PHEOCHROMOCYTOMA PRESENTING WITH POLYDIPSIA AND POLYURIA IN A CHILD

Poliüri ve polidipsi ile gelen feokromasitomalı çocuk: Olgu sunumu

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Abstract : *Pheochromocytomas are rare tumors in childhood and can mimic many unrelated diseases due to their various presenting symptoms. While hypertension is the most prevalent finding of pheochromocytomas, polyuria and polydipsia are rare and interesting symptoms. In this study we presented a child with unilateral pheochromocytoma, whose first symptoms were polyuria-polydipsia, and hypertension, which were important clues for pheochromocytoma. With the help of clinical and laboratory findings, patient was diagnosed as pheochromocytoma and referred to surgery; with the removal of the tumor the symptoms disappeared. When the family members were screened for multiple endocrine neoplasia (MEN) syndromes, a bilateral pheochromocytoma was diagnosed in his sister and she was also operated on immediately. In this article we emphasized that polyuria-polydipsia may be the first symptoms of pheochromocytoma in children, the importance of blood pressure measurement in initial physical examination and the familial pattern of pheochromocytoma.*

Key Words: *Pheochromocytoma; polyuria; child; hypertension.*

Özet : *Feokromositoma çocukluk çağında nadir görülen tümörlerdendir ve farklı semptomları ile birçok hastalığı taklit edebilir. Hipertansiyon feokromositomada en sık bulgu olmasına rağmen, poliüri-polidipsi nadir ve ilginç semptomlardır. Bu yazıda tek taraflı feokromositomalı, ilk semptomu poliüri-polidipsi olan ve fizik muayenede hipertansiyon tespit edilen vaka takdim edilmiştir. Klinik ve laboratuar bulguları ile tanı konulan olguda cerrahi rezeksiyon sonrası bulgu ve belirtiler kayboldu. Multipl endokrin neoplazi (MEN) açısından aile bireyleri tarandı ve kız kardeşine de feokromositoma tanısı konularak opere edildi. Bu makale ile poliüri ve polidipsinin feokromositomanın ilk semptomları olabileceğini, ilk muayenede tansiyon ölçülmesinin önemini ve feokromositomanın ailesel olabileceğini vurgulamak istedik.*

Anaktar Kelimeler: *Feokromositoma; poliüri; çocuk; hipertansiyon*

Pheochromocytoma is a cathecholamine-secreting tumor, most often benign (90 %), and is derived from chromaffin cells of surrenal medulla. Pheochromocytoma is a rare disease in childhood, and it frequently occurs between 6 to 14 years of age. In contrast to adults, children with pheochromocytoma have a higher incidence of bilaterality (in >20 % of affected children), a higher association with the multiple endocrine neoplasia (MEN) syndromes, and a lower incidence of malignant neoplasms (1-4).

Classical clinical triad includes headache, sweating and nausea while adult population present with triad of headache, palpitation with or without tachycardia, and sweating (3). However, presentation may be atypical. Pheochromocytoma is an uncommon, but an

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important cause of surgically correctable hypertension resulting from hyperproduction of epinephrine and norepinephrine (5). Untreated patients die because of uncontrolled hypertension. Other symptoms that can be seen in pheochromocytoma are tremor, exhaustion, anxiety, dyspnea, flushing or warmth, blurred vision, dizziness, convulsion, tinnitus, dysarthria, polyuria and polydipsia (3,6). The polyuria and polydipsia in pheochromocytoma are associated with hyperglycemia (3,6). The symptoms of our patient were headache, polyuria and polydipsia. In this article we wanted to emphasize on polyuria and polydipsia, which are rare symptoms in pheochromocytoma and the importance of initial blood pressure measurement, which helped the clinician reach the diagnosis of this disorder.

Case

A fourteen-year-old boy with a history of sweating, palpitation, headache, polyuria and polydipsia for two years, was admitted to our hospital. He was drinking fluids excessively and his urinary output was 3 L/day (2.7 ml/Kg/hr). On initial physical examination his body weight was 47 Kg, and height was 159 cm. The blood pressure was high, (160/100 mmHg -170/120 mmHg, >97 %), and the pulse rate was 100 beats/min. Other findings were normal.

Laboratory data revealed normal serum electrolyte levels, liver and kidney function tests. Fasting blood glucose level was mildly elevated: 135 mg/dl. Urine vanilmandelic acid (VMA) level was 80 mg/g creatinine. Blood epinephrine and norepinephrine concentrations were 97 pg/mL and 15.331 pg/mL respectively. Because of elevated levels of VMA, epinephrine and norepinephrine some imaging modalities were used to image the cathecholamine-secreting tumor. Cranial computed tomography was within normal ranges, but abdominal ultrasonography revealed a homogenous, regular marginated mass lesion in the left surrenal area. In abdominal tomography a mass lesion in the left surrenal area, with regular margins (3x4x5 cm in diameter), central hypodensity and soft tissue density was demonstrated. During follow up period blood pressure increased up to 160- 200 mmHg. After the control of blood pressure with the administration of propranolol (2 mg/kg/ bid, per oral), sodium nitroprussid (5 g/Kg/min, IV infusion) and prazosin (3 mg/Kg, tid, per oral), a well capsulated tumor tissue 3x4x5cm in diameter originating from left surrenal gland, was totally excised with left surrenalectomy. No perioperative complications developed. Microscopic examination of the tumor established the diagnosis of pheochromocytoma (Figure 1, 2). When family members were screened for MEN syndrome, a bilateral pheochromocytoma was diagnosed in his 16-year-old sister and she was also operated on immediately. During the 9-month postoperative period no recurrence has occurred.

