TÜBİTAK

CASE REPORT

Turk J Med Sci
2008; 38 (3): 281-283

E-mail: medsci@tubitak.gov.tr

Neuroblastoma in Adulthood

Turgut YAPANOĞLU[1]
İsa ÖZBEY[1]
Yılmaz AKSOY[1]
Nesrin GÜRSAN[2]
Özkan POLAT[1]

Abstract: Neuroblastoma is a common malignancy in children, but rarely occurs in adults. Neuroblastoma occurring in adolescents, compared to those occurring in children, are more frequently diagnosed in advanced stages and have common sites of metastases. Currently there are no standard treatment guidelines for patients with neuroblastoma in adulthood. If treated with standard protocols as those used for children, their survival is poor, particularly patients with localized disease seem to have a more aggressive course compared to children.

Key Words: Neuroblastoma, adult

Yetişkinde Saptanan Nöroblastom

Özet: Nöroblastom, çocukluk çağında en sık görülen malign hastalıktır, fakat yetişkinlerde nadir görülür. Yetişkinlerde görülen nöroblastomu, çocuklarda görülen nöroblastomla karşılaştırdığımızda, yetişkinlerdeki nöröblastom daha sık olarak ileri evre ve metastatik olarak karşımıza çıkmaktadır. Günümüzde yetişkin nöroblastomunun standart bir tedavi protokolu yoktur. Çocuklarda uygulanan tedavi protokolü yetişkinler için uygulandığında yetersiz kalmaktadır ve özellikle lokalize hastalık, çocuklara göre yetişkinlerde daha agresif seyretmektedir.

Anahtar Sözcükler: Nöroblastom, yetişkin

[1] Department of Urology, Faculty of Medicine, Atatürk University, Erzurum TURKEY

[2] Department of Pathology, Faculty of Medicine, Atatürk University, Erzurum TURKEY

Introduction

Neuroblastoma is a tumor derived from neural crest cells. It is the most common tumor in infants, the second most common solid tumor in children (1). Neuroblastoma rarely occurs in adults and, although abdomen and retroperitoneum are the primary sites in adults, the primary sites in children are adrenal glands, but may arise anywhere within the sympathetic nervous system (2).

Case Report

A previously healthy 54-year-old man presented with a chief complaint of right flank pain, body weight loss, pain in the right hip, and disabling fixed flexion contracture of the right hip started 4 weeks before. Clinical examination revealed mild tenderness and a mass in the right inferior quadrant. The computed tomographic scan showed a large mass sized $30 \times 13 \times 8$ cm below the right posterior pararenal space and adjacent to the iliopsoas muscle (Figure 1). Chest radiographs and bone scan showed no evidence of metastatic disease. By extraperitoneal approach, the mass was completely excised. Histology of the tumor was neuroblastoma, which included small round blue cells. Tumor cells were small and hyperchromatic without visible cytoplasm. Pale, pink-staining neurofibrillary stroma was visible in some areas, identifying the tumor as neuroblastoma. This fibrillary material represents neuritic processes, which may form the center of Homer Wright rosettes (Figure 2A). Imunohistochemically, the tumor cells were stained with vimentin (Figure 2B), neuron-specific enolase (NSE), S-100 (Figure 2C) and chromogranin (Figure 2D).

Because the mass was not considered as neuroblastoma preoperatively, metaiodobenzylguanidine (MIBG) scan, urinary homovallinic acid, and vanillylmandelic acid were not studied. However, after the pathological examination confirmed the

Received: September 24, 2007
Accepted: March 19, 2008

Correspondence

Turgut YAPANOĞLU
Department of Urology,
Faculty of Medicine,
Atatürk University,
25240 Erzurum - TURKEY

turgutyapanoglu@mynet.com

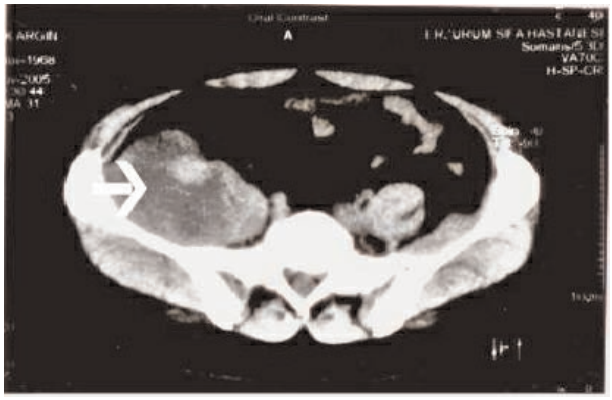

Figure 1. The computed tomographic scan showed a large mass (white arrow) sized as 30 × 13 × 8 cm under the right posterior pararenal space and adjacent of the iliopsoas muscle.

neuroblastoma, MIBG scan showed no abnormal uptake, and a 24-h urine collection done 4 weeks postoperatively revealed normal levels of homovallinic acid and vanillylmandelic acid. So, the patient was diagnosed with stage I disease. The patient was discharged from hospital on postoperative day 5 and followed for 18 months.

Discussion

Neuroblastoma affects children in the first year of life and is very rare among adolescents and young adults (1). Adolescents more frequently have an advanced stage of the disease, a low excretion of urinary catecholamines, absence of biological abnormalities and chronic but fatal disease (3). The prognosis of the patients with neuroblastoma depends on numerous variables; the most important of them are age of patient at diagnosis and stage of the tumor. The low age at diagnosis and the low stage are 2 important favorable prognostic features (4).

