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Right adrenal gland neuroblastoma infiltrating the liver and mimicking mesenchymal hamartoma: A case report

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ABSTRACT

INTRODUCTION: Neuroblastoma is the most common extracranial solid pediatric malignancy. The most common site is abdomen with predominance of suprarenal medulla. Infiltration of the tumour to the liver is rare. No cases were reported in the literature about the misdiagnosis of neuroblastoma as mesenchymal hamartoma in the liver.
PRESENTATION OF CASE: We represent a rare case of neuroblastoma misdiagnosed as mesenchymal hamartoma in liver in a six-month-old female infant presented with fever and abdominal mass. Abdominal computed tomography (CT) revealed large cystic lesion occupying most of the right liver enchroaching upon right suprarenal region and displacing the right kidney inferior suggestive for mesenchymal hamartoma. Right adrenalectomy with en-bloc resection of the adjacent liver segments was done. Postoperative pathology revealed neuroblastoma with positive specific immunohistochemistry (IHC).
DISCUSSION: Although neuroblastoma is the second most common pediatric abdominal malignancy with specific diagnostic modalities, a misdiagnosis of a case with neuroblastoma as mesenchymal hamartoma is rare. Histopathological diagnosis of neuroblastoma with positive IHC is essential as shown in our case.
CONCLUSION: We represent a rare case of neuroblastoma which arose from the right adrenal gland and infiltrated the adjacent liver substance mimicking mesenchymal hamartoma of the liver. Neuroblastoma is rarely presented with pyrexia of unknown origin. Neuroblastoma should be considered in differential diagnosis of abdominal mass in all infants and children.

1. Introduction

Neuroblastoma (NB) is the most common extracranial solid pediatric malignancy representing 8–10% of all childhood malignancies. It is an embryonal tumor which originates from the sympathetic nervous system [1]. NB has been described as an extremely heterogeneous because of its highly variable biologic behavior [2], ranging from spontaneous regression to a progressive course with fatal outcome [1,3]. About 90% of patients are diagnosed with neuroblastoma before the age of 5 years (median age at diagnosis of 18 months) [4]. Nearly 65% of tumours arise in the abdomen and they are usually localized to the medulla of the adrenal gland [5]. Most patients are symptoms-free, but some may present with constitutional symptoms (malaise, fever, and weight loss), an enlarging mass, pain, abdominal distention, lymphadenopathy, or respiratory embarrassment secondary to compression or hepatomegaly [6]. Biochemical diagnostic findings include increased levels of serum or urine catecholamines or its metabolites and/or nonspecific biomarkers such as lactate dehydrogenas, ferritin, and neuron-specific enolase that may be associated with advanced stage or prolapse [7]. For diagnostic imaging, computed tomography (CT) scan and/or magnetic resonance imaging (MRI) are the gold standard for detection of the tumour site, size and degree of involvement [8]. Histopathological examination with specific immunohistochemistry is essential for final diagnosis. The standard lines of treatment in management of neuroblastoma include chemotherapy, radiotherapy and/or surgical resection [9]. Neuroblastoma arising from suprarenal medulla infiltrating the right liver with radiological finding suggestive for mesenchymal hamartoma is rare. We report a case of neuroblastoma infiltrating the right liver misdiagnosed as mesenchymal hamartoma.

