Case Report

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A Case of Superior Vena Cava Syndrome in Pregnancy Within a Developing Country in Sub-Saharan Africa

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Abstract

Introduction: Superior Vena Cava Syndrome is a rare but potentially life-threatening condition that can occur during pregnancy. The case presented here describes a dramatic presentation culminating in a potentially preventable maternal death at a tertiary hospital in East Africa.

Case presentation: A HIV positive, 28 years old primigravida at a gestational age of 34 weeks presented to a tertiary health facility in Northern Tanzania with generalised facial swelling and difficulty in breathing. She was referred from primary health facility where she was treated presumptively as a case of Ludwig's angina. On examination she was dyspnoeic and had extensive supraclavicular and cervical lymphadenopathy. On second day post-admission, she was induced, but later her condition deteriorated and developed severe dyspnoeic, confusion, convulsions and died. A Narrative of this case, challenges encountered in investigating, establishes a diagnosis and management are explored in this case report. Furthermore pathophysiology and treatment options for superior vena caval occurring in pregnancy will be reviewed.

Conclusion: Superior vena syndrome is a fatal condition and rare to occur in pregnancy. A rigorous and timely approach to investigate and manage is of a critical importance. Additionally a collaborative multidisciplinary team approach is necessary to prevent complication.

Keywords: Superior Cava Syndrome, Pregnancy, East Africa, Case report

Introduction

Superior Vena Cava Syndrome is often dramatic in its presentation and rarely seen in pregnancy. Prompt recognition, diagnosis, and treatment can result in improved outcomes for mother and fetus. In developing countries with low resources and limited access to medical care present additional challenges to providing care for these patients.

Superior vena cava syndrome occurs in about 15,000 people in the United States each year; the incidence in Sub-Saharan Africa is unknown. It was first described

in the mid 1700s in a patient with a syphilitic aortic aneurysm, indeed early reports showed most of the cases were related to an infectious process, most commonly syphilis or tuberculosis. Today the etiology tends to be more commonly related to a neoplastic process mostly such as lung cancer, particularly adenocarcinoma or lymphoma involving the mediastinum [1].

Superior vena cava is located on the right side of the mid mediastinum and extends about 6-8 cm cephalad from the right atrium. It is easily compressible by an extrinsic mass given its location. The prevalence of Superior Vena Cava Syndrome in patients with either lung cancer or

lymphoma is between 2-5%. In addition this syndrome may be seen secondary to venous thromboembolism associated with a venous catheter or in cases of vasculitis involving the vena cava [3]. Overall survival is determined by the underlying cause as the symptoms of SVCS respond well to prompt recognition and treatment.

Case Presentation

A 28 years primigravida at 34 weeks of gestation by dates and 30 weeks by mid-trimester ultrasound was referred to Kilimanjaro Christian Medical Center (KCMC) in Moshi, Tanzania from a district hospital located 70 km away. She was a member of the Maasai, a semi-nomadic tribe living in northern Tanzania. She presented with worsening facial swelling at 24 weeks of gestation. At the district hospital, a provisional diagnosis of Ludwig's Angina with a differential diagnosis of non-Hodgkin's Lymphoma and Tuberculous lymphadenitis was entertained. She was treated presumptively with Ceftriaxone and Gentamycin.

She was diagnosed with HIV 4 years prior to admission, currently with no AIDS defining illness and had been treated with antiretroviral therapy for 2 years. Her recent CD-4 count was 693 cells/mm³.

She had history of facial swelling which had rapidly increased, accompanied with mouth and facial pain, with speaking and swallowing difficulty, accompanied with drooling and orthopnea.

Physical examination revealed an alert, ill appearing woman. Her blood pressure was 130/70 mmHg, pulse rate 90 bpm, respirations 18 breath per minute. The head, face, and neck were massively swollen with difficulty in opening and closing her eyes and mouth. She had hard fixed cervical lymphadenopathy (Figure 1a). Chest showed fine crackles in both bases and heart sounds were normal. Abdomen was soft and non-tender with distended veins on the surface (Figure 1b). Fundal height was 30 cm and fetal heart tones were 142 beats per minute.

