CASE REPORT

NEUROBLASTOMA DIAGNOSED AFTER SURGERY FOR A BENIGN CONDITION

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ABSTRACT

We report a case of neuroblastoma diagnosed in an 8-month-old infant who was being followed-up due to antenatal hydronephrosis. Postnatal imaging findings were suggestive of bilateral ureteropelvic junction (UPJ) obstruction. A left pyeloplasty was performed during the postnatal third month. An early postoperative computed tomography (CT) scan, performed because of considerable decrease in hemoglobin/hematocrit (Hb/Htc) values, revealed negative results for any kind of localized collection of fluid or tumor. At the postoperative fourth month, postnatal ultrasonography (USG) revealed a solid left adrenal mass. Blood and urine test results were suggestive of a neuroblastoma. On surgical exploration an adrenal mass was excised. Pathologic examination confirmed the diagnosis of neuroblastoma. The patient had stage 1 disease with favourable histopathologic features. This low-risk tumor, can be treated with primary surgery alone with excellent outcomes. Neuroblastomas either detected by mass screening programs or diagnosed incidentally on urinary tract imaging can be expected to be well localized and have a good prognosis.

Keywords: Neuroblastoma, Incidental Diagnosis, Antenatal Hydronephrosis, Surgical Treatment, Screening Programs

PYELOPLASTİDEN SONRA TANISI KONAN NÖROBLASTOM

ÖZET

Bu yazıda antenatal hidronefroz nedeniyle takip edilen ve pyeloplastiden sonra nöroblastom tanısı alan 8 aylık bir erkek çocuğun klinik özellikleri bildirilmiştir. Postnatal 3. ayda gerçekleştirilen sol pyeloplastinin erken postoperatif döneminde belirgin hemoglobin, hematokrit düşüşü olması nedeniyle çekilen tomografide tümör veya kolleksiyon lehine bulgu izlenmedi. Fakat postoperatif 4. aydaki kontrol ultrasonografisinde sol sürrenalde kitle görüldü. Kan ve idrar testleri nöroblastom lehine sonuçlandı. Bunların üzerine yapılan eksplorasyonda sol sürrenaldeki kitle eksize edildi ve iyi histopatolojik özelliklere sahip, evre 1 nöroblastom tanısı konmuş oldu. Bu vakada olduğu gibi insidental olarak veya tarama programları ile tanısı konan düşük riskli tümörler sadece cerrahi müdahale ile tedavi edilebilir.

Anahtar Kelimeler: Nöroblastom, İnsidental Tanı, Antenatal Hidronefroz, Cerrahi Tedavi, Tarama Programları

INTRODUCTION

Neuroblastomas are the most common extracranial solid tumor in children and accounts for 8% to 10% of all cancers in children. Patients diagnosed under 1 years of age (40%) have a much better prognosis than those diagnosed in later childhood[1]. The wide range of clinical presentations makes the diagnosis, clinical management and follow-up a challenge. Here, we report a case of a neuroblastoma diagnosed in an 8 months old

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infant who was being followed-up due to antenatal hydronephrosis.

