Cystic neuroblastoma in an older child

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ABSTRACT

Cystic neuroblastoma presenting beyond the neonatal period is rare and seems to have a different presentation and outcome as compared to those diagnosed in the antenatal and perinatal period. Presented herein is a case of cystic neuroblastoma diagnosed in a 9-year-old girl who had a poor outcome despite complete excision, aggressive chemotherapy and radiation therapy.

KEY WORDS: Cystic neuroblastoma, outcome, postnatal diagnosis

Cystic neuroblastoma is a rare clinical entity with most of the cases being diagnosed antenatally or perinatally. Very few cases of cystic neuroblastoma diagnosed beyond the perinatal period have been reported. All these have been in children less than 2 years of age. Presented herein is a case of cystic neuroblastoma diagnosed in a 9-year-old girl who had a poor outcome despite complete excision, aggressive chemotherapy, and radiation therapy.

CASE REPORT

A 9-year-old girl presented with complaints of a vague right upper abdominal pain of 1-month duration, and a lump in the right side of the abdomen noticed 15 days ago. There was no history of anorexia or loss of weight. Examination revealed a large smooth cystic mass in the right hypochondrium extending to the epigastrium and the right lumbar regions. The mass was moving with respiration, it was fluctuant and mildly tender. Ultrasonography revealed a 16 × 15-cm cystic mass, with multiple septations occupying the entire right lobe of the liver [Figure 1]. Serology for echinococcus was reactive. With a clinical diagnosis of hydatid cyst of the liver, a laparotomy was performed; a 16 × 15 × 18-cm extrhepatic cystic mass was found in the right suprarenal location pushing the entire liver on to the left [Figure 2]. The inferior venacava (IVC) was found to be engulfed by the cyst. Approximately 700 ml of thin blood-stained fluid was aspirated from the cyst, which was then excised. There was no lymphadenopathy in the para-caval or para-aortic group of nodes. The contralateral adrenal was normal; the excision was considered complete. No normal adrenal tissue was identified. Histopathtology was returned as neuroblastoma, stroma poor type (Shimada's classification). Immunohistochemistry was positive for neuron-specific enolase, S-100, synaptophysin, and chromogranin and was negative for MIC-2. Postoperatively done alpha fetoprotein, 24-hour urinary HVA and VMA, skeletal survey, bone scan, bone marrow aspiration and bone marrow biopsy were all normal. The disease was staged as...
Stage 2A (INSS staging). The child received five courses of chemotherapy consisting of cyclophosphamide 150 mg/m²/day, days 1-7 and Doxorubicin 60 mg/m² on day 8, at 3-week intervals. A contrast-enhanced CT (CECT) scan done 3 months after completion of this 4 months of chemotherapy was normal. Six months after completion of chemotherapy, the patient again complained of pain in the right upper abdomen and an examination revealed a 7 × 7-cm hard nodular mass with indistinct margins, which was moving with respiration. A CECT scan revealed a solid 10 × 10-cm mass in the right suprarenal area that was engulfing the IVC and displacing it to the left [Figure 3]. The right kidney was displaced inferiorly and anteriorly. A fine needle aspiration cytology of the mass confirmed the recurrence of neuroblastoma and so the child was started on chemotherapy consisting of cyclophosphamide - 150 mg/m²/day - days 1-7 on weeks 1, 3, 6, 13, 17, 21, 29 – given intravenously in week 1 and thereafter orally; cisplatin - 90 mg/m²/day – day 1 on weeks 2, 7, 10, 14, 25, 35; doxorubicin - 35 mg/ m²/day - day 1 on weeks 2, 7, 14, 22, 30; etoposide - 100 mg/m²/day - days 1 and 2 on weeks 4, 10, 18, 25, 33. She also received local radiation therapy (3000 cGy). Despite chemotherapy and radiation therapy, the mass progressively increased in size and so the parents decided to discontinue therapy. The child succumbed to the disease 14 months from the time of diagnosis.

DISCUSSION

Cystic neuroblastoma is an extremely rare type of neuroblastoma and most of the cases have been diagnosed either antenatally or perinatally.[1-5] Only a few cases have been diagnosed beyond the perinatal period.[2,6-10] Most of these cases diagnosed beyond the perinatal period have been adrenal in location.[2,4,7-10] with only a few being extra-adrenal or mediastinal in location.[2,6] All these cases have been in children less than 2 years of age, while the present case is the first report of this entity in a 9-year-old child.

The main differential diagnosis of a cystic supra-renal mass detected on an ultrasound performed antenatally or in a neonate is neuroblastoma, an adrenal hemorrhage or a dilated upper pole renal calyx.[5,11,12] Antenatally or perinatally diagnosed suprarenal masses that are either palpable or are progressively increasing in size are more likely to be neuroblastomas.[5] Common indicators of neuroblastomas like raised urinary excretion of homovanillic acid and vanillylmandelic acid or the presence of calcification are said to be of little help in diagnosing cystic neuroblastomas.[5] Surgical excision of such masses is the only way of obtaining a firm diagnosis of MIBG-negative, nonsecreting tumors.

Antenatally or perinatally diagnosed cystic neuroblastosomas have a uniformly favorable outcome[1-5] owing to their usually favorable stage (Stages 1, 2, or 4S) and biologic features. The same cannot be said about the cases diagnosed beyond the perinatal period. Of the few reported cases in the children, two were autopsy reports,[9,10] two had metastasis at presentation of whom one died[2,3] and another two developed recurrence and died.[2,10]

Presentation at 9 years of age and the cystic nature of the mass reported on ultrasonography to be in the right lobe of the liver, in addition to the serology being reactive for echinococcus, were the reasons for the wrong clinical diagnosis in the present case. As cystic neuroblastomas are rare, only a high index of suspicion supported by careful preoperative investigations will enable a preoperative diagnosis. The number of cases reported in the literature are two few to decide about the optimum treatment but it would probably be safer to err on the side of instituting appropriate chemotherapy after excision in all these children, especially those diagnosed beyond the perinatal period.

REFERENCES