Case Report

Spinal cord compression of primary extragonadal giant yolk sac tumor

A Guzel¹, M Tatli^{*1}, D Belen² and H Seckin²

¹Department of Neurosurgery, Faculty of Medicine, University of Dicle, Diyarbakir, Turkey; ²Clinic of Neurosurgery, Dişkapi Research and Education Hospital, Ankara, Turkey

Study design: Case report.

Objective: To report an adult male patient with a primary extragonadal giant yolk sac tumor presenting with acute spinal cord compression.

Setting: Faculty of Medicine, University of Dicle, Diyarbakir, Turkey.

Method: A 31-year-old man was referred to our department with a diagnosis of Pott's disease, a complaint of back pain and gait difficulty for 2 weeks. Neurological examination showed spastic paraparesis and hypoesthesia below the L2 dermatome level. He also had urinary incontinence. Abdominal computed tomography and lumbar magnetic resonance imaging study revealed a giant cystic mass lesion located in the psoas muscle. Posteriorly, the third lumbar vertebral body was destructed and the tumor was compressing the dural sac. A combined anterior and posterior approach was performed. Pathological diagnosis was a yolk sac tumor.

Result: His neurological status improved during the postoperative course. A chemotherapy protocol was given including bleomycin, etoposide and cisplatin. Five months after the last chemotherapy, he was brought to the emergency unit in sepsis and died despite antibiotherapy.

Conclusion: Yolk sac tumor should be considered in young adult male patients presenting with acute paraparesis even without any signs or medical history of a testis tumor. These tumors may be unresectable; however, decompression of neural structures and stabilization of the spine with instrumentation may cause substantial improvement in neurological deficit and pain relief.

Spinal Cord (2007) 45, 254–257. doi:10.1038/sj.sc.3101942; published online 30 May 2006

Keywords: adult; lumbar column; paraparesis; primary yolk sac tumor

Introduction

Yolk sac tumor is a malignant germ cell neoplasm. It primarily originates from testes or ovaries.^{1–4} In 2–5% of cases, it originates from extragonadal regions such as mediastinum, retroperitoneum, pelvis or pineal region. In rare instances, the tumor may invade the spinal column and this accounts for 1–5% of adult male patients.^{1,4–7} Retroperitoneal extension of the tumor has been reported in only 1% of male patients with germ cell tumors whose testes are clinically normal.⁸ There are very few cases with a primarily retroperitoneal tumor with spinal column or spinal cord invasion, and these patients are usually at the pediatric age group.^{2,4,9,10} In this article, we report an adult male patient with a primarily extragonadal giant yolk sac tumor located in the retroperitoneal region, presenting with acute spinal cord compression.

Case report

A 31-year-old male patient was referred to our department with a diagnosis of Pott's disease, a complaint of back pain and gait difficulty for 2 weeks. His past medical history included surgery for bilateral testicular varicoceles under spinal anesthesia 2 months earlier at another institution. There was no history for any mass lesions during varicocele surgery nor a report of a biopsy. His physics examination was normal. On neurological examination, a paraparesis with 3/5 of normal motor strength and bilaterally hyperactive deep tendon reflexes were detected. There was hypoesthesia under the level of the second lumbar vertebral roots together with urinary and anal incontinence. His routine blood chemistry tests were normal, but his lactate dehydrogenase (LDH), β -human chorionic gonadotropin (β -HCG) and α -fetal protein (α -FP) levels were sky-high. Both abdominal computed tomography (CT) and lumbar magnetic resonance imaging (MRI) revealed a heterogeneously enhancing cystic mass lesion (approximately $22 \times 17 \times 15$ cm in diameter) located in the psoas muscle and extending into the midline. The mass was pushing the abdominal aorta to the right, compressing

*Correspondence: M Tatli, Department of Neurosurgery, Faculty of Medicine, Dicle Üniversitesi, Tıp Fakültesi, Nöroşirürji AD. 1. Kat, (21180) Diyarbakir, Turkey

The informed consent of patient was obtained from his father after the nature of the publication had been fully explained

the right iliac artery. It was also compressing the left kidney and the urinary vesicle. Posteriorly, the third lumbar vertebral body was destructed and the tumor was extending into the spinal column, compressing the dural sac (Figures 1, 2, 3a and b). Thoracal CT was normal.