A biophysical profile was 8 out of 8 with 30 weeks gestation by ultrasound. She had mild anemia with hemoglobin of 9.2 g/dl, creatinine was 0.9 mg/dl, and liver functions were normal, except an elevation in aspartate aminotransferase (AST) of 40 IU/L. Antibiotics were changed to Ceftriaxone and Metronidazole, and Dexamethasone was started for fetal lung maturation due imminent need for premature delivery for maternal indications.

She was admitted to the antepartum ward for observation with the provisional diagnosis of superior vena cava syndrome secondary to either Non Hodgkin Lymphoma or TB adenitis (scrofula), or Ludwig's Angina.

Most likely primary diagnosis was lymphoma with HIV due to the absence of other symptomatology or radiographic evidence of Tuberculosis. The skull x-ray was normal while the chest x-ray showed lymphadenopathy of the mediastinum (Figure 2).

Efforts to get a definitive histological tissue diagnosis and transfer her to the closest center for external beam radiation and chemotherapy depending on the histologic diagnosis were made. This nearest center is approximately 6 hours away by bus and at a significant cost to the patient and her family.

Over the next 48 hours, she was confused although maintained normal vital signs. It was felt that her deterioration was related to cerebral edema and the decision to induce her was made with the hopes of improving her status. On the 4th hospital day within a few hours of her induction, her mental status deteriorated.

Admission to the intensive care unit was warranted but there were no beds and all ventilators were in use. Induction was continued as it was felt that she would not survive general or spinal anesthesia or Cesarean section. On the fifth hospital day, she developed sudden oxygen de-saturation associated with generalized seizure. She was treated with anticonvulsants and oxygen but suffered a cardiac arrest. Cardiopulmonary resuscitation was attempted but unfortunately she died 8 hours after induction. No perimortem C-section was attempted and the fetus was also lost. An autopsy was not performed at the request of her family, but the cause of death was felt to be secondary to acute increased intracranial pressure with herniation. The underlying pathologic process was never confirmed.

Discussion

This case exemplifies the challenges of superior vena cava syndrome during pregnancy complicated by the lack of resources in much of the world. Superior vena cava syndrome is a rare event that occurs in about 15,000 people in United States yearly but the incidence in Sub-Saharan Africa is unknown. William Hunter originally described SVCS in 1757 in a patient with a syphilitic aortic aneurysm [2] and subsequent reports showed most of these cases were related to an infectious process, most

Figure 1a: Enlarged Right Supraclavicular Lymph Node

Figure 1b: Distended veins on the chest

Figure 2: Widened upper mediastinum with lymphadenopathy

commonly Syphilis or Tuberculosis. Today the etiology commonly relates to neoplastic processes mostly such as lung cancer -particularly adenocarcinoma or lymphoma involving the mediastinum [1].

The superior vena cava provides the main drainage of venous blood from the head, neck, and upper thorax. It is located on the right side of the mid mediastinum and extends about 6-8 cm cephalad arising from the right atrium. It is easily compressible by an extrinsic mass given its location with a prevalence of superior vena cava syndrome in patients with either lung cancer or

lymphoma between 2-5%. In addition this syndrome may occur secondary to venous thromboembolism associated with a venous catheter or rare cases of vasculitis involving the vena cava [3].

In pregnancy, additional planning is required in terms of monitoring the mother and her fetus and plan for time and route of delivery. The most useful imaging is a CT of the head, neck, and chest after contrast, which will differentiate between extrinsic and intrinsic (thrombosis) causes of obstruction. Biopsy either of a supraclavicular lymph node or via bronchoscopy prior to invasive

procedures such as mediastinoscopy is often preferred.

The severity depends on both the speed of onset as well as the degree of narrowing of the vascular lumen. When blood return from the superior vena cava is obstructed it flows back to the heart via a vascular network to the inferior vena cava. The most striking feature is that of edema of the head and face often with identification of collateral vessels in the chest and abdominal wall. The diagnosis is mainly clinical, based on signs and symptoms as noted with special attention to the rate of progression as well as other symptomatology relating to the underlying disease [4-6].

