

Pediatric paraganglioma of the posterior mediastinum

A case report and review of literature

Miao Yuan, MD^a, Chang Xu, MD^{a,*}, Gang Yang, MD^a, Weiya Wang, MD^b

Abstract

Rationale: Paraganglioma is rare in children and most pheochromocytomas originate in the adrenal gland.

Patient concerns: The clinical characteristics, diagnosis, and managements in a 9-year-old boy who presented with vomiting and occasional headache with a blood pressure of 210/170 mm Hg was retrospectively reviewed. CT scan of the chest revealed a 7 × 5-cm-sized soft tissue mass in the left paraspinal area. Biochemical reports revealed elevated levels of serum norepinephrine, urine norepinephrine, urine dopamine, and serum neuron specific enolase.

Diagnoses: The immunohistochemical studies suggested that the tumor was a paraganglioma of the posterior mediastinum.

Interventions: The patient underwent mass resection with thoracotomy. Before operation, the patient was prepared by orally administering captopril, propranolol hydrochloride, and phenoxybenzamine. Body fluid volume was also prepared by vein and mouth in 3 days.

Outcomes: The patient was followed up postoperatively for 1 year without recurrence.

Lessons: We should be highly vigilant the pediatric tumor of the posterior mediastinum with vomiting and headache as the first clinical manifestation.

Abbreviations: AFP = alpha fetoprotein, CEA = carcinoembryonic antigen, cgA = chromogranin A, CT = computed tomography, IHC = immunohistochemical, nl = normal limits, NSE = serum neuron specific enolase, syn = synaptophysin.

Keywords: children, paraganglioma, posterior mediastinum

1. Introduction

Pheochromocytomas and paragangliomas, which arise from chromaffin cells of the adrenal medulla and extra-adrenal paraganglia, respectively, are uncommon. Approximately 95% of catecholamine-secreting tumors are located in the abdomen and pelvis.^[1] Pediatric paraganglioma of the posterior mediastinum is reported to be exceedingly rare, and there is a lack of guidelines for its perioperative managements. Here, we report a rare case of a paraganglioma of the posterior mediastinum in a 9-year-old boy.

2. Consent

The patient's parents provided informed consent for the publication of his clinical and radiological data. This case report

was approved by Medical Ethical Committee of West China Hospital, Sichuan University.

3. Case report

A 9-year-old boy presented to a local hospital with vomiting and occasional headache with a blood pressure of 210/170 mm Hg. No obvious diseases were observed on digestive endoscopy and abdominal computed tomography (CT) scan, and no remarkable improvement by medicine treatment. CT scan of the chest revealed a 7 × 5-cm-sized soft tissue mass in the left paraspinal area from T3 to T7 with destruction of the adjacent thoracic vertebra and ribs (Fig. 1). Biochemical reports revealed elevated levels of serum norepinephrine, urine norepinephrine, urine dopamine, and serum neuron specific enolase. Serum epinephrine, urine epinephrine, alpha fetoprotein, and carcinoembryonic antigen were within the normal range (Table 1). The admitting diagnosis was tumor in the posterior mediastinum: paraganglioma? Before operation, the patient was prepared by orally administering captopril, propranolol hydrochloride, and phenoxybenzamine by mouth. The patient's blood pressure remained stable at approximately 110/80 mm Hg. In addition, body fluid volume was also prepared by vein and mouth in 3 days before surgery.

Thoracotomy was performed through the left fifth intercostal space. Intraoperatively, several membranous and fascicular adhesions existed in the thoracic cavity. The irregular ovoid mass measured 8 × 7 × 5 cm. The tumor originated from the nerve root and adhered to the surrounding tissue. It invaded the spine and chest wall. The mass was tough and rich in blood supply.

Editor: N/A.

The authors have no conflicts of interest to disclose.