Drainage of the cyst was decided for diagnostic and decompressive purposes. A drain was inserted inside the cyst under ultrasonographic guidance. Hemorrhagic fluid with jet flow due to high pressure was drained. Cytological examination of the fluid did not reveal any tumor cells. Nevertheless, owing to the massive destruction of L3 vertebral body and extensive structure of the lesion, a diagnosis of invasive tumor with cyst formation and intracystic hematoma was made. The fluid was drained for 2 days and reached an amount of 500 ml before it ceased to drain. After the drainage of the fluid, repeated ultrasonography (USG) disclosed the heterogeneous nature of the mass. On the 7th day of

Figure 1 Axial abdominal CT scan revealing a large cystic heterogeneous mass (22 × 17 × 15 cm in diameter) surrounding the left anterolateral region of the spine (arrow)

Figure 2 T2-weighted axial MRI revealing the hyperintense mass located anteriorly to L3 vertebra (arrow) and tumor infiltration to the vertebral body

admittance, his paraparesis worsened to 1/5 of normal motor strength, and the patient was scheduled for surgery.

Surgery started with the transabdominal approach to the tumor. A grayish-red, highly vascularized, encapsulated mass filling the abdominal cavity was seen. The mass was firmly attached to the surrounding structures such as liver, gallbladder, pancreas, left kidney, abdominal aorta and vein as well as iliac arteries. It was considered unresectable, and the patient was closed after a biopsy. Intraoperative frozen pathological examination revealed malignant tumor of unknown origin. The patient was then turned and positioned in the prone position, and L3 total and L4 subtotal laminectomy was performed through a midline incision. The tumor was compressing the dura anteriorly, but only a biopsy could be taken owing to the massive hemorrhage. A posterior transpedicular instrumentation through L1–L5 was performed. Pathological diagnosis was reported as a yolk sac tumor (Figures 4 and 5).

His neurological status improved during the postoperative course. He was ambulatory without assistance on the 6th postoperative week with full continence. He was put on BEP (bleomycin 40 mg/q.d. i.v., etoposide 140 mg/qd i.v. and cisplatin 40 mg/q.d. i.v.) protocol by the medical oncology department. Bleomycin was given for 3 days in four rounds and the EP protocol was given in 5 days in two rounds at 4-week intervals. His β -HCG level was 0.75 ng/dl, α -FP level was 33 ng/dl and LDH level was 350 μ /dl after 2 weeks of chemotherapy. These values consistently remained low during weekly follow-ups. His follow-up brain and thoracal CTs were normal. Abdominal USG examinations did not show enlargement of the tumor.

Figure 3 (a and b) Contrast-enhanced T1-weighted sagittal (a), and T2-weighted sagittal (b) MR images demonstrating the hyperintense retoperitoneal solid mass locating along the anterior borders of L1–L5 (arrows). L3 vertebra is invaded and destructed by the tumor and spinal canal is severely compressed at the same level

Figure 4 Photomicrograph of the pathological specimen of the tumor demonstrates loose meshwork lined by flat or cuboidal cells, rounded or festooning pseudo-papillary processes with central vessels (Schiller–Duval bodies) and atypical tumor cells (H&E, $\times 100$)

After 5 months of his last chemotherapy, the patient was brought to the emergency unit with tachycardia, high fever and low blood pressure. Laboratory investigations showed anemia and leukocytosis. The tumor was seen unchanged on the USG examination. There were no brain or mediastinal metastases in CT examinations. He was diagnosed as a sepsis and blood, urine and throat cultures were obtained. Vancomycin 1 gm q.i.d. was initiated empirically. The patient's general condition worsened rapidly, and he died on the second day of his last admission.

