVE CARE CASE OF THE MONTH

“Malignancy related Superior Vena Cava Syndrome”

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Case: RC was a 60-year-old woman with widely metastatic breast cancer, who had progressed on multiple lines of chemotherapy. Her goal was to continue therapy for as long as possible in the hopes of attaining as much time with her family as she was able. This included full resuscitative measures if her condition declined. In the summer, she presented with worsening of upper extremity edema, dyspnea and dysphagia. A contrast-enhanced chest CT demonstrated progression of cancer in the right chest, mediastinum and lymph nodes with severe narrowing of a segment of the superior vena cava (SVC). Based on her imaging and symptomology, she was diagnosed with SVC syndrome. She was admitted to the general medicine service, and consultations were obtained from radiation oncology and vascular surgery. The surgical service contemplated SVC angioplasty but deferred, as she was medically stable at that juncture. A plan for palliative radiation was formulated but proved challenging to implement, as she could not comfortably lie flat for the prescribed treatments. As a result, radiation was delayed for five days, during which time her condition worsened and she was admitted to the intensive care unit for mechanical ventilator support.

Discussion: Superior Vena Cava (SVC) syndrome can result from any process that obstructs blood flow in the SVC. In the post antibiotic era, up to 90% of cases of SVC syndrome have been the result of invasion or external compression of the SVC by a malignant process arising from the right lung, lymph nodes or mediastinal structures. More recently, with increased use of intravascular devices, such as central vein catheters, the incidence of thrombosis has risen, and benign causes may now account for 20-40% of cases. This discussion will focus on the management of malignant causes of SCV syndrome.

Elevation of central venous pressures results in interstitial edema of the head and neck. This edema may narrow nasal passages, and cause swelling of the larynx and pharynx resulting in dyspnea, stridor, cough, hoarseness and dysphagia. Cerebral edema may also occur and can result in cerebral ischemia, herniation and death. If cerebral edema is present, patients may complain of headache, confusion or present with coma. The most common clinical findings are facial edema and distension of veins in the head and neck. The rapidity and severity of symptoms depends on the rate of obstruction of the SVC in relation to the recruitment of venous collaterals as alternate pathways for return of blood to the right atrium. As venous collaterals dilate over several weeks, upper body venous pressures decrease. A rapidly evolving malignant obstruction may not allow adequate time for development of collateral flow. On the other hand, a slowly evolving malignant process allows for extensive collateralization and thus presents in a more insidious manner. In general, the signs and symptoms of SCV syndrome gradually progress over several weeks and then improve over time with the development of venous collaterals.

Of malignant causes NSCLC accounts for 50% of cases followed by small cell lung in 25% and non-Hodgkin's lymphoma in 10% of cases. For unclear reasons, Hodgkin's Lymphoma rarely causes SVC syndrome despite its common presentation in the mediastinum. Clinical history together with CT imaging will be able to differentiate between benign and malignant etiologies of SCV syndrome in most cases. Histologic diagnosis and severity of symptoms guide appropriate therapy.

In the past, SVC syndrome was treated as a potentially life threatening medical emergency and managed with immediate radiation treatment. More recently, a clearer understanding of the pathophysiology and natural history of SVC syndrome has shown that it evolves over weeks to months. Deferral of therapy does not pose a hazard to the medically stable patient. In fact, radiation therapy prior to biopsy can impair accurate histologic diagnosis and thus compromise medical care.

Patients who present with severe laryngeal edema resulting in respiratory compromise or cerebral edema resulting in signs of a depressed central nervous system do represent true medical emergencies. These patients require immediate medical treatment with endovascular stent placement and radiation therapy. Endovascular stenting provides more rapid relief than can be achieved with radiation treatment. In malignancy related SVC syndrome, endovascular stents resulted in symptomatic improvement in 90% of cases, with re-occlusion occurring on average in 13% of cases. Thrombotic occlusion of the SVC is not a contraindication for endovascular stent placement.

Other supportive interventions include raising the head to decrease hydrostatic pressure, glucocorticoids, and diuretics. For medically stable patients with small cell lung cancer, non-Hodgkin's lymphoma and germ cell tumors, chemotherapy may be the treatment of choice and can achieve long-term remission. Due to considerable associated morbidity, surgery should be reserved for carefully selected cases when severe symptoms persist despite the aforementioned treatment options.

The average prognosis of patients with malignancy related SVC syndrome is six months, but varies widely depending on the underlying malignancy. Treatment of the SVC syndrome and the underlying cancer can result in cure or long-term remission in some patients.

Resolution of Case: RC ultimately completed all 12 planned radiation treatments while on mechanical ventilation. Repeat imaging towards the end of her radiation therapy showed improvement in her mediastinal and para-tracheal disease. Her hospital stay was one month in duration; she successfully weaned from the ventilator and discharged from the hospital on hospice care.

Personal details in the case published have been altered to protect patient privacy.

For palliative care consultations please contact the Supportive and Palliative Care programs at PUH/MUH, 647-7243, pager # 8511, Shadyside, 647-7243, pager # 8513, Perioperative/Trauma Pain, 647-7243, pager # 7246, UPCI Cancer Pain Service, pager 644-1724, Interventional Pain 784-4000, Magee Women's Hospital, pager 412-647-7243 pager # 8510, VA Palliative Care Program, 688-6178, pager # 296. Hillman Outpatient: 412-692-4724. For ethics consultations at UPMC Presbyterian-Montefiore and Children's pager 958-3844.

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Resolution of Case continued

She initially went to an inpatient hospice unit for ongoing symptom management, and from there she was able to return home with hospice support. She died two months later.

Review of this case highlights a missed opportunity for multidisciplinary care. As RC's respiratory failure evolved, consideration was not given to endovascular stenting and interventional radiology was not consulted. Perhaps endovascular stenting would have resulted in quicker control of RC's symptoms, avoided mechanical ventilation and a protracted hospital admission. Perhaps RC's goal of spending more time with her family could have been more completely fulfilled.

References:

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