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Medicine (2018) 97:27(e11212)

Received: 30 January 2018 / Accepted: 28 May 2018

<http://dx.doi.org/10.1097/MD.00000000000011212>

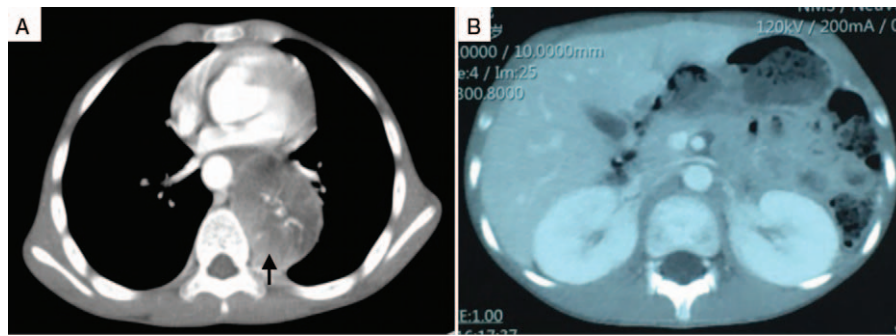


Figure 1. A, CT scan of the chest showed a 7 × 5-cm-sized soft tissue mass on the left paraspinal with destroying adjacent thoracic vertebra and ribs (arrow, the tumor). B, CT scan of the abdomen showed normal. CT = computed tomography.

Table 1
Levels of biochemical test.

Factors/parameters	Levels	Normal limits	Change
Serum norepinephrine, ng/L	23,231	272–559	↑
Urine norepinephrine, ug/24 h	2983.72	16.3–41.5	↑
Urine dopamine, ug/24 h	1807.10	107.2–246.6	↑
NSE, ng/mL	24.26	<15	↑
Serum epinephrine, ng/L	121	54–122	Normal
Urine epinephrine, ug/24 h	8.97	7.5–21.9	Normal
AFP, ng/mL	2.40	<8	Normal
CEA, ng/mL	2.22	<3.4	Normal

AFP = alpha fetoprotein, CEA = carcinoembryonic antigen, NSE = serum neuron specific enolase.

There were intraoperative changes in the patient’s blood pressure, which ranged from 85/50 mm Hg to 180/130 mm Hg. During the resection, the surgeon closely communicated with the anesthesiologist to decide the operative process. Histological studies demonstrated that the mass was a tumor (Fig. 2). Immunohistochemical (IHC) studies demonstrated that tumor cells stained positive for synaptophysin (syn, +) and chromogranin A (cgA, +). The positive rate of Ki67 (MIB-1) staining was 2% to 5%. The S100 and PCK staining was negative (Fig. 3). The immunohistochemical studies suggested that the tumor was a paraganglioma. Postoperatively, the patient’s blood pressure was stable and within the normal range. On postoperative day 2, the concentration of serum epinephrine was 236 ng/L and serum norepinephrine was 4686 ng/L. On postoperative day 4, serum norepinephrine (321 ng/L) and epinephrine (76 ng/L) were

normal. The patient was discharged on postoperative day 6. After operation, the patient did not exhibit hypertension and his blood pressure was normal without medicine. After 1 year, follow-up chest CT did not reveal tumor recurrence (Fig. 4).

4. Discussion

Patients with paragangliomas often present with symptoms secondary to catecholamine excess. The common symptoms are headache, palpitations, sweating, nausea, vomiting, and weight loss. Hypertension is observed in 70% to 90% of children with pheochromocytoma.^[2] Except the presenting symptoms, the catecholamines in the blood and metabolites in the urine are always elevated.^[3] Radiological tests are helpful to determine the tumor location. It is important to distinguish from the other tumors before operation. Furthermore, α-adrenergic or/and β-adrenergic blockade, which are considered to decrease the risk of cardiac dysrhythmia accompanied with pheochromocytoma during the surgery, are used to maintain normal blood pressure normal.^[4,5] Meanwhile, patients with pheochromocytoma usually have low blood volume^[6] and it is necessary to counteract hypovolemia after removing the tumor due to sudden decrease in serum catecholamine levels.^[7] Our patient presented the same symptoms such as vomiting, headache, and hypertension, as those previously reported in the literature. Preoperative biochemical studies revealed that serum and urine norepinephrine were elevated and serum and urine epinephrine were within the normal range. The results suggested that the tumor mainly produced norepinephrine. Postoperatively, serum norepinephrine continued to decline. However, serum epinephrine increased

