

## **Anesthetic Management for a Patient with Kidney Tumor with Inferior Vena Cava Thrombus Utilizing Cardio-Pulmonary Bypass: A Case Report**

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*Abstract.* Primitive neuroectodermal tumors are very rare in adults or children, and rarely affect the genitourinary system and usually are large tumors on presentation. These tumors are highly aggressive and can metastasize to lung, pleura, bones and liver. The present case is a 15-year-old female, who was presented with a huge renal mass with tumor extending into the inferior vena cava. She underwent complete removal of the primary kidney tumor, and removal of the IVC tumor utilizing cardio-pulmonary bypass and deep hypothermic circulatory arrest.

*Keywords:* Primitive neuroectodermal tumors, Cardio-pulmonary bypass, Inferior vena cava.

### **Introduction**

Primitive neuroectodermal tumors (PNET) are very rare and small cell tumors in adults or children<sup>[1]</sup>. These tumors most often occur as bones or soft tissue masses in the trunk or axial skeleton. Primary renal PNET is an extraordinary rare neoplasm. Primitive neuroectodermal tumor (PNET) occurs mainly in older children and adolescents with 75% of cases before the age 35 years, and a mean age of 20. These tumors are usually large at presentation and highly aggressive in nature, and are

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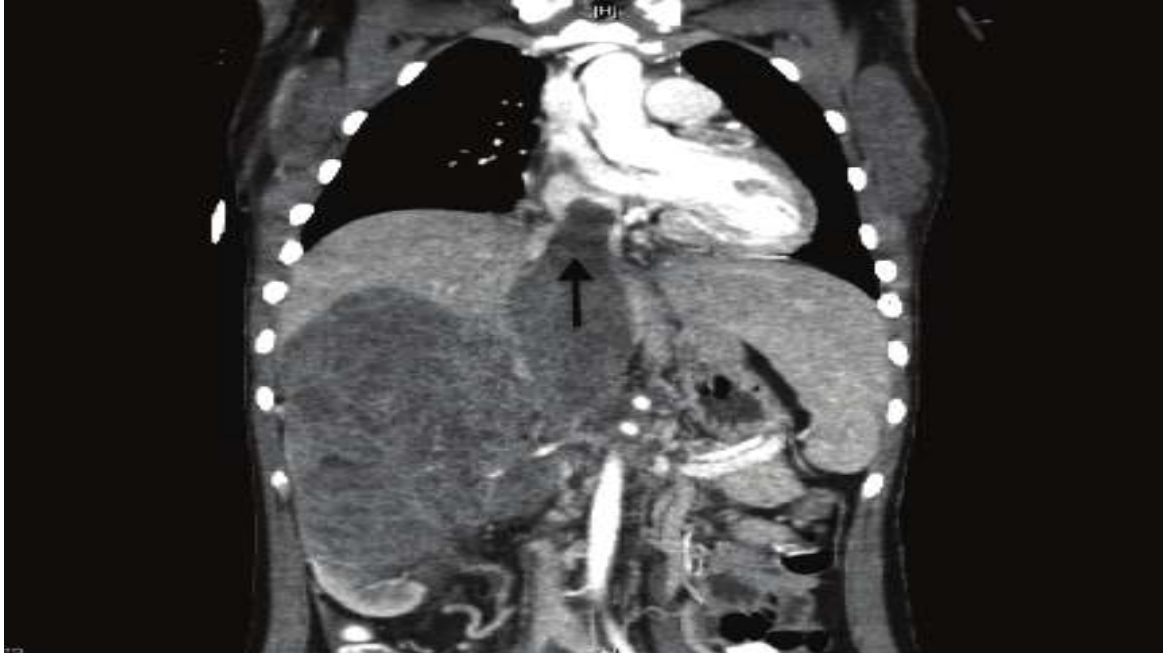
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present with flank pain and/or hematuria<sup>[2]</sup>. Metastases to the lung, pleura, bones and liver can occur with PNET of the kidney<sup>[2]</sup>. Renal PNET shows poor response to chemotherapy<sup>[1]</sup>. This report presents an adolescent patient who underwent successful resection of PNET of the kidney, and inferior vena cava thrombus (IVC) through a laparotomy incision and sternotomy utilizing cardio-pulmonary bypass (CPB) and deep hypothermic circulatory arrest (DHCA).