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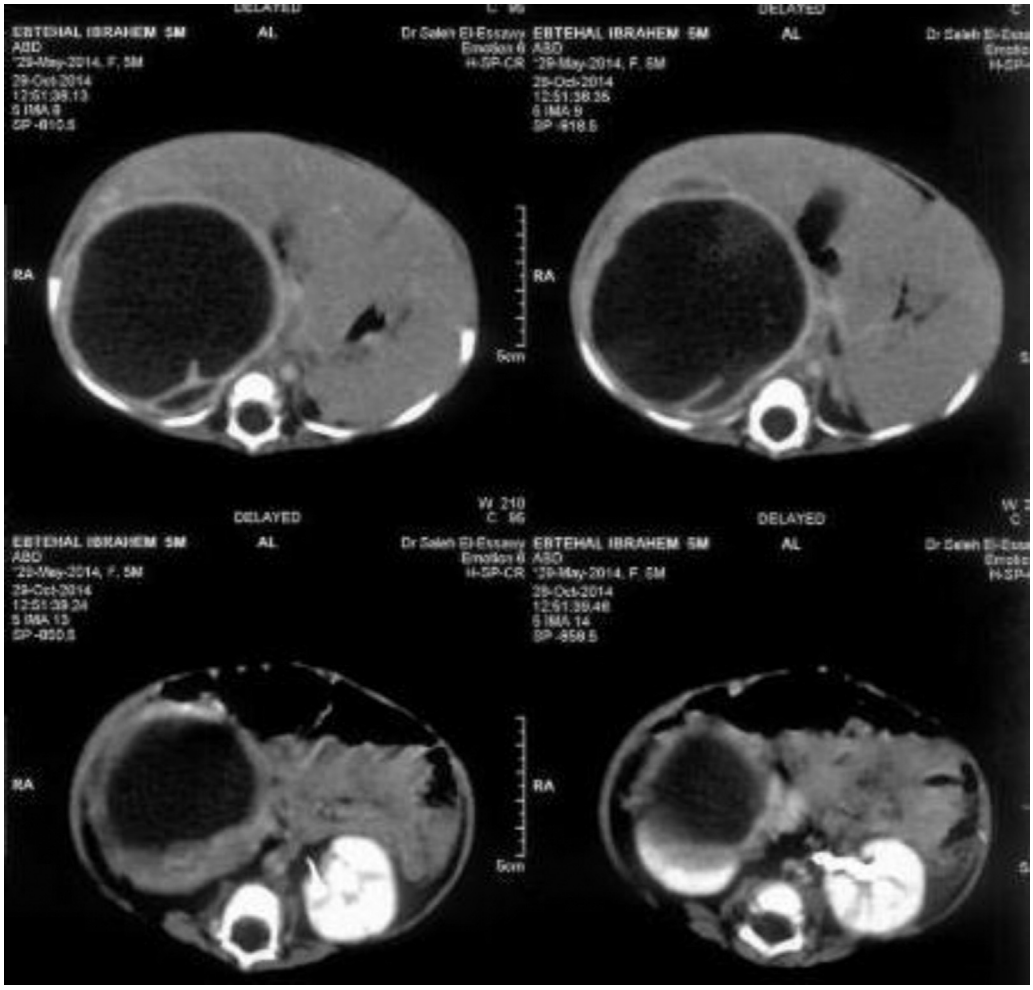

Fig. 1. CT abdomen shows large well defined, non-enhanced hypodense cystic lesion occupying most of the right liver lobe (segment V,VI,VII) measuring (8 × 8 × 8.3 cm) with exophytic component enchroaching upon right suprarenal region and displacing the right kidney inferior suggestive for mesencymalhamartoma.

1.1. Presentation of case

A six-month-old female infant was referred to our center by right liver mass mostly mesenchymal hamartoma on radiological basis for surgical intervention. She was admitted in children hospital for pyrexia of unknown origin with no response to antibiotics and antipyretics. Fever was associated with right hypochondrial pain and abdominal enlargement without jaundice for one month. Perinatal history was uneventful. On examination, there was right hypochondrial tenderness and hepatomegaly with liver span equals 8 cm (normal liver span for this age is 5–5.5 cm). Laboratory investigations showed microcytic hypochromic anemia (Haemoglobin = 8.4 gm/dl), leukocytosis (White blood cells' count = 13.1×10^3), thrombocytosis (Platelets = 620×10^3), slightly elevated alpha-feto protein (AFP) (AFP = 26.6 ng/ml) and normal liver function tests (LFTs). Abdominal ultrasonography (US) showed well-defined heterogeneous mass with cystic degeneration and foci of calcification affecting most of the right liver measuring about 7.4 × 8.4 cm. Abdominal computed tomography (CT) with contrast showed large well defined, non-enhanced hypodense cystic lesion occupying most of the the right liver (segment V,VI,VII) measuring (8 × 8 × 8.3 cm) with exophytic component enchroaching upon right suprarenal region and displacing the right kidney inferiorly with no enlarged abdominal lymph node (LN). The mass was highly suggestive for the right liver mesenchymal hamartoma as shown in (Fig. 1). No liver biopsy was taken. The provisional diagnosis was mesenchymal hamartoma or hepatoblastoma for surgical resection and histopathological confirmation. The mother of the infant underwent informed written consent for surgery.