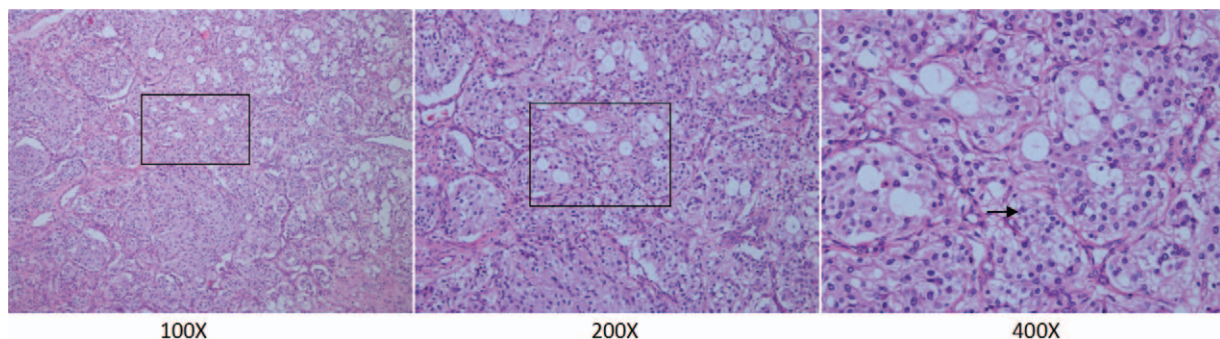


Figure 2. Histologic studies showed eosinophilic granular cytoplasm with hyperchromatic and round nuclei (arrow, hematoxylin and eosin, magnification ×100, ×200, ×400).