Discussion

Pheochromocytoma in children especially occurs in the second part of the childhood and is more frequently seen in males with a male female ratio of 2:1 (1-5). In contrast to adults, bilateral involvement is more frequent (7). Approximately 10 % of pediatric pheochromocytoma is thought to be familial. In addition to familial syndrome of pediatric isolated pheochromocytoma, pheochromocytomas in children are associated with other inherited diseases, including von Hippel-Lindau, tuberous sclerosis, Sturge-Weber syndrome, and as a component of multiple endocrine neoplasias (MEN IIA-IIB) (8-10). Our case was a boy in late childhood, who had unilateral surrenal involvement and had a sister with bilateral surrenal involvement.

Clinical findings of pheochromocytoma are usually the result of hyperproduction of epinephrine and norepinephrine and a clinical diagnosis could be made with these symptoms. The most prominent symptom is hypertension; while intermittent headache, palpitation, sweating and flushing are other common symptoms(1,10-12). The first symptom of our patient was headache, and it was detected that he had hypertension. Dorairajan et al.(13) retrospectively reviewed 162 cases of endocrine-based hypertension and reported an incidence of 48.7 % of

pheochromocytoma. In a referral hospital based study, in malignant hypertension patients, pheochromocytoma was responsible for 4.4 % of cases (14).

Polydipsia and polyuria, the other complaints of our patient are rare symptoms in pheochromocytoma (15), but may be seen in 25 % of pheochromocytoma. After the surgical excision of the surrenal mass these symptoms regressed in our patient. In previous studies, it was noted that polyuria and polydipsia might be found as a result of increased glycolysis/glycogenolysis and alpha-receptor–mediated inhibition of insulin release. This insulin inhibition causes an increase in blood sugar levels and glucose intolerance. As a result, patients may present with diabetes mellitus or glucose intolerance, most commonly during paroxysms (3,5,16). There is a mild to moderate glucose intolerance in surrenal disorders. Also lipolysis increase in pheochromocytoma (17). Marked hyperglycemia, glycosuria and polyuria are uncommon and ketosis is rare. However, in our case fasting blood glucose levels were mildly elevated, and could not be the cause of polyuria or polydiypsia. The cause may have been increased metabolism which is the result of excess epinephrine and norepinephrine.

Measuring blood pressure is one of the basic evaluation criteria of patients in small clinics, although, sometimes it is overlooked or perhaps ignored by the clinician. Our case is a good example demonstrating the importance of basic health index measurements (weight, height, fever, blood pressure, respiratory and hearth rate). By detecting the hypertension, with the help of anamnesis, physical examination and the basic laboratory examinations the diagnosis was clear in our case.

Pheochromocytoma may be isolated, familial or a component of MEN syndromes. In a twenty-five-year study of Reddy et al (18), 2 of the 11 cases were associated with inherited syndromes (multiple endocrine neoplasia 2A and von Hippel-Lindau). In the control of family members we detected a bilateral pheochromocytoma in the sister, but there were no other findings for MEN syndromes, thus our case was isolated familial pheochromocytoma.

We present this case to emphasize that polydipsia and polyuria may be the presenting sign of pheochromocytoma. The importance of blood pressure measurement in initial physical examination and also the familial pattern of the disease were also emphasized.

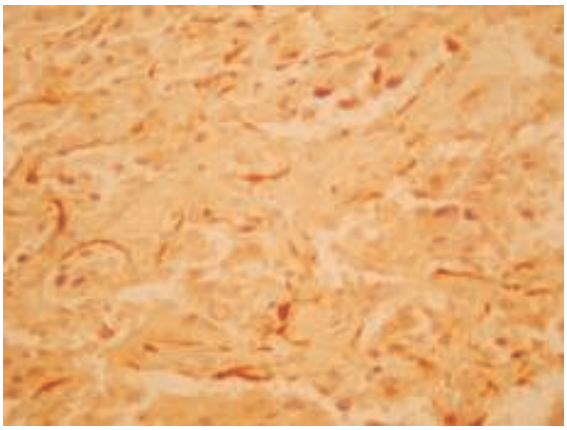

Fig 1: Pheochromocytoma with delicate sustentacular network outlined by the immunostain for S 100 protein (S100, x400)

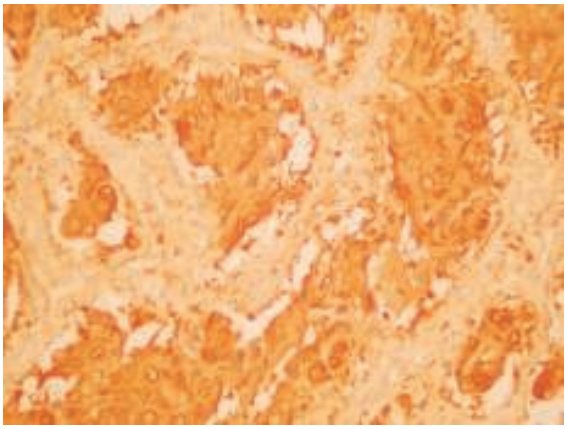

Fig 2: Diffuse immunostaining for neuron specific enolase (NS) in pheochromocytoma (NSx400)

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