### Case Presentation

The legal guardian signed a written informed consent to publish this case report. A 15-year-old female presented with a one-week history of right upper quadrant pain and fullness. Abdominal ultrasound was done, which showed a mass that could be originating from either the kidney or the liver. Past medical history revealed bronchial asthma that was controlled with an inhaled beta agonist. Abdominal CT scan showed a kidney tumor, which was 10 × 18 cm with IVC thrombus extending to the right atrium (Fig. 1). An image-guided biopsy confirmed the diagnosis of PNET of the kidney. Liver and kidney function were both normal despite the mass around both structures. Initial trans-thoracic echocardiogram confirmed that the IVC was almost completely obstructed by a large echogenic mass with extension into the right atrium. No other abnormalities were found and cardiac function is within the normal range. At this point, the patient started on enoxaparin due to the fear of embolization. Then, bridged to heparin infusion 24 hrs prior to the surgical procedure and the infusion was stopped 2 hrs before coming to the operating room. The patient received four cycles of chemotherapy to decrease the tumor size, but with no evidence of decrease tumor size on CT. At this point, it was felt that the best strategy was to proceed through surgically resection of the tumor and the IVC thrombus. The patient was clearly anxious, and on the day of the procedure, intravenous midazolam was given for preoperative sedation: a total dose of 120 mcg/kg was given *via* the *in-situ* peripherally inserted central line in the right upper limb. Once in the operating room, all monitors were applied, including electrocardiogram (ECG), non-invasive blood pressure (NIBP), pulse oximeter and end tidal carbon dioxide (EtCO<sub>2</sub>) once intubated. Both esophageal and rectal temperatures were monitored during the case. General anesthesia was induced using propofol 2 mg/kg, fentanyl 5 mcg/kg and pancronium 0.15 mg/kg and the trachea were intubated.

Anesthesia was maintained with isoflurane in air/oxygen mixture 60/40% and intermittent doses of fentanyl and pancronium. The plan was to keep the patient intubated after the surgery as to extend the surgical incision.



**Fig. 1.** CT Abdomen showing the tumor arising from the right kidney and extending into the IVC (arrow).

Thus, opted not performed epidural for postoperative pain management, because of the likely hood of using CPB and DHCA, and the risk of coagulopathy. One dose of methylprednisolone 30 mg/kg was given in anticipation for the institution of DHCA. Invasive monitoring was inserted after the induction of general anesthesia, which included right radial arterial line and a left internal jugular central venous line. Two 14 gauge intravenous catheters were inserted for volume transfusion, if needed. The risks and benefits of central vein cannulation were discussed, and for that reason, the left internal jugular was used and a small 4 F line was inserted, and to take the line as early as possible in the postoperative period. In this case, the CVP monitoring is crucial as to anticipate massive fluids changes, and the need of central access if vasoactive medications need to be started post CPB and DHCA. Bilateral near infrared spectroscopy probes were used to monitor brain saturation. Both sternotomy and laparotomy were performed. Once the sternotomy was performed, purse string was applied on both, the ascending aorta and the right atrial appendage in case massive pulmonary embolism occurs, and the need for urgent institution of CPB. The tumor

