

Huge Retroperitoneal Germinoma Presenting with Pathological Fracture of the Spine

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Primary retroperitoneal germ cell tumors are extremely rare neoplasms. The most common presenting features are abdominal pain and palpable abdominal masses. Pathological fractures of the spine presenting as bilateral lower leg weakness are exceptionally rare. We describe a 16-year-old girl who developed progressive paraplegia after a minor falling injury. Radiological study demonstrated a huge retroperitoneal tumor with invasion of the T12 vertebral body and spinal canal. A posterior surgical approach was used to perform laminectomy (T12, L1), removal of the intraspinal tumor and internal fixation with transpedicular screws (T10, T11 to L2,3), and posterolateral fusion. Postoperative combination chemotherapy for six cycles with cisplatin (100 mg/m² per day for 1 day every 3 weeks), bleomycin (15 units intravenously weekly for 18 weeks) and etoposide (100 mg/m² per day for 3 days every 3 weeks) were given and the tumor responded dramatically. The patient had fully recovered without evidence of sequelae or recurrence at 2 years after operation. To the authors' knowledge, this is the first case in which a huge retroperitoneal germinoma presented as pathological fracture of the spine and spinal cord compression. The effectiveness of the postoperative cisplatin-based chemotherapy against this tumor made major retroperitoneal surgery to remove the main tumor mass unnecessary is also demonstrated. (*Chang Gung Med J* 2002;25:844-9)

Key words: retroperitoneal germinoma, spinal pathological fracture, chemotherapy.

Primary tumors of the retroperitoneum are rare and consist of neoplasms of various tissue types. Eighty-five percent of these tumors are malignant.⁽¹⁾ Lymphomas or sarcomas are the most common histopathological types. Only 5 to 25% of retroperitoneal tumors are germ cell tumors.⁽²⁾ Most patients with retroperitoneal germ cell tumors have signs or symptoms suggestive of abdominal masses. Spinal cord compression by a retroperitoneal germ cell tumor has not been reported. Here we describe a patient with a huge retroperitoneal germinoma presenting with spinal pathological fracture and spinal cord compression.

CASE REPORT

This previously healthy 16-year-old girl suffered from low back soreness after a fall on the bus. Five days after the fall, in November 1999, she visited the emergency department due to aggravation of back pain, weakness and numbness of both lower limbs, and urine retention.

Physical examination revealed a mid-abdominal mass, measuring about 10 cm in diameter, and back mobile pain. Muscle power of the right leg was grade 2/5 and that of the left leg was grade 3/5. Plain radiography of the thoraco-lumbar spine

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showed widening of the paraspinous line and a compression fracture of the T12 vertebral body. Magnetic resonance (MR) imaging showed a huge retroperitoneal tumor occupying bilateral para-aortic and para-spinal regions. The T12-L1 epidural space was invaded by the tumor and the spinal cord was compressed from the right side (Fig. 1). Right

