An adult adrenal neuroblastoma presented as a right shoulder pain: A case report

Azzawi M. Hadi,. Idreiss J. Khalaf, Yasser I. Abbass.
Dept. of Surgery, College of Medicine, Tikrit University

Abstract

Neuroblastoma of the adrenal gland is an extremely rare tumor in adulthood although it is one of the most common malignancies in childhood. In this report, a 25-year-old woman who had a left adrenal mass on preoperative imaging. She was operated and the mass was resected in en-block manner along with the left kidney. The histopathological examination of the specimen revealed the diagnosis of neuroblastoma. She had metastasis to the cervical part of the spinal cord operated upon before the time of diagnosis as a suprarenal tumor and received chemotherapy after the operation. Although neuroblastoma of adrenal gland is rare in adulthood, it should be considered in the differential diagnosis for patients with adrenal masses.

Key words: Neuroblastoma, adrenal gland.

Introduction

Neuroblastoma is the most common tumor in infants less than one year of age, the third most common tumor of childhood and more than 90% of all cases occur in children less than 10 years of age (1-2). If some cases occur in adolescence period, it has been very occasionally reported in adults.

The most common locations for primary disease in adults are abdomen, followed by the thorax, pelvis and occasionally head and neck and are very similar to the locations of primary disease in children (1-3). Neuroblastoma of the adrenal gland in adults is an extremely rare disorder and only 25% of the neuroblastoma cases arise from the adrenal glands, although abdomen is the most common site of origin (75%) in adulthood (3). Because of the rarity of adult adrenal neuroblastoma, little is known about its clinicopathological features and imaging findings, hence differential diagnosis of neuroblastoma from the other adrenal masses represents a major difficulty in the preoperative period. Herein, we present a case of adrenal neuroblastoma in an adult patient with advanced disease.

Case report

A 25-year old woman presented with weight loss and right shoulder pain and episodes of low grade fever during the last three months. The physical examination shows pale and tired lady pulse rate was 84 beat per minute and blood pressure was 130/80 mmHg. The routine laboratory tests were within normal limits other than hemoglobin was 90gm/dl high ESR 110mm/hr. This pain responds poorly to systemic NSAIDs and local steroids injection. Cervical spine and chest X-ray was normal, but MRI of the cervical spine shows spinal cord lesion which was excised (fig 1-2). The histopathological examination reveals a case of neuroblastoma.

Two months later she consults a physician for abdominal discomfort and fullness of the left hypochondrial area. On examination there was a hard mass of smooth outline in that area. Abdominal ultrasonography revealed a solitary retroperitoneal lesion with dimensions of 125x90 mm, in the left upper quadrant, adjacent to the upper pole of the left kidney.

Computed tomography (CT) showed a complex, heterogeneous 125x70x90 mm mass arising from a left suprarenal position with no demonstration of a normal adrenal gland and was interpreted as an adrenal tumor which displaced the left kidney inferiorly and invades its upper pole and the tail of pancreas anteriorly. The mass was lobulated and had a clear cut border from the surrounding structures except the borders between the mass and the upper pole of left kidney were not clear (Fig.3).

The left suprarenal gland was explored through a left thoracoabdominal incision. Peroperatively a huge, lobulated tumor located in the left adrenal gland just inferior-posterior to the tail of pancreas with well defined borders was found. The mass infiltrate the upper pole of the left kidney, so that the left adrenal and kidney were removed in mass (Fig.4). There was no lymph node involved in the field. Histopathological examination of the mass showed neuroblastoma. It originated from the left adrenal gland. The aim of the surgery was for de-bulking of the tumor to help the chemotherapy.

She received chemotherapy postoperatively. Six cycles of chemotherapy were given at three weeks interval and as a changing sequence of slow intravenous infusion of cisplatin (100 mg/m2) at first day and intravenous injection of vincristin (1.5 mg/m2) at first and fifth days and 24-hour infusion of ifosfamide (2 g/m2) at first day and intravenous injection of vincristin (1.5 mg/m2) at first and fifth days. Although she had nausea, vomiting, some weight loss and some degree of bone marrow depression during the chemotherapy. She presented latter with better general condition at the last follow up visit.