was dissected without any complications and the IVC was isolated below the diaphragm. Following an appropriate dose of heparin, the ascending aorta, the superior vena cava, and the IVC below the diaphragm were cannulated and cardiopulmonary bypass (CPB) was established. When the patient cooled to a core temperature of 20°C, the ascending aorta was cross-clamped, one dose of cardioplegia was given, and CPB was discontinued. The atrium was opened and the thrombus was immediately obvious. It was removed and CPB was re-initiated after 9 minutes of total arrest. Once adequate core temperature achieved, the patient weaned from CBP without difficulty. The patient required a total of 4 units (1200 ml) of packed red blood cells and one dose of platelets to achieve homeostasis. The patient was transferred to the intensive care unit intubated and sedated, and was extubated within 18 hrs after the procedure with no neurological deficit. Once the patient was extubated, she started on morphine *via* patient controlled analgesia (PCA) and then switched to oral morphine. The patient was discharged home after two weeks, and would continue with chemotherapy.

### **Discussion**

Pediatric patients presenting for major surgical procedures present a challenge for the anesthesia care team and provoke major anxiety to both, the patient and the family. This case in particular had many issues during the preoperative period. The size of the tumor and the proximity to major blood vessels increase the risk of major blood loss, and it is crucial to be prepared for such event. Insertion of large bore peripheral intravenous catheters is important; especially in this case as inserting a large bore central line may increase the risk of central vein thrombosis. Blood products should be available in the operating room in case of sudden major bleeding encountered. Pain management is another challenge in this case; in the ideal situation thoracic epidural will be an optimal for pain control. The incision was quite large in which both sternotomy and laparotomy was performed, and in such case, thoracic epidural will provide excellent pain control. The risk of an inserting a thoracic epidural was discussed as we felt there is a high chance of using DHCA. The associated coagulation derangement, epidural hematoma is a real risk especially if bloody tap occurred upon inserting the epidural catheter. Intra-venous morphine was used as the primary pain management for this case. Other options such as transverse abdominal block (TAP);

paravertebral block is not suitable for this case. The presence of a tumor thrombus in the right atrium presents a real risk of massive pulmonary embolism, which could occur during the induction phase of anesthesia or during dissection of the tumor. Intravascular tumor extension into the IVC occurs in about 15% of cases with renal carcinoma and into the right atrium in approximately 1% of patients. Tumor thrombus can be staged according to the nevus classification as level 1 (renal vein only), level 2 (infrahepatic IVC), level 3 (retrohepatic IVC close to the main hepatic veins), and level 4 (supradiaphragmatic IVC or right atrium). The perioperative mortality is about 2-4% mainly due to massive pulmonary embolism<sup>[3]</sup>. CPB ideally should be used only in patients with level 4 thrombus<sup>[4]</sup>. CPB has many applications in both cardiac and non-cardiac procedures<sup>[5]</sup>. It will support both circulation and oxygenation, and make it safer in-patients for which maintaining oxygenation or circulation is difficult due to severe airway or major vessels compression. CPB and the use of deep hypothermic arrest have been used in cases in which catastrophic blood loss is anticipated specially in small children, such cases as massive liver tumors<sup>[6]</sup>. In this case, the use of CPB and DHCA was of great value in achieving stable hemodynamic once the abdominal IVC was opened to resect the tumor, and by bypassing the IVC through the CPB circuit the venous return is not compromised. The use of DHCA in this case was also of value at the stage intra-cardiac tumor in which the surgeon has a clear field, and minimizing the risk of tumor spread from the CPB suction tubing. The presence of cancer always complicates the management of these cases, due to the fear of spreading the cancer cells, the effect of CPB on the immune system and the body defense against tumor cells. Invasion of the IVC with the tumor or thrombus formation sometimes necessitates the use of CPB to resect the tumor, especially if there is extension into the right atrium. Early tumor resection may need to be done early if the tumor is bigger in size or embolization to the lungs occurs. Some data from the adult population showed no difference in recurrence or mortality in patients with malignancy that underwent cardiac procedure with CPB or off-CPB<sup>[7]</sup>. DHCA involves cooling the patient down to 10-15°C mainly to decrease the metabolic rate and provide brain protection<sup>[5]</sup>. The cooperation and communication between the anesthesia team and the surgical team in complex surgical procedure is crucial, especially if part of the surgical team is not familiar with the implications of using CPB and DHCA. One of these concerns is using the CPB to suction debris from the surgical

