Unusual primary secreting germ cell tumor of the spine

Case report

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The authors describe a young man with a rare primary spinal germ cell tumor that secreted β–human chorionic gonadotropin. The tumor was resected, and six courses of adjuvant chemotherapy consisting of cisplatin, bleomycin and etoposide were administered together with irradiation to the craniospinal area. An additional dose of radiation was delivered to the tumor site after the first four chemotherapy sessions. The patient was well without any neurological deficit or iatrogenic sequela 33 months after diagnosis. The occurrence of this rare tumor located primarily in the spine warrants attention in pathological studies of spinal tumors in young patients.

KEY WORDS • spinal tumor • secreting germ cell tumor

GERM cell tumors arising in the central nervous system represent 5% of all childhood and adolescent intracranial malignancies. Of these, secreting germ cell tumors represent approximately 20%. To date a spinal origin of germ cell tumors has been reported in fewer than 20 cases. We describe the sixth germ cell tumor found to secrete β-HCG.

Examination. This 20-year-old previously healthy man complained of lumbar pain and right lower-limb dysesthesia. One month after the symptoms commenced, he noted worsening motor weakness in the lower extremities which resulted 2 days later in paraplegia and associated neurogenic bladder. Spinal MR images (Fig. 1A–C) revealed an intraspinal nodule at L-2 that originated in the ependymal canal and extended 4 cm vertically, occupying the entire vertebral space. The tumor was isointense on T2-weighted and hyperintense on T1-weighted images; the contrast-enhanced image was heterogeneous. The patient was admitted with paraparesis and more distally paralysis, and bilateral hypesthesia was present from L-3 to his feet.

Operation. The patient underwent an emergency L1–3 laminectomy. The dura mater was made taut by the underlying tumor, which protruded immediately after the incision had been made. With the aid of an operating microscope it was possible to remove an intradural, soft, hypervascular, hemorrhagic, brown nodule originating in the ependymal canal at the distal apex of the conus medullaris. The tumor infiltrated the terminal filum and displaced the caudal roots upward. All the nerve roots were spared and the tumor was totally resected. One week after the operation only mild bilateral motor weakness remained. The original histological diagnosis was anaplastic ependymoma. The patient was then referred to our institution where neuropathological investigation was performed.

Pathological Findings. Histologically the tumor was largely composed of solid areas of cytotrophoblastic mononuclear cells with intermingled syncytiotrophoblastic giant cells (Fig. 2 upper). In some areas the neoplasm was composed of immature stromal tissue with a myxoid background containing an immature fetal-type gland. Immunohistochemical staining was intensely and diffusely immunoreactive for β-HCG in both the syncytiotrophioblastic components (Fig. 2 lower). However, the latter stained focally positive for keratin and placental alkaline phosphatase. Alpha-fetoprotein was not detected. A diagnosis of a mixed nonseminomatous germ cell tumor consisting of a choriocarcinoma and an immature teratoma was made.

Postoperative Course and Treatment. A thorough imaging evaluation was performed consisting of thoracoabdominal computed tomography scanning, testicular ultrasono-
raphy, and MR imaging of the entire central nervous system; none of the examinations revealed a different primary tumor site or residual disease. The β-HCG level was 16.84 mIU/ml in serum and 223.39 mIU/ml in the cerebrospinal fluid; the α-fetoprotein value was 6.99 and 13 IU/ml in serum and cerebrospinal fluid, respectively. Malignant cells were not found in cerebrospinal fluid obtained during a lumbar puncture. The final clinicopathological diagnosis was therefore a mixed secretory germ cell primary tumor of the spine.

Chemotherapy consisting of cisplatin 25 mg/m²/day for 4 days, bleomycin 18 mg/m² on Days 2 and 16, and etoposide 125 mg/m²/day for 4 days was undertaken. After the first course of chemotherapy the cerebrospinal marker values were within the normal range. The chemotherapeutic protocol was repeated for a total of four 4-week courses prior to the delivery of radiation therapy. Craniospinal irradiation at a total dose of 30 Gy (150 cGy daily fraction) plus a 9-Gy dose to the tumor site was then delivered and two additional chemotherapy courses completed the treatment. At 33 months after the diagnosis, there was complete disease remission and a neurological examination revealed normal findings.

Discussion

Intramedullary spinal cord germ cell tumors are generally represented by metastases, prevalently located in cervical segments. A primary location of germ cell tumors in the spine is very rare and so far has been described in 15 other cases, mostly arising in the dorsolumbar tract; in 12 of these cases, the patients were Japanese. Of these cases, only five had a β-HCG-secreting component like the one that we have described; our case is only the second one reported in the literature in which the patient is not Asian. The two patients reported on by Hisa, de Monléon, and their colleagues were young children who presented at diagnosis with features typical of precocious puberty and required medical attention before neurological symptoms appeared. This kind of clinical history is
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not exceptional when dealing with secretory germ cell tumors, with the tumors noted both in gonadal and extragonadal locations.16-17 In another case, that of a prepubertal boy, the authors only described the neurological impairment and the pain leading to diagnosis.21 Because the pubertal development of our patient was complete, like those referred to by Sasaki,16 Takahashi,21 and their colleagues, the β-HCG secretion was not able to produce any detectable systemic endocrine alteration as described earlier.13,15 As a matter of fact, the presenting symptoms of our patient were merely related to the neurological impairment secondary to the lumbar location of the tumor and were therefore more easily attributable, due to the age of the young man, to a glial tumor such as an ependymoma, which was the first diagnosis.

The treatment that we chose to apply was the same used at our institution for a secretory germ cell tumor in the brain,16 and in our case complete remission of the tumor was obtained and maintained longer than 2 years after diagnosis. Note that the biological response to chemotherapy was rapid, as also reported by Takahashi, et al.,22 with levels of β-HCG in the serum returning to within the normal range after the first course. The other few therapeutic experiences described in the literature have shown that any less intensive treatment can result in local or distant recurrence11-13 requiring aggressive retreatment. In addition, our patient differed from most of the other patients in that he has been able to recover completely from his severe symptoms without any sequela.

Besides the more common histological subtypes of neoplasms arising in the spine in this age group, such as astrocytomas, ependymal tumors, and primitive neuroectodermal tumors, this case outlines the unexpected possibility of occurrence of a secretory germ cell tumor and its successful treatment.

References


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