On exploration, there were no signs of peritoneal seeding or involvement of adjacent organs. The liver was completely healthy with large mass mainly in segment VI and VII adherent to transverse colon with displacement of the right kidney downward. After surgical assessment of the tumor, Right adrenalectomy with en-bloc resection of the adjacent liver segments was performed with safety margin about 1 cm of liver tissue after dissection of the mass from the right renal vessels. No residual tumour tissue was left (R0).

1.2. Postoperative pathology

Grossly the mass was friable measuring about 9 × 8 × 8 cm. Cut section revealed large areas of hemorrhage and necrosis with infiltration of adjacent liver tissue. Microscopically the tumor cells are small with hyperchromatic nuclei and scanty cytoplasm, and are arranged in sheets separated by thin fibro-vascular septa between the nests of tumor cells . Few ganglion cells 9.5% of tumor population are detected. There are wide areas of necrosis. No evidence of Homer Wright rosettes or pseudorosettes was detected. Surgical margins were free from tumor infiltration as shown in Figs. 2–5.

Postoperative course was uneventful and the patient was discharged after 6 days in good general condition. The patient was

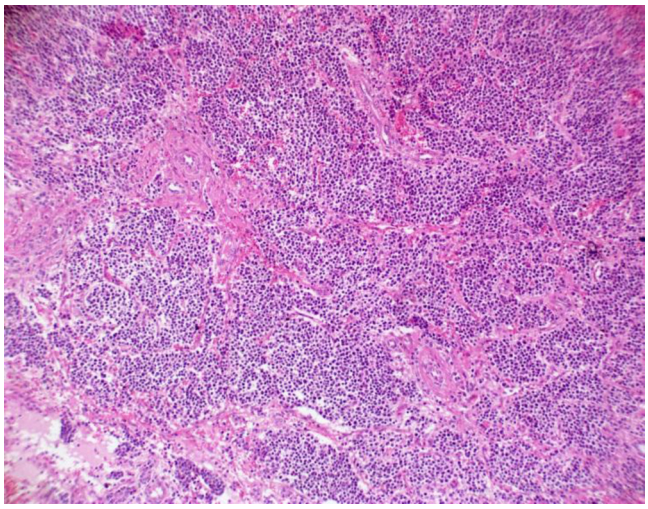

Fig. 2. The tumor is formed of small cells separated by fibro-vascular septa (H&E staining 100×).

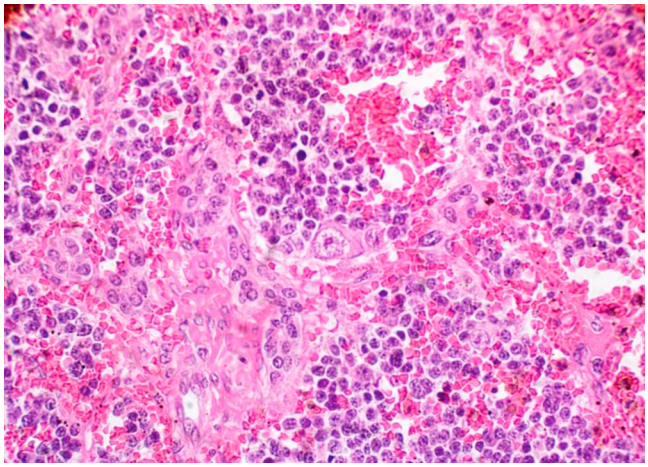

Fig. 3. Malignant small cells with ganglion cell differentiation (arrow) (H&E staining 400×).