Discussion

Teillum¹¹ was the first to describe the origin of yolk sac tumors as the embryonic vitellin. Malignant germ cell tumors account for about 3% of neoplasms in children, and endodermal sinus tumor (EST) is the most common histological subtype with a high degree of malignancy.¹² They may originate from gonads or from extragonadal organs and tissues.^{1,8,9,11,13–18} The exact pathogenesis of extragonadal germ cell tumors is unknown, but it has been suggested that the malignancy process starts during the early embryological days. The primordial cells located in the midline of the embryo are thought to be affected during this period and develop a malignant tumor later.^{13–18}

Only two cases have been reported in the literature with metastasis to the spinal column and compression of the spinal cord.^{10,19} However, we have not found in the literature any adult cases located in the retroperitoneum and presenting primarily with spinal cord compression.

In the presented case, the history of surgery for bilateral varicoceles under spinal epidural anesthesia led us to suggest a surgical infection. However, normal body temperature, normal sedimentation rate, leukocyte counts and normal CRP levels helped us to rule out this

Figure 5 Immunohistochemical staining of the tumor tissue shows strong immunoreactivity to AFP in the tumor cells (immunoperoxidase $\times 100$)

diagnosis. High β -HCG and α -FP levels suggested a diagnosis of a tumor of germ cell origin. Examination of the cyst content was non-contributory. Pathological examination confirmed the definitive diagnosis as nonseminomatous germ cell tumor.

In germ cell tumors, even the testicular biopsy is not sufficient to conclude that the tumor is primarily extragonadal or metastatic.^{7,20,21} Both these tumors bear the same cytogenetic properties as well. The extragonadal location of a germ cell tumor may also indicate a high-grade tumor, suggesting a poor prognosis.^{4,11,22}

The treatment of EST causing cord compression has traditionally been laminectomy with or without radiation, followed-up by chemotherapy.^{2,4,23} However, management of such tumors is always a challenge, especially if the patient suffered from progredient paraparesis with the need for intervention. The management of such a giant lesion can be discussed critically, especially if the neurologic state has diminished. The first aim should be the preservation of the neurologic function and the second the observation of histologic specimen. Pashankar *et al*²³ reported that they used a different method. After performing a biopsy of the paraspinal mass, they opted to use only chemotherapy without laminectomy or radiation therapy to decompress the epidural mass. With this approach, they had excellent tumor response and a complete neurologic recovery despite a near-complete paraplegia at presentation.

In conclusion, yolk sac tumor should be considered in young adult male patients presenting with acute paraparesis even without any signs or medical history of testis tumor. CT and MRI are the preferred radiological examinations in the diagnosis of these tumors and CT-guided biopsy is of value for preoperative diagnosis. These tumors may be unresectable; however, decompression of neural structures and stabilization of the spine with instrumentation may cause substantial improvement of neurological deficit, and pain relief may also be achieved.

Acknowledgements

We thank Fahri Yilmaz, MD from the department of pathology for his work in pathological diagnosis.