Neuroblastomas can metastasize widely through both lymphatic and vascular routes. The most common sites of metastasis are bone marrow, bones, lymph nodes, liver, skin, and testes. There is a greater frequency of metastases among adolescents in unusual parenchyma of the lung or brain, which are affected more frequently in other malignancies that are typical among adults. MIBG scans have high specificity for detecting metastases, but less specificity for detecting central nervous system (CNS) metastasis. CNS metastases may be difficult to diagnose

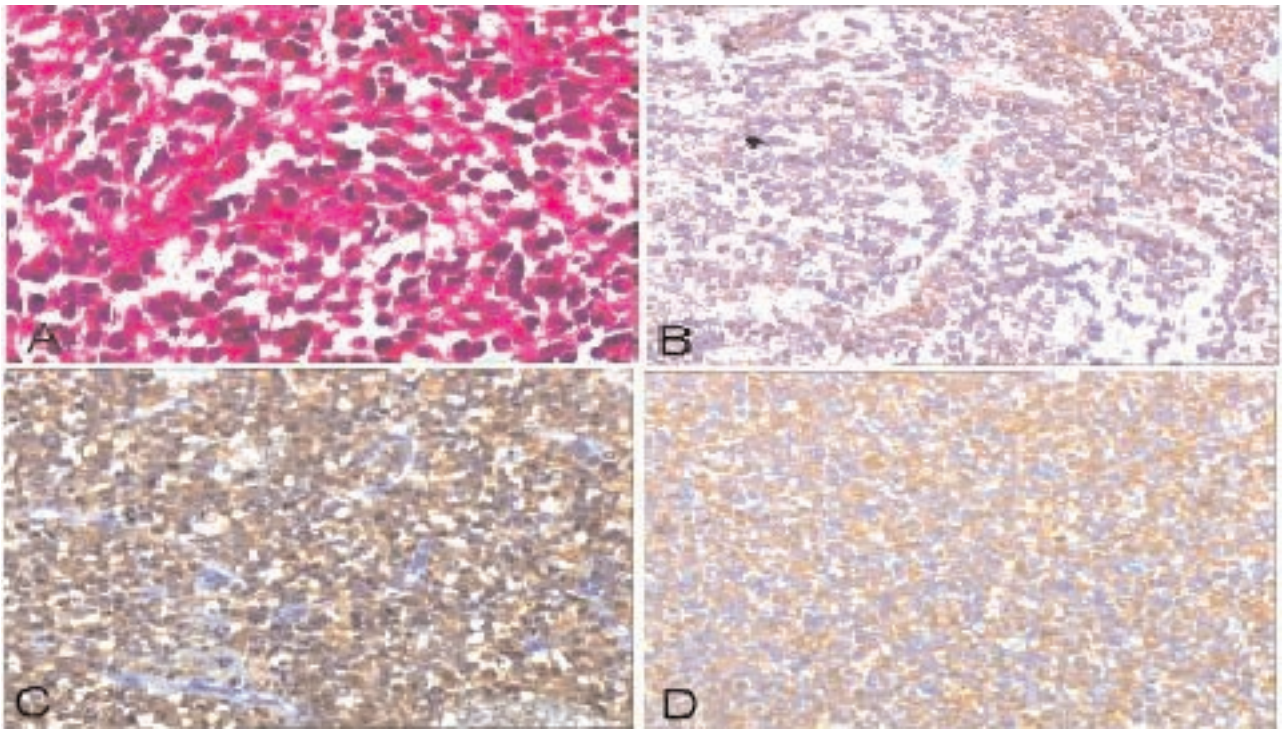

Figure 2. Various hematoxylin-eosin and immunohistochemical staining of neuroblastoma
(A) Neuroblastoma. Note the rosettes (H&E ×200),
(B) Immunohistochemical staining of neuroblastoma cells, showing diffuse immunoreactivity to chromogranin,
(C) Immunohistochemical staining of neuroblastoma cells, showing diffuse immunoreactivity to S-100,
(D) Immunohistochemical staining of neuroblastoma cells , diffuses immunoreactivity to vimentin.

clinically, because they may remain silent for a long period of time. Therefore, particularly when dealing with adolescents who have neuroblastoma, a careful neurological evaluation almost always should be performed to determine CNS metastasis (4).

Although currently there are no standard treatment guidelines for patients with neuroblastoma in adulthood (5), complete surgical resection is sufficient for low risk and non-metastatic tumors, but combination therapy with chemotherapy and radiotherapy is necessary for disseminated and recurrent diseases. We did not perform chemotherapy or radiotherapy, because our patient had a low risk and no distant metastasis.

In conclusion, we suggest that although they are rare in adulthood, neuroblastoma should be kept in mind with almost all cases of suprarenal masses, and if pathological examination confirmed the diagnosis of neuroblastoma, the treatment should be performed according to the pediatric guidelines because no treatment recommendations exist for the treatment of neuroblastoma in adults.

References

1. Menegaux F, Olshan AF, Reitnauer PJ, Blatt J, Cohn SL. Positive association between congenital anomalies and risk of neuroblastoma. Pediatr Blood Cancer 2005; 45: 649-655
2. McLean TW, Iskandar SS, Shimada H, Hall MC. Neuroblastoma in an adult. Urology 2004; 64: 1232.
3. Conte M, De Bernardi B, Milanaccio C, Michelazzi A, Rizzo A, Montobbio G et al. Malignant neuroblastic tumors in adolescents. Cancer Lett 2005; 228: 271-274.
4. Conte M, Parodi S, De Bernardi B, Milanaccio C, Mazzocco K, Angelini P et al. Neuroblastoma in adolescents: the Italian experience. Cancer 2006; 106: 1409-1417.
5. Franke A, Jentsch-Ullrich K, Schalk E, Mohren M, Koenigsmann M, Buhtz P Metastatic adrenal neuroblastoma in an adult. Onkologie 2005; 28: 353-355.