Discussion

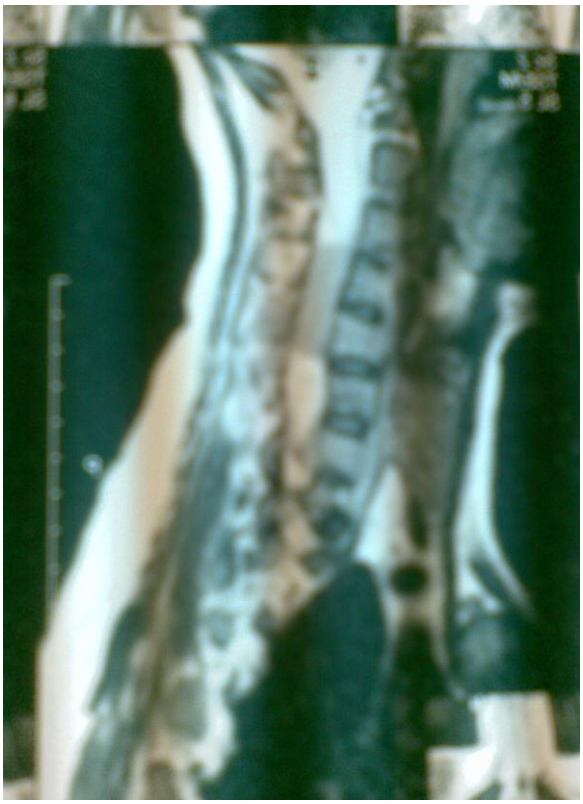
Because of the rarity of adult adrenal neuroblastoma, little is known about its clinicopathological features and imaging findings. The clinical presentation of the disease varies and usually depends on the size, site of tumor and involvement of the adjacent organs or metastatic sites, but may occur without any specific symptoms as occurred in this case. Biochemical studies demonstrated that 85 to 90 percent of neuroblastoma patients have elevated catecholamine metabolites in urine but hypertension in those patients is rare (4). Although the computed tomography (CT) scan is a very useful technique in making the diagnosis and in determining resectability in adrenal masses, MRI is increasingly used because it can reveal tissue-specific characteristics which allow the examiner to differentiate metastases and primary tumors from benign solitary and cystic masses. It is as sensitive as CT scanning in terms of assessing tumor size and resectability ; moreover it has the added advantage of being superior to CT in assessing vessel encasement, vessel patency, spinal cord compression, in showing tumors thrombus in the inferior vena cava and also up to the right atrium (5, 6). In our case MRI defined the borders of the lesion better than CT. Elevated urinary catecholamine metabolites urged us to focus primarily on pheochromocytoma despite the lack of hypertension, a great proportion of neuroblastoma patients have also elevated catecholamine metabolites in urine. There is not enough data in the English literature concerning the catecholamine metabolite level in adrenal neuroblastoma of adulthood. Complete surgical resection whenever possible is the most effective therapy. The role of chemotherapy in neuroendocrine tumors is difficult to assess because of their rarity and variations in biological behavior (7). Chemotherapy adjunct to surgery in neuroblastoma patients is recommended in patients with locally advanced disease. The most active agents are cyclophosphamide, cisplatin, doxorubicin, carboplatin and ifosfamide (8). The usefulness of postoperative radiotherapy was limited in the patients who had microscopic residual disease (1). The disease had had an aggressive course even though complete surgical resection and adjuvant chemotherapy. It has been reported that the neuroblastoma in adults may have a very different outcome from that in paediatric patients (3). Since the disease is rare in adults, but in some cases survival may be worse than for younger patients, even in patients with localized disease (3, 9-11).

As a result, adrenal neuroblastoma in adulthood is a very rare disease but should be considered in the differential diagnosis of atypical adrenal masses. Urinary metabolites of catecholamine may be elevated in adrenal neuroblastoma. Both CT and MRI of abdomen are very useful imaging modalities in the assessment of resectability in adrenal masses including adrenal neuroblastoma (10).