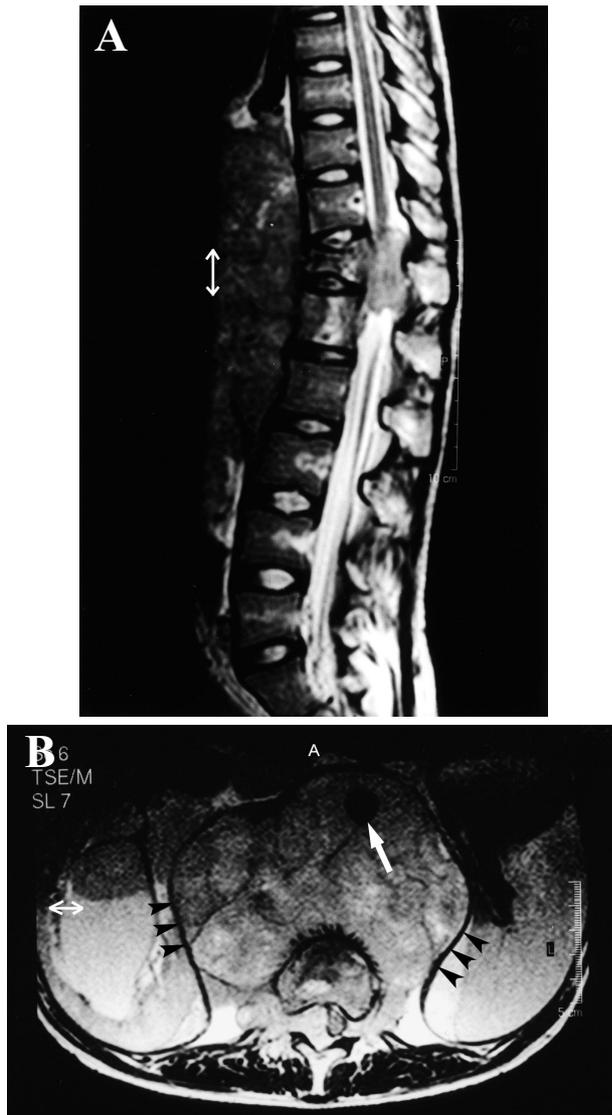


Fig. 1 Unenhanced sagittal (A) and axial (B) T2-weighted MR images showing a huge retroperitoneal tumor (arrowheads) with encasement of the aorta (arrow) and invasion to the T12 vertebra. Intraspinous extension to the right epidural space with considerable compression of the spinal cord is seen.

ureteral encasement with resultant hydronephrosis was also noted. Serum alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (β -HCG) were within reference ranges. Gynecologic studies including pelvic ultrasound and computed tomography examinations showed no organic anomalies.

A T12 and L1 laminectomy was performed via the posterior approach. After the spinal canal was opened, the spinal cord was found to be displaced to the left side by a solid and grayish tumor. The intraspinal tumor was easily removed with the help of an ultrasonic surgical aspirator. The spinal cord returned to its normal position upon removal of the tumor. Screw fixations of T10, T11 to L2, L3 transpedicular area with posterolateral fusion were then performed. Right double-J stent insertion for right hydronephrosis was performed in another operation 1 week after the screw fixations.

Grossly, the excised tumor fragments, measuring up to 2.5 \times 1.2 \times 0.3 cm, appeared solid and gray. On microscopic examination, the tumor cells were arranged in sheets or in a diffuse fashion, bounded by substantial fibrous septa, which were densely infiltrated by lymphocytes. The neoplastic cells were polygonal with large, round to oval, vesicular nuclei with prominent centrally located nucleoli (Fig. 2). The nuclear membrane was distinct and the nuclear chromatin was unevenly distributed. Brisk

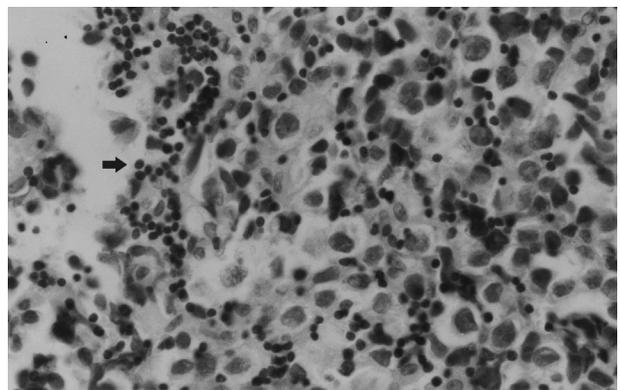


Fig. 2 The excised specimen revealing sheets of polygonal neoplastic cells containing large, vesicular nuclei with distinctive nuclear membrane and conspicuous centrally located nucleoli. Delicate fibrous septa that were heavily infiltrated by lymphocytes are shown (arrow). (Haematoxylin and eosin, \times 200)

mitotic activity was noted. The cytoplasm of the tumor cells had a clear to eosinophilic appearance. Neither glands, cords, tubules, papillae nor Schiller-Duval bodies could be identified. Further immunohistochemical studies revealed strong immunoreactivity for placental alkaline phosphatase (PLAP), and cytokeratin (AE1/AE3) but results were negative for AFP, β -HCG, and carcinoembryonic antigen (CEA). The lesion was diagnosed as a germinoma.

The postoperative course was uneventful. Brain computerized tomography scanning revealed no intracranial lesions. Muscle power of both legs improved rapidly and she could walk independently 2 weeks after the operation. Urination recovered to normal function 1 month after the operation. She received six cycles of cisplatin-based chemotherapy during the period from December 1999 through May 2000. The regimen included cisplatin (100 mg/m²

per day for 1 day every 3 weeks), bleomycin (15 units intravenously weekly for 18 weeks) and etoposide (100 mg/m² per day for 3 days every 3 weeks). Two episodes of neutropenic fever were noted during the interval between the six chemotherapy treatment cycles, one was between the first and second cycle and the other was between the fourth and fifth cycle. The episodes each resulted in 2 weeks delay in chemotherapy. There were no sequelae after completion of the six cycles of chemotherapy during 15 months follow up. MR imaging performed in November 2001 (24 months after the operation and 19 months after completion of chemotherapy) showed a remarkable decrease of the size of the retroperitoneal tumor (Fig. 3). Serum lactate dehydrogenase, β -HCG, AFP, and alkaline phosphatase at 1 year after completion of the chemotherapy were in reference ranges.