References

- Gerl A, Clemm C, Lamerz R, Wilmanns W. Cisplatin-based chemotherapy of primary extragonadal germ cell tumors. A single institution experience. *Cancer* 1996; **77**: 526–532.
- Resnick DK, McLaughlin MR, Albright AL. Primary endodermal sinus tumor presenting with spinal cord compression. Case report. *J Neurosurg* 1997; **86**: 151–153.
- Toner GC, Geller NL, Lin SY, Bosl GJ. Extragonadal and poor risk nonseminomatous germ cell tumors. Survival and prognostic features. *Cancer* 1991; **67**: 2049–2057.
- Wada S et al. Primary retroperitoneal pure yolk-sac tumor in an adult male. *Scand J Urol Nephrol* 2001; **35**: 515–517.
- Goss PE et al. Extragonadal germ cell tumors. A 14-year Toronto experience. *Cancer* 1994; **73**: 1971–1979.
- Hartmann JT et al. Prognostic variables for response and outcome in patients with extragonadal germ-cell tumors. *Ann Oncol* 2002; **13**: 1017–1028.
- Scholz M, Zehender M, Thalmann GN, Borner M, Thoni H, Studer UE. Extragonadal retroperitoneal germ cell tumor: evidence of origin in the testis. *Ann Oncol* 2002; **13**: 121–124.
- Saltzman B, Pitts WR, Vaughan Jr ED. Extragonadal retroperitoneal germ cell tumors without apparent testicular involvement. A search for the source. *Urology* 1986; **27**: 504–507.
- Kurisaka M, Moriki A, Mori K, Sonobe H. Primary yolk sac tumor in the spinal cord. *Childs Nerv Syst* 1998; **14**: 653–657.
- Lee JK et al. Metastatic spinal cord compression of testicular yolk sac tumor. *Childs Nerv Syst* 2002; **18**: 171–174.
- Chong CD, Logothetis CJ, von Eschenbach AC, Ayala AG, Samuels ML. Successful treatment of pure endodermal sinus tumors in adult men. *J Clin Oncol* 1988; **6**: 303–307.
- Davidoff AM, Hebra A, Bunin N, Shochat SJ, Schnauffer L. Endodermal sinus tumor in children. *J Pediatr Surg* 1996; **31**: 1075–1078.
- Bokemeyer C et al. Extragonadal germ cell tumors of the mediastinum and retroperitoneum: results from an international analysis. *J Clin Oncol* 2002; **20**: 1864–1873.
- Chong SM, Wee A, Yeoh SC, Nilsson B, Chan S. Retroperitoneal endodermal sinus tumor. Report of a case with an abnormal cervicovaginal smear. *Acta Cytol* 1994; **38**: 562–567.
- Dede M, Pabuccu R, Yagci G, Yenen MC, Goktolga U, Gunhan O. Extragonadal yolk sac tumor in pelvic localization. A case report and literature review. *Gynecol Oncol* 2004; **92**: 989–991.
- Gunawardena SA, Siriwardana HP, Wickramasinghe SY, Madurawe MN, Fernando R. Primary endodermal sinus (yolk sac) tumour of the liver. *Eur J Surg Oncol* 2002; **28**: 90–91.
- Park NH, Ryu SY, Park IA, Kang SB, Lee HP. Primary endodermal sinus tumor of the omentum. *Gynecol Oncol* 1999; **72**: 427–430.
- Schmoll HJ. Extragonadal germ cell tumors. *Ann Oncol* 2002; **4**: 265–272.
- Colak A, Benli K, Berker M, Onol B. Epidural metastasis of testicular yolk sac tumor: an unusual cause of spinal cord compression. Case report. *Pediatr Neurosurg* 1991–92; **17**: 139–141.
- Böhle A, Studer UE, Sonntag RW, Scheidegger JR. Primary or secondary extragonadal germ cell tumors? *J Urol* 1986; **135**: 939–943.
- Fossa SD et al. Testicular carcinoma *in situ* in patients with extragonadal germ-cell tumours: the clinical role of pretreatment biopsy. *Ann Oncol* 2003; **14**: 1412–1418.
- Schmoll HJ, et al., European Germ Cell Cancer Consensus Group. European consensus on diagnosis and treatment of germ cell cancer: a report of the European European consensus on diagnosis and treatment of germ cell cancer: a report of the European Germ Cell Cancer Consensus Group (EGCCCG). *Ann Oncol* 2004; **15**: 1377–1399.
- Pashankar FD, Steinbok P, Blair G, Pritchard S. Successful chemotherapeutic decompression of primary endodermal sinus tumor presenting with severe spinal cord compression. *J Pediatr Hematol Oncol* 2001; **23**: 170–173.