field into the bypass circuit. Many other applications for the use of CPB have been reported in different surgical specialties. From the anesthesia prospective, patient with massive airway tumors such as thyroid cancer or intra-tracheal obstructing tumor can be managed with CPB for preventing severe hypoxia<sup>[5]</sup>. Preoperative assessment is crucial for this type of cases, in our case the tumor was arising from the right kidney and because of the size of the tumor it was in close proximity to the liver. Both renal and liver function test were important to evaluate because of the role of both organs in fluid homeostasis, drugs metabolism and excretion. It is even more important for the kidney function because it was evident that the patient will lose that kidney and paying close attention to the left kidney function, avoid nephrotoxic medications, and preserve renal perfusion. Cardiac assessment is also important because the tumor thrombus in the right atrium, which could extend and involve the tricuspid valve and lead to destruction of the valve. The results in tricuspid valve regurgitation could lead to obstruction of the inflow to the right ventricle. Involvement of the right ventricle can also occur as the tumor may attach to the right ventricular wall and may result in right ventricular dysfunction. Echocardiogram is the main diagnostic tool in this case. In this patient, the tumor thrombus was in the right atrium and not attached to the tricuspid valve, and that was also confirmed intra-operatively on opening the heart and inspecting the tricuspid valve. One also have to keep in mind that atrial communication in term of patent foramen ovale (PFO) could lead to embolization to the left side of the heart which can lead to systemic embolization. No PFO were seen in this patient and no evidence of brain emboli on pre-operative MRI. Cardiac function was normal before and after the procedure, and with no evidence of involvement of the right ventricle. Invasive monitoring is very important in this type of cases. Central line insertion is important for fluid management and the delivery of vasoactive medications when needed. The insertion of central line in this type of cases is associated with risk of venous thrombosis, especially if the line in close proximity to the right atrium and the tumor thrombus. In this patient, the central line was inserted into the left internal jugular vein and the smallest central line available was used, which was 4F and 8 cm long. The position of the central line was not confirmed intra-operatively, but the position was confirmed in the postoperative chest X-ray. The central line was removed on the second postoperative day and the patient had no evidence of central vein thrombosis in the postoperative period.

## Conclusion

Primitive neuroectodermal tumor (PNET) is a very rare tumor, both in adult and pediatric population that can arise from the kidney and invade the venous system and the thrombosis can extend into the right atrium. CBP can be used in cases in which the tumor extends into the IVC and to the right atrium. This approach will reduce risk of tumor fragmentation and cellular embolization.

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# التعامل التخديري لمريض مصاب بورم في الكلية مع خثرة في الوريد الأجوف السفلي بالاستفادة من تقنية الجهاز القلبي الالتفافي : تقرير الحالة

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المستخلص. أورام الأديم العصبي الظاهر نادرة الحدوث في الأطفال والبالغين. أورام الأديم العصبي الظاهر نادرا ما تصيب الجهاز البولي والتناسلي وعادة تكون أوراماً كبيرة عند الظهور. هذه الأورام هي عدوانية للغاية ويمكن أن ينتقل إلى الرئة والجنبه والعظام والكبد. هذه الحالة هي لطفلة تبلغ من العمر ١٥ عاماً مع تورم من هذا النوع في الكلية مع امتداد الورم إلى الوريد الأجوف السفلي. تم إخضاعها إلى عملية استئصال للتورم من الكلية مع استئصال الانتشار إلى الوريد الأجوف السفلي وذلك باستخدام تقنية الالتفاف القلبي الرئوي، وتقنية تبريد الجسم الدموي العميق.