Primary endodermal sinus tumor presenting with spinal cord compression

Case report

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Endodermal sinus tumor is an uncommon malignant germ-cell neoplasm. These tumors usually present in childhood or young adulthood as testicular or ovarian masses; however, mediastinal and intracranial tumors have been described. The authors report the occurrence of a primary paraspinal endodermal sinus tumor in a 21-month-old boy who presented with thoracic spinal cord compression. A review of the literature failed to reveal a similar case. The clinical presentation, radiographic characteristics, operative findings, and patient outcome are discussed.

KEY WORDS • alpha fetoprotein • endodermal sinus tumor • spinal cord compression • spinal tumor • yolk sac tumor • children

Examination. At the time of admission the child was irritable and afebrile. Cranial nerve and upper-extremity neurological function were normal. The child’s lower extremities were hypotonic, and only minimal withdrawal was noted with intense painful stimulation. The weakness was more severe proximally with the quadriceps muscles graded as 1/5 and the gastrocnemius muscle as 3/5 bilaterally. A Babinski reflex was present and was associated with four-to-five beats of ankle clonus bilaterally. Rectal tone was markedly diminished; however, the bulbocavernous reflex was intact.

Magnetic resonance (MR) imaging was performed immediately and demonstrated a large left suprarenal mass with epidural extension and marked spinal cord compression at T9–L1 (Fig. 1).

Operation. The patient was taken immediately to the operating room. An osteoplastic laminotomy was performed at T9–L1 and revealed the epidural tumor. The tumor was extremely vascular and even minimal manipulation provoked severe bleeding. Intraspinal tumor was removed with the ultrasonic aspirator and rongeurs. Tumor specimens were obtained for histopathological review. Postoperatively, the child did not recover lower-extremity function.

Histological Analysis. Histopathological review revealed round-to-oval hyperchromatic nuclei with prominent nucleoli and varying amounts of eosinophilic cytoplasm (Fig. 2). Some of the cells contained hyaline droplets that were periodic acid Schiff-positive and resis-
tant to diastase digestion. The final histopathological diagnosis was endodermal sinus tumor. A metastatic workup (including bone marrow aspirate and MR imaging of the brain) revealed no evidence of disseminated disease. The patient’s serum α-fetoprotein was 85,660 ng/ml (normal range < 20 ng/ml).

The child was diagnosed as having EST Stage III and subsequently entered into CCG protocol 8882 “Germ-Cell Tumor” for chemotherapy with bleomycin, VP-16, and cisplatin.

Postoperative Course. The child completed his first cycle of chemotherapy on postoperative Day 12 and was discharged home on postoperative Day 14. Two days later he became suddenly unresponsive. He was pronounced dead on arrival in the emergency room. An autopsy was performed and revealed the known paravertebral EST without evidence of disseminated disease or residual epidural mass. Postmortem histological examination of the tumor revealed similar findings to those made antemortem, and the final pathological diagnosis remained EST. Vitreous fluid sodium was noted to be elevated (164 meq/L OS and 167 meq/L OD); however serum sodium 3 days prior to death was 132 meq/L. No evidence for pulmonary embolism was found and the cause of death remains unknown.

Discussion

Spinal cord compression from epidural tumor occurs in 2 to 5% of children with malignant systemic tumors. The most frequent tumor types involved are neuroblastoma, Ewing’s sarcoma, rhabdomyosarcoma, and osteogenic sarcoma. Germ-cell neoplasms, including teratoma and embryonal cell carcinoma, are responsible for the compression in less than 5% of reported cases. Treatment for spinal cord compression from these malignant childhood tumors is directed toward rapid decompression of the neural elements, establishment of a tissue diagnosis, and administration of appropriate adjuvant therapy. Endodermal sinus tumor usually presents as a testicular or ovarian mass, although mediastinal, retroperitoneal, presacral, and intracranial primary locations have been described. The tissue of origin for the EST appears to be the extraembryonic mesoderm and endoderm (that is, tissues destined to become the yolk sac and placenta). Extragonadal occurrence of these tumors is thought to be the result of abnormal persistence of germ cells along embryonic migratory pathways. Intracranial occurrence is traditionally limited to the suprasellar cistern and pineal region, although a single case of a cerebellar primary tumor has been described. Systemic ESTs can and do metastasize, and metastases to the brain and spine, although extremely rare, have been described.

Two cases of spinal involvement with metastatic endodermal sinus tumor have been described. One case involved a 62-year-old woman with disseminated EST from a presacral primary tumor. She presented with a conus medullaris syndrome and eventually died of the disease in spite of multiple surgical procedures and chemotherapy. The second case of EST metastatic to the spine causing cord compression was reported by Raffel, et al., in their series of 33 children with spinal cord compression from malignant tumors. No further information about the case was given.

The diagnosis of EST may be inferred from elevated serum α-fetoprotein measurements; however, final diagnosis is dependent on the demonstration of characteristic features by histopathological examination. Although multiple histological types have been described, some common features do exist among the architectural variants. Among these features are Schiller–Duvall bodies, hyaline globules, and immunohistochemical staining for α-fetoprotein.

The mainstays of treatment for EST are cytoreductive surgery followed by chemotherapy, most commonly multiagent chemotherapy including vincristine, actinomycin-D, and cyclophosphamide. The results of therapy in cases

Fig. 1. Axial (left), coronal (center), and parasagittal (right) MR images demonstrating the extent of epidural spread from the supraprenal tumor mass.

Fig. 2. Photomicrograph of tissue demonstrating round-to-oval hyperchromatic nuclei with prominent nucleoli and varying amounts of eosinophilic cytoplasm. Intracytoplasmic hyaline droplets are well demonstrated in this section. PAS, original magnification × 300.
of gonadal EST have been mixed with long-term survival reported in between 0 and 75% of patients, depending on the stage of the tumor and adjuvant therapy used. 3,8,12 However, patients with intracranial disease have consistently done poorly, with three of 18 survivors (range of follow up 7 months–5.5 years) noted in the cases reviewed by Eberts and Ransburg, 6 and three of 19 in the additional cases reviewed by Kirikae, et al. 10 (range of follow up 7.5 months–3.5 years). 6,10 Experience with spinal tumors is obviously limited. Although postmortem examination was unrevealing in this case, three cases of fatal tumor embolism have been reported, as has a case of a spontaneous hemothorax, in patients with EST. 2,9

References

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