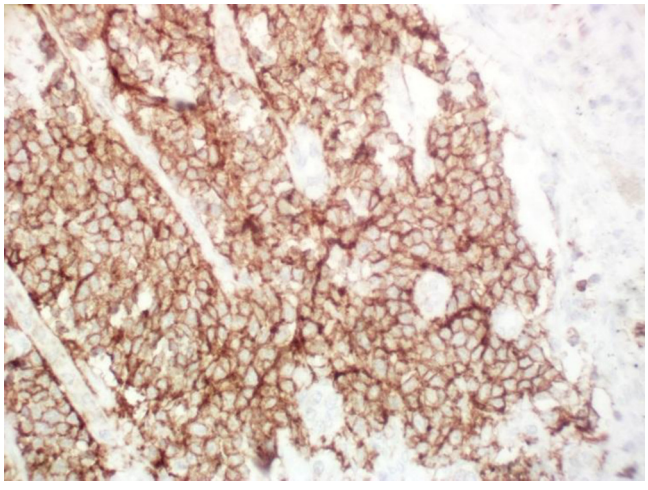

Fig. 4. Positive membranous reaction of CD56 immunohistochemistry (400×).

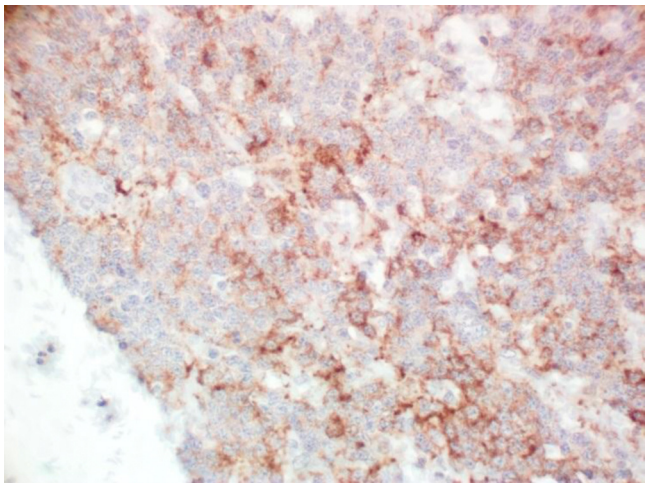

Fig. 5. Positive cytoplasmic reaction of chromogranin A immunohistochemistry (400×).

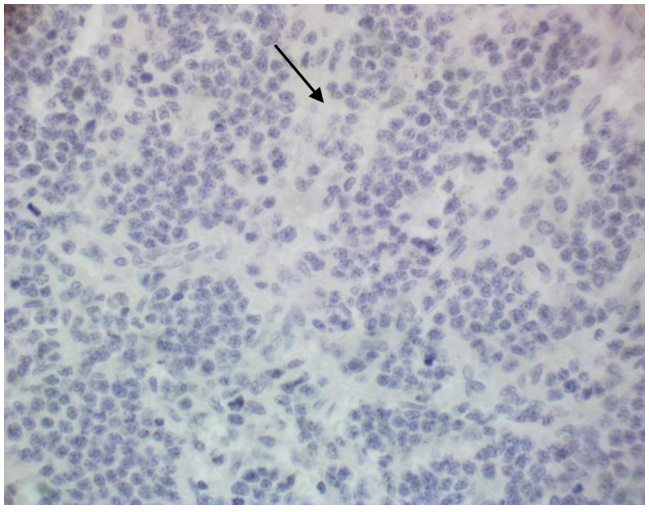

Fig. 6. Negative reaction of HepPar-1 immunohistochemistry (400×).

referred to pediatric oncology department for receiving adjuvant chemotherapy in the form of cisplatin and adriamycin Fig. 6.