Treatment should address both the underlying disease process as well as the obstruction causing the superior vena cava syndrome. Elevation of the patient's head and prompt tissue diagnosis with initiation of systemic chemotherapy is the mainstay of treatment. Corticosteroids, diuretics, and external beam radiation may promote relief of symptoms but without proven long-term benefit.

In addition vascular stent placements by Interventional Radiology or surgical bypass have been suggested in severe cases for relief of life threatening symptoms. In this case pregnancy does not confuse the clinical presentation, diagnosis, or treatment options though certainly poses added risks to both mother and child.

In pregnancy cerebral blood flow in the larger arteries gradually decreases throughout gestation associated with a decrease in arteriolar resistance to maintain constant blood flow at the level of the tissue [7]. In the case of diminished outflow such as with superior vena cava syndrome could theoretically affect auto-regulation with changes in the cerebral perfusion and intracranial pressure with increased risk of acute cerebral edema. In addition, there is the potential for the common occurrence of supine hypotension related to maternal positioning and the weight of the gravid uterus to have a more effect on diminishing cerebral blood flow due to decreased cardiac output.

Respiratory status can also be compromised either due to extrinsic compression of the underlying disease process, and must be followed closely looking for signs of acute decomposition.

Finally, this patient represents challenges in the practice of high-risk obstetrics in a resource-limited setting where

access to rapid diagnostic testing and therapeutic options are limited. The assumption was that the underlying etiology of SVCS was lymphoma associated with HIV. This was based on the fact that she was at low risk of lung cancer and had no other symptomatology of an infectious etiology like Tuberculosis or Syphilis. Imaging studies were limited to an obstetrical ultrasound, chest and skull x-rays. At the time of this patient's admission there was no pathologist on site to interpret results of any tissue obtained and had there been a diagnosis there would have been no ability to administer advanced chemotherapy, external beam radiation therapy, or vascular stent placement.

The decision to attempt vaginal delivery in this patient as opposed to planned C-section was a difficult one but made in conjunction with the departments of Medicine and Anesthesiology at KCMC. In review of the anaesthesia literature there was a consensus that avoidance of general anesthesia is imperative as it is associated with a high morbidity and mortality related to problems with airway edema, existing respiratory compromise, and positive pressure -ventilation that could further decrease venous return from the head.

In addition the most commonly used anaesthetic for C-section at KCMC is a single bolus subarachnoid block, which in this case could have resulted in a rapid and unpredictable hypotension. The safest method would be to proceed with continuous lumbar epidural block initiated with slow incremental doses of local anesthesia [8,9], again, which was not available. Finally, whether or not to pursue perimortem C-section at the time of cardiac arrest in the hopes of enhancing survival for the mother as well as fetus [9]. Given existing resources and training it is doubtful that this would have changed the outcome of this case. That said, it should certainly be a part of clinical decision making in cases where a pregnant woman suffers a cardiac arrest

Conclusion

This clinical presentation should prompt the clinician to establish a rapid diagnosis in order to provide effective treatment of the SVC syndrome and the underlying disease process, which in this case may have been lifesaving for both mother and child. It is important to be aware of this rare but potentially lethal syndrome that can be seen in pregnancy especially within areas with a higher prevalence of tuberculosis and HIV disease.

Conclusively, this case highlight the importance

of prompt recognition and establishment of primary diagnosis and institute a timely management when such critical condition is suspected. Additionally a multidisciplinary approach to these patient in planning for delivery is crucial.

Consent

Since the subject was an illiterate, verbal consent was sought instead of a standard written consent. A standardised checklist on ethical approval from KCMC ethical committee was used mainly provided detailed information on the main purpose of obtaining the picture and use of information for potential publication purposes to further understand the condition was narrated. Also assurance that her de-identified portrait would be used in the document was provided.

Declaration

Ethics approval and consent to participate

Verbal informed consent was approved from the patient for the usage of de-identified documents for potential publication purposes.

Authors contributions

BM managed the case, and has provided significant contribution in the manuscript writing. PM has provided significant contribution in the development of manuscript. JS has provided significant contribution in literature search and managed the patient.

Competing of interest

The authors declares to have no competing interest in writing this article

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Consent to Publish

Verbal consent was obtained from the subject prior to obtaining picture and other detailed information.

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