CASE REPORT

Our patient was being followed-up due to antenatally diagnosed bilateral hydronephrosis. Postnatal ultrasonography (USG), intravenous pyelography (IVP) and nuclear imaging (Tc99m DMSA and DTPA scan) findings were suggestive of bilateral ureteropelvic junction (UPJ) obstruction. Diminished renal parenchymal thickness and relatively low radiopharmaceutic uptake (30%) on the left side were the main findings. On the third postnatal month, a left pyeloplasty was performed. During the early postoperative period due to a considerable decrease in hemoglobin/hematocrit values (Hb: 7.2 gr/dl, Htc: 21%) a computerized tomography (CT) was carried out. The CT scan was negative for any retroperitoneal collection or space occupying lesion (Figure 1). The patient was followed conservatively and discharged from the hospital after an uneventful postoperative period. After discharge, the patient was recruited for follow-up visits at regular intervals. At the fourth postoperative month, USG revealed that a solid, hypoechoic mass, 3x2 cm in diameter, was located near the left adrenal gland. Initially it was considered to be a hematoma based on early postoperative findings and the benign radiologic appearance. A CT scan localized an adrenal, hypovascular mass, 3 x 4.5 cm in diameter, with regular contours (Figure 2). There was no sign of local invasion or calcification. Moreover, grade 3 hydronephrosis was still evident in the right pelvicalyceal system. The CT scan was negative for any lymphadenopathy or metastatic lesion. Vascularization was present on doppler USG which was the main criterion to distinguish the mass from a hematoma. Serum blood tests (for neuron-specific enolase, lactic dehydrogenase and ferritin) and urinary catecholamine metabolite levels (for homovanillic acid, vanillylmandelic acid) were supportive of a neuroblastoma. Based on these findings an open surgical exploration was performed. At this one session, a left adrenal mass was completely excised and a pyeloplasty was performed on the right side with two seperate flank incisions. Histopathological evaluation confirmed the diagnosis of a stroma poor, differentiated neuroblastoma with a favorable prognosis according to the Shimada classification[1]. The tumor measured 4 cm in its greatest dimension. The surgical margins were free of any tumoral invasion. After an uneventful postoperative period the patient was discharged from the hospital on the fourth day. A metastatic evaluation was carried out postoperatively with bone marrow aspirates, a chest CT scan, radionuclide bone scan and a metaiodobenzylguanidine (MIBG) scan. The patient had stage 1 disease with favorable histopathologic features.

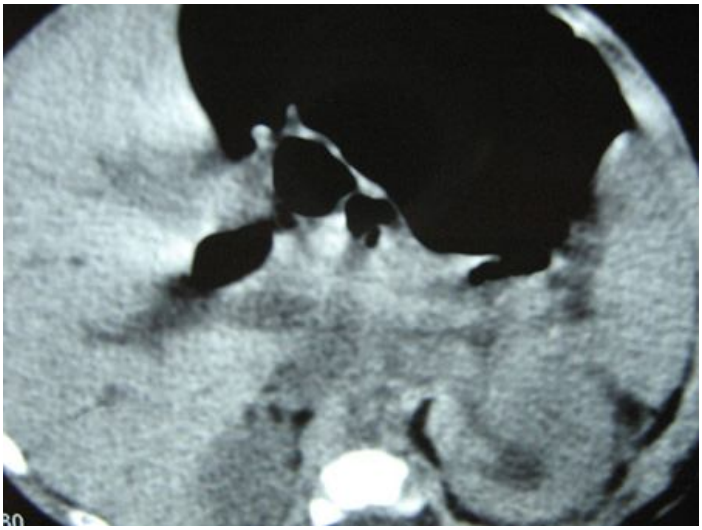

Figure 1: Early postoperative CT scan after pyeloplasty, demonstrating no sign of retroperitoneal collection or tumor