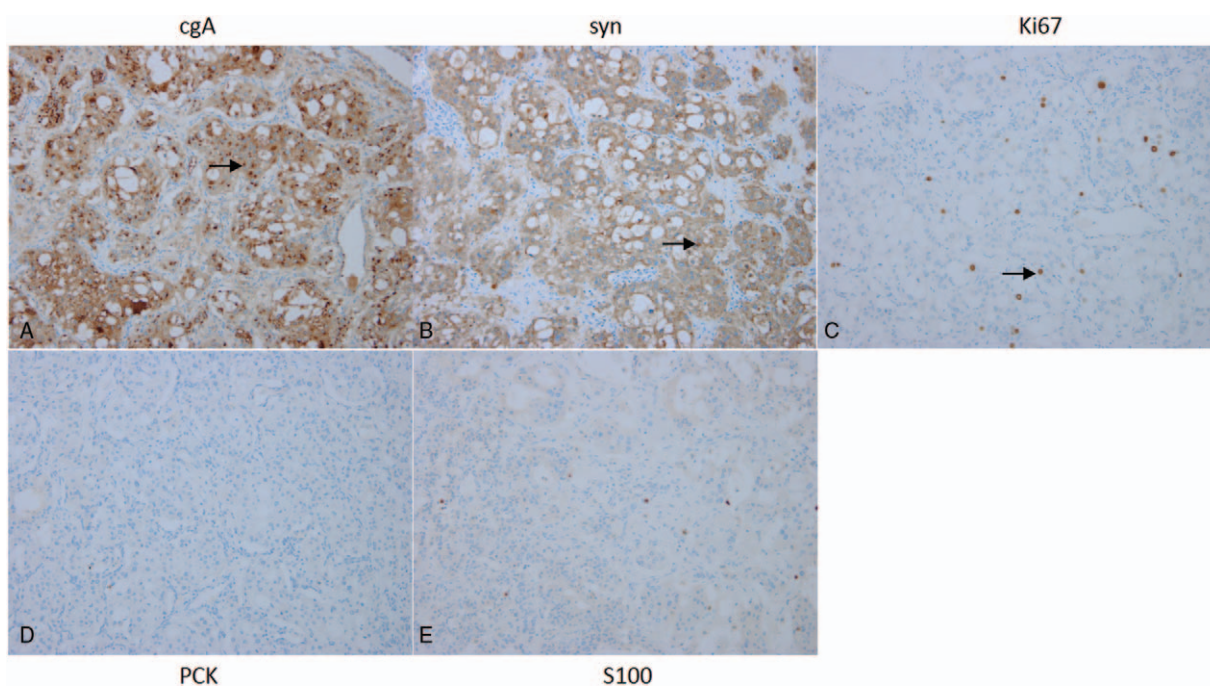


Figure 3. cgA and syn antibody IHC staining showed positive reactivity (A, B, arrow). The positive rate of Ki67 staining was 2% to 5% (arrow); S100 and PCK antibody IHC staining showed negative reactivity (D, E); magnification $\times 200$. IHC = immunohistochemical.

on postoperative day 2 and then was within the normal range, which may be due to the intraoperative extrusion of the tumor. To maintain stable intraoperative blood pressure, blood volume was prepared for physiological need of sodium solution intravenously and orally on preoperative day 3.

Renard et al^[8] reviewed the surgery and perioperative care required for patients with pheochromocytoma and paraganglioma. Surgical resection for paraganglioma was regarded as the gold standard of treatment options. Intravenous midazolam and propofol were used as anesthesia. Tsirlin et al^[11] suggested that fentanyl, ketamine, and morphine should be avoided because they may stimulate catecholamine release. Intraoperatively, tumor vessels were first managed to prevent catecholamine entering the vascular structure. Meanwhile, it was necessary to closely communicate with the anesthesiologist to control

hemodynamics. Once the blood pressure was elevated, the operation was suspended. Vasopressin was used for hemodynamic resuscitation to control acute catecholamine deficiency after tumor removal.^[9]

The tumor was excised for histological and immunohistochemical examinations. The accurate diagnosis of paraganglioma could be based on immunohistochemical staining of syn, cgA, Ki-67, S100, and PCK. However, the definition of malignancy in paraganglioma was based on not only histological and immunohistochemical characteristics but also the presence of metastasis and the size of the tumor.^[10] Owing to the relative rarity of the tumor, chemotherapy for children was absent and unsatisfactory.^[11] In our patient, preoperative CT of the chest and abdomen suggested no other problems besides the mass in the posterior mediastinum. Meanwhile, surgical resection was complete. Postoperatively, the symptoms of vomiting and headache disappeared. The patient's blood pressure remained normal without the use of hypotensors. Biochemical test results revealed that serum and urine norepinephrine and epinephrine levels returned to normal. Our patient decided to undergo long-time follow-up without chemotherapy. During the follow-up, monitoring the patient's symptoms and blood pressure was important. CT of the chest and biochemical studies are essential 3 months, 1 year, and then each year postoperatively.

5. Conclusions

The experience of treating pediatric paraganglioma of the posterior mediastinum is insufficient. Our treatment was based on some case reports and experience with adults. Our patient was rehabilitated and discharged using a series of perioperative managements. However, pathological diagnosis could not provide benign or malignant diagnosis, and long-term follow-up is still needed.

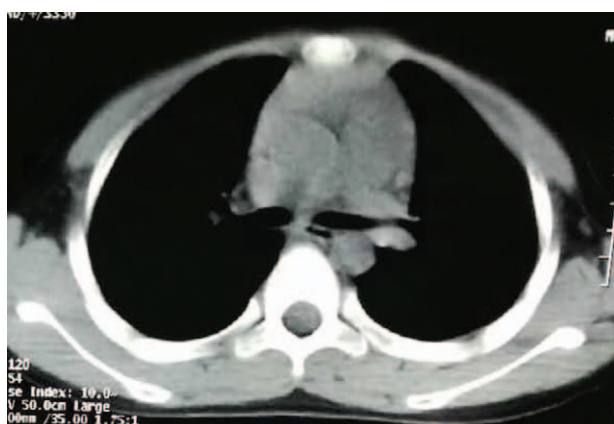


Figure 4. Postoperative CT scan of the chest showed negative for tumor recurrence.

Author contributions

Conceptualization: Chang Xu.

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Formal analysis: Gang Yang.

Writing – original draft: Miao Yuan.

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