1.3. Discussion

Neuroblastoma is the most common extracranial solid pediatric malignancy representing 8-10% of all childhood malignancies. It is an embryonal tumor arises from the sympathetic nervous system. It accounts for approximately 15% of all cancer-related deaths in the pediatric population [1]. Neuroblastoma has been described as an extremely heterogeneous because of its highly variable biologic behavior [10], ranging from spontaneously regression to a progressive course with ultimate fatal outcome [1,3]. Our case was six-month-old infant at the time of diagnosis which is a common age for diagnosis of neuroblastoma as reported in literature [4]. More than 50% of patients with neuroblastoma are diagnosed as high-risk neuroblastoma with features including large unresectable and widely metastatic tumour resulting in poor prognosis as regard long-term survival [11]. Nearly 65% of tumours arise in the abdomen with half of those localized to the medulla of the adrenal gland [12] as reported in our case. In our report, the girl presented by fever of unknown origin for 2 weeks followed by abdominal enlargement and pain as reported in previous studies [6]. Biochemical diagnostic tests of neuroblastoma include serum or urinary

level of catecholamines or its metabolites (dopamine, vanillylmandelic acid, and homovanillic acid) and/or nonspecific biomarkers such as lactate dehydrogenase, ferritin, and neuron-specific enolase [7]. None of these laboratory tests were done preoperatively because the liver mass was not suspicious of neuroblastoma on radiological basis. For diagnostic imaging, abdominal CT scan of the abdomen is the gold standard for detection of the tumour site and the degree of involvement. A magnetic resonance imaging is more sensitive in cases of spinal extention [8]. In our case, the abdominal CT revealed large cystic lesion in the right liver just displacing the right kidney and the right suprarenal gland and it was highly suggestive for mesenchymal hamartoma which is classic for it as reported in the literature [13], while radiologic findings of neuroblastoma include focal or multifocal solid tumor with calcification in 40%–50% of cases, which is not essential for diagnosis [14]. Other diagnostic tests include, but not done in our case, iodine-131-meta-iodobenzylguanidine (MIBG) which is useful in detection of both primary tumour and metastasis but not widely used in clinical practice and flurodeoxyglucose positron emission tomography (FDG-PET) which maybe valuable in localizing soft tissue metastasis [15]. Confirmation of the diagnosis is confirmed pathologically by tissue biopsy either by complete resection or open biopsy in case of unresectable tumours and/or BM aspiration [16]. No preoperative US or CT guided biopsy from the mass was done to avoid dissemination and seeding of the tumour cells as hepatoblastoma was considered in the differential diagnosis of our case.

The standard lines of treatment in management of neuroblastoma include chemotherapy, radiotherapy and/or surgical resection. Many new promising therapies were introduced based on molecular mechanisms for differentiation, cell survival and apoptosis [17]. The surgical resection offers definitive therapy with excellent outcome for management of locoregional low-risk neuroblastoma [18], while high-risk patients are treated with intensive multimodality therapies. In our case, the decision was surgical resection from the start if the tumour was resectable for final pathological diagnosis as the provisional diagnosis was whether hepatoblastoma or mesenchymal hamartoma.

2. Conclusion

We represent a rare case of right adrenal gland neuroblastoma infiltrated the adjacent liver segments mimicking mesenchymal hamartoma of the liver. Neuroblatoma is rarely presented with pyrexia of unknown origin. Neuroblastoma should be considered in differential diagnosis of abdominal mass in all infants and children.

Conflict of interest

Prof. Ahmed Abu-elenen and other co-authors have no conflict of interest.

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Ethical approval

Local ethical board committee of the Gastroenterology Center (GEC), Mansoura University.

Consent

The patient underwent informed written consent of the surgery and publication.

Guarantor

None.

Author contributions

All authors have made substantial contributions to all of the following: (1) acquisition of data, analysis and interpretation of data, (2) revising it critically for important intellectual content and (3) final approval of the version to be submitted.