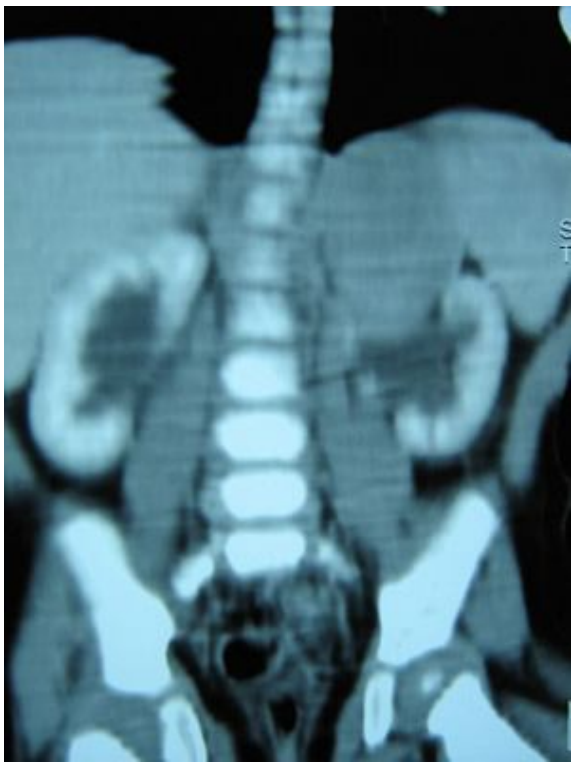

Figure 2: CT images 3 months after pyeloplasty, demonstrating an adrenal mass

DISCUSSION

As the most common extracranial solid tumors in children, neuroblastomas account for 8% to 10% of all cancers in children. The median age at diagnosis is 22 months and 80% of the children are diagnosed < 4 years of age[1]. Multiple cytogenetic abnormalities, amplification of the N-myc oncogene and changes in the normal diploid chromosomal content have been identified as a part of the disease process[2]. However, the exact pathogenetic mechanism remains uncertain. A variety of symptoms may be evident related to the site of the primary or metastatic tumor. Retroperitoneal and abdominal tumors (62%-65%) may present as a palpable mass or may cause abdominal pain, weight loss, anorexia, vomiting and symptoms related to a mass effect[3-6]. One fourth to one third of patients with a neuroblastoma present with a localized disease. Treatment depends on clinical stage, resectability and histopathology. Those with a localized, low-risk and resectable tumor can undergo primary curative surgery. On the other hand, intermediate-and high-risk patients are treated with a multimodal approach that combines surgery, chemotherapy and radiation[1].

In our case, the patient was asymptomatic and the mass was discovered incidentally on follow-up imaging studies. Diagnosis of this localized, small tumor before 1 year of age is definitely expected to improve treatment outcome. Similarly, mass screening of infants (< 6 months of age) by using urinary catecholamine metabolites, has reduced the occurrence of disseminated tumors and increased neuroblastoma detection rates[7]. Screening for neuroblastoma mainly detects tumors that might have regressed spontaneously[7,8]. As for our case, a neuroblastoma detected by mass screening usually shows a good prognosis. This low-risk tumor in our case could be treated with primary surgery alone with excellent outcomes. Overall 5 year survival reaches 90% with supplemental treatment needed in only 10% of these patients[1] On the other hand, mass screening may also detect unfavorable tumors in early stages.

The prognosis for children with a symptomatic neuroblastoma is dependent both on age and stage, with children aged under 1 year and those with tumours of stages I, II, and IVS having a much better prognosis[1]. Comparisons of the yield of cancers detected by screening and the expected cumulative incidence of neuroblastomas throughout childhood suggest that screening overdiagnoses many non-progressive cases, with consequent physical and psychological morbidity. In view of the lack of impact of screening programs on neuroblastoma mortality, and the fact that serious complications may arise from therapy, routine screening programs for children are not being supported[9,10].

Therefore in this particular age group imaging studies, carried out for a variety of reasons,

should be interpreted cautiously bearing in mind unusual presentation types of neuroblastomas. To our knowledge, this is the first case of neuroblastoma diagnosed incidentally on follow-up imaging studies after urinary tract surgery.

CONCLUSION

Neuroblastomas either detected by mass screening programs or diagnosed incidentally on urinary tract imaging for a variety of reasons can be expected to be localized and have a good prognosis. Urinary tract imaging, especially before 1 year of age, should be carried out cautiously bearing this possibility in mind.

REFERENCES

1. Lee KL., Ma JF, Shortlife LD. Neuroblastoma management, recurrence, and follow-up. Urol Clin N Am 2003: 30: 881-890.
2. Schilling FH, Spix C, Berthold F. Neuroblastoma screening at one year of age. N Engl J Med 2002; 346: 1047-5103.
3. Alexander F. Neuroblastoma. Urol Clin North Am 2002; 27: 393-402.
4. Castleberry RP. Biology and treatment of neuroblastoma. Pediatr Clin North Am 1997; 44: 919-937.
5. Simpson JK, Gaze MN. Current management of neuroblastoma. Oncologist 1998; 3: 253-562.
6. Ritchey M. Pediatric urologic oncology. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ, editors. Campbell's Urology. 8th edition. Philadelphia: W.B. Saunders; 2002:2469-2475.
7. Woods WG, Gao R, Shuster JJ. Screening of infants and mortality due to neuroblastoma. N Engl J Med 2002; 346: 1041-1046.
8. Yamamoto K, Ohta S, Ito E. Marginal decrease in mortality and marked increase in incidence as a result of neuroblastoma screening at 6 months of age: cohort study in seven prefectures in Japan. J Clin Oncol 2002; 20: 1209-1214.
9. Barrette S. Treatment complications in children diagnosed with neuroblastoma during a screening program. J Clin Oncol 2006; 24: 1542-1545.
10. Chamberlain J Screening for neuroblastoma: a review of the evidence. J Med Screen 1994; 1:169-175.