DISCUSSION

Germ cell tumors are rare and occur primarily in young individuals. They are found in both sexes and may arise in gonadal and extragonadal sites.⁽³⁾ The most common sites of extragonadal germ cell tumors are midline and can include the mediastinum, retroperitoneum, sacrococcygeal region and pineal region. The retroperitoneum or mediastinum is the most common site of origin of primary extragonadal germ cell tumors.^(4,5) Germinomas are the most common histological type of germ cell tumors, accounting for nearly half of germ cell tumors in the cranium and ovary.^(6,7) In the retroperitoneum, however, most germ cell tumors are teratomas or mixed germ cell tumors.^(2,5) Retroperitoneal germinomas are rare and their incidence is unknown. The present case showed the characteristics of a pure germinoma, including normal serum AFP, and β -HCG and homogenous reaction for PLAP stain but no evidence of a reaction for AFP, β -HCG or CEA stain using results of immunohistochemical studies. In addition, the tumor cells in this case were also immunoreactive to cytokeratin. Cytokeratin has been used to distinguish germinoma from other germ cell tumors.⁽⁸⁾ However, subsequent studies detected cytokeratin in 9-80% of seminomas tested.^(9,10) This makes the usefulness of cytokeratin to differentiate germinoma from nongerminomatous germ cell tumor questionable.



Fig. 3 Gadolinium-enhanced sagittal T1-weighted MR images after surgery and chemotherapy showing re-expansion of the thecal sac without any intraspinal tumor mass.

With the exception of the endocrine-secreting type, retroperitoneal tumors are usually diagnosed late in the disease course as patients typically present with tumors that are already extensive. Pain is usually the most common complaint and is often referred to the abdomen, back, or flank. Other symptoms include abdominal swelling, weight loss, weakness, nausea, constipation, edema, anorexia, adenopathy, anuria, fever, or painful varicocele. An abdominal mass or tenderness is the most common physical finding. Our patient developed progressive paraparesis after a minor injury. Physical examination showed a mid-abdominal mass while plain radiography demonstrated pathological fracture of the lower thoracic vertebra. MR imaging revealed a huge retroperitoneal tumor with intraspinal extension and spinal cord compression. To our knowledge, retroperitoneal germinoma with pathological fracture of the spine and intraspinal extension has not been previously reported.

Most retroperitoneal germ cell tumors are treated with surgical excision of the main tumor mass and postoperative combination chemotherapy. In our patient, surgery was undertaken via the posterior approach for spinal cord decompression, stabilization, and pathological diagnosis but not for main tumor excision. After postoperative chemotherapy, the volume of the retroperitoneal main tumor mass decreased dramatically. This finding suggests that major retroperitoneal operation to remove a huge retroperitoneal germ cell tumor mass may be unnecessary.

One of the most commonly used chemotherapy regimens for germ cell tumors of the ovary is cisplatin-based. Gerl et al found that extragonadal seminomas (germinomas) responded well to cisplatin-based chemotherapy and concluded that the majority of patients with extragonadal seminomas, regardless of the site of presentation, can expect to be cured.⁽⁵⁾ In our patient, MR imaging revealed that the tumor volume decreased dramatically after six courses of cisplatin-based chemotherapy. This result further suggests the effectiveness of cisplatin-based chemotherapy in extragonadal germinomas as has been reported.^(5,11,12)

Germinomas are very sensitive to radiation therapy and may be cured even in cases of gross metastatic disease. Loss of fertility remains a problem with

radiation therapy.⁽¹³⁾ After the pathological diagnosis was made in our patient, chemotherapy was given instead of radiation therapy due to the huge size of the tumor, and location in a region in which radiation therapy would be hazardous to the abdominal organs, as well as to preserve her fertility.

In summary, a patient presenting with a huge retroperitoneal germinoma with a pathological fracture of the spine is extremely rare. After spinal decompression and stabilization procedures, combination chemotherapy with bleomycin, etoposide and cisplatin was effective against this extensive tumor.

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