References

[1] J.R. Park, A. Eggert, Caron H. Neuroblastoma, biology, prognosis and treatment, Pediatr. Clin. North. Am. 55 (2010) 97–120.
[2] C. Spix, G. Pastore, R. Sankila, C.A. Stiller, E. Steliarova-Foucher, Neuroblastoma incidence and survival in European children (1978–1997): report from the automated childhood cancer information system project, Eur. J. Cancer 42 (13 (September)) (2006) 2081–2091.
[3] A. Salim, D. Mullassery, B. Pizer, H.P. McDowell, Losty P.D. Neuroblastoma, a 20-year experience in UK regional centre, Pediatr. Blood Cancer 57 (7 (December 15)) (2011) 1254–1260.
[4] K.K. Matthay, J.G. Villablanca, R.C. Seeger, D.O. Stram, R.E. Harris, N.K. Ramsay, et al., Treatment of high-risk neuroblastoma withintensive chemotherapy radiotherapy autologous bone marrow transplantation and 13-*cis*-retinoicacid Children's Cancer Group, N. Engl. J. Med. 341 (16 (October 14)) (1999) 1165–1173.
[5] G.M. Brodeur, J.M. Maris, Neuroblastoma, in: P.A. Pizzo, D.G. Poplack (Eds.), Principles and Practice of Pediatric Oncology, 4th ed., Lippincott Williams and Wilkins Co., Philadelphia, 2002, pp. 895–937.
[6] G.M. Haase, M.P. La Quaglia, Neuroblastoma, in: M.M. Ziegler, R.G. Azizkhan, T.R. Weber (Eds.), Operative Pediatric Surgery, McGraw-Hill, New York, 2003, pp. 1181–1192.
[7] S. Kim, D.H. Chung, Pediatric solid malignancies: neuroblastoma and Wilm's tumour, Surg. Clin. North Am. 86 (2 (April)) (2006) 469–487, xi.
[8] N.C. Colon, D.H. Chung, Neuroblastoma, Adv. pediatr. 58 (1) (2011) 297–311.
[9] A.M. Davidoff, Neuroblastoma, Semin. Pediatr. Surg. 21 (1) (2012) 2–14.
[10] G.M. Brodeur, A. Nakagawara, Molecular basis of clinical heterogenesity in neuroblastoma, Am. J. Pediatr. Hematol. Oncol. 14 (2 (May)) (1992) 111–116.
[11] S.G. DuBois, Y. Kalika, J.N. Lukens, G.M. Brodeur, R.C. Seeger, J.B. Atkinson, et al., Metastatic sites in stage IV and IVS neuroblastoma correlate with age, tumour biology, and survival, J. pediatr. Hematol. Oncol. 21 (3 (May–June)) (1999) 181–189.
[12] Kushner B.H. Neuroblastoma, A disease requiring a multidude of imaging studies, J. Nucl. Med. 45 (7 (July)) (2004) 1172–1188.
[13] M.D. Stringer, N.K. Alizai, Mesenchymal hamar toma of the liver: a systematic review, J. Ped. Surg. 40 (11 (November)) (2005) 1681–1690.
[14] C.E. Herzog, R.J. Andrassy, F. Eftekhari, Childhood can cers: hepatoblastoma, Oncologist 5 (6 (December)) (2000) 445–453.
[15] D.R. Taggart, M.M. Han, A. Quach, S. Groshen, W. Ye, J.G. Villablanca, et al., Comparison of iodine-123 metaiodobnzylguanidine (MIBG) scan and (18f) Flourodeoxy glucose positron emission tomography to evaluate response after iodine-123 MIBG therapy for relapsed neuroblastoma, J. Clinoncol. 27 (32 (November 10)) (2009) 5343–5349.
[16] L. Wood, S. Lowis, An update on neuroblastoma, Pediatr. Child Health 18 (3 (March)) (2008) 123–128.
[17] G. Cecchetto, V. Mosseri, B. De Bernardi, P. Helardot, T. Monclair, E. Costa, et al., Surgical risk factors in primary surgery for localized neuroblastoma: the LNESG1 study of the European International Society of Pediatric Oncology Neuroblastoma Group, J. ClinOncol. 23 (33 (November 20)) (2005) 8483–8489.
[18] H.J. Conter, V. Gopalakrishnan, V. Ravi, J.L. Ater, S. Patel, D.M. Araujo, Adult versus pediatric neuroblastoma: the MD, Anderson Cancer Center experience, Sarcoma 2014 (2014) 371515.