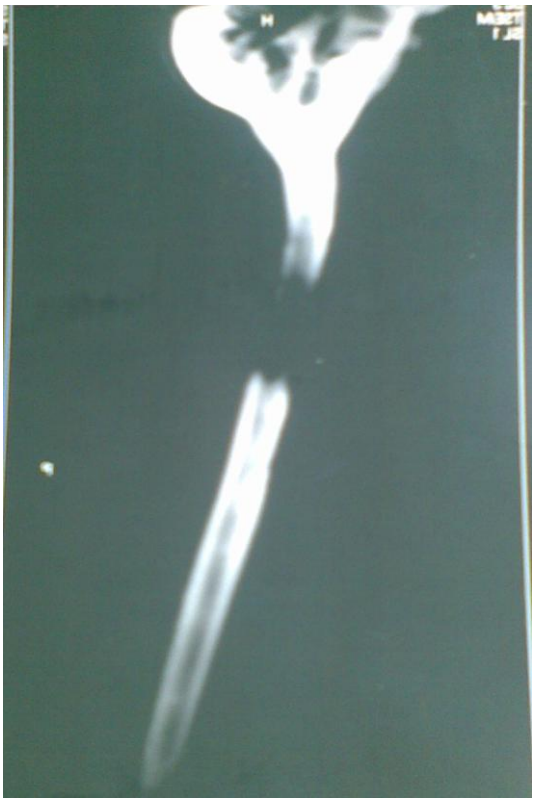
References

1. Albanese C.T. Pediatric surgery. *In*: Norton J. A., Bollinger R. R., Chang A. E. *et*

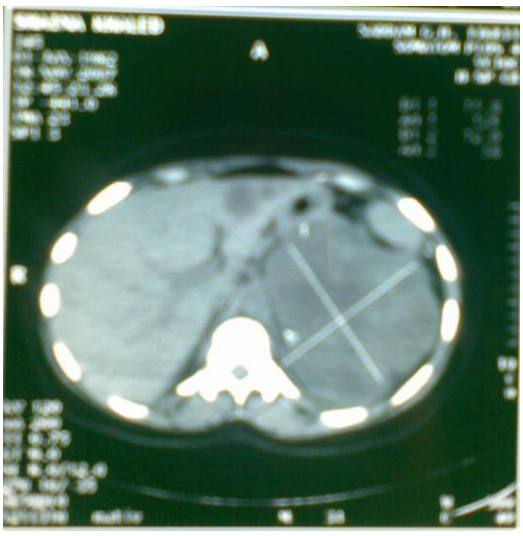
al. (eds.). Surgery : Basic andclinical evidence. 1st ed. New York. Springer, 2001: 2061-94.
2. Albanese C.T., Syndorak R.M., Harrison R.M. Paediatric surgery. *In* : Wayl W , Doherty G.M. (eds.). Current Surgical Diagnosis and Treatment. 11th ed. New York. McGraw-Hill, 2003: 1293-1299.
3. Franks L.M., Bollen A., Seeger R.C. *et al*. Neuroblastoma in adults and adolescents : an indolent course with poor survival. *Cancer*, 1997, **79** : 2028-35.
4. Gann D.S., Demaria E. J. Adrenal. *In*: Schwartz S. I., Shires T. G., Spencer F. C. (eds.). Principles of surgery. 6th ed. New York. McGraw-Hill, 1994:1561-1609.
5. Zielke M. Q., Reed N. S. Adrenal imaging procedures. *In* :Clark O. H., Duh Q. Y. (eds.). Textbook of endocrine surgery.1st ed. Pennsylvania. W.B. Saunders, 1997: 4446-74.
6. Custadio C. M., Semelka R. C., Balci N. C. *et al*. Adrenal neuroblastoma in an adult with tumor thrombus in the inferior vena cava. *J Magn Reson Imaging*, 1999, **9** : 621-3.
7. Hatton M. Q., Reed N. S. Chemotherapy for neuroendocrinetumors : the Beatson oncology centre experience. *Clin Oncol*, 1997, **9** : 385-9.
8. Genc F. A., Aksoy M., Kapran Y. *et al*. Adrenal neuroblastoma in an adult: Report of a case. *Surg Today*, 2003, **33** : 879-81.
9. Aleshire S. L., Glick A. D., Cruz V. E., Bradley C. A., Parl F. F. Neuroblastoma in adults. Pathologic findings and clinical outcome. *Arch Pathol Lab Med*, 1985, **109** : 352-6.
10. Lopez R., Karakousis C., Rao U. Treatment of adult neuroblastoma. *Cancer*, 1980, **45** : 840-4.
11. Prestidge B. R., Donaldson S. S. Treatment results among adults with childhood tumors : a 20-year experience. *Int J Radiat Oncol Biol Phys*, 1989, **17** : 505-14.



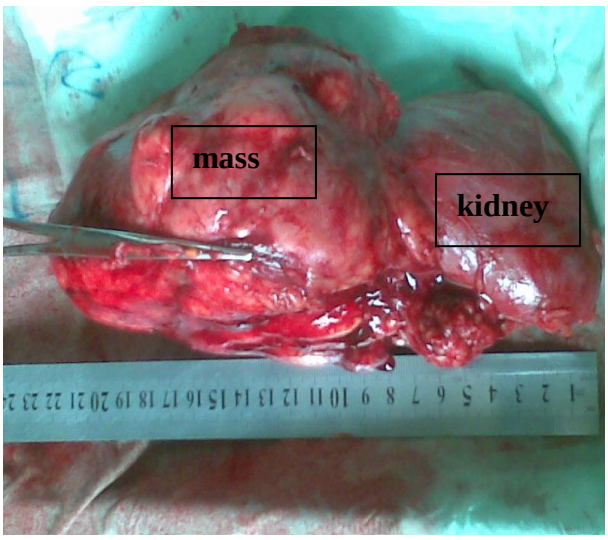
Fig(1)MRI of the cervical spinal cord.



Fig(2)MRI of the cervical spinal cord.



Fig(3) spiral CT of the abdomen shows irregular non-homogenous mass of left suprarenal gland



Fig(4) Suprarenal mass attached to the upper pole of the left kidney removed completely.