CASE REPORT

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Retroperitoneal Yolk Sac Tumor in Adult Woman Presenting as Spinal Cord Compression and Fatal Pulmonary Tumor Embolism

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A 35-year-old woman, previously treated for systemic metastases from retroperitoneal yolk sac tumor, presented with progressive painful paraparesis. Preoperative images showed severe cord compression by the metastatic infiltration of the lumbar vertebrae and epidural mass as well as a huge retroperitoneal mass. While performing unremarkable surgery in prone position, the patient abruptly fell into hypoxic insults and circulatory arrest. Intraoperative pulmonary tumor embolism was deemed a cause of death. When planning operative procedure for this dangerous malignancy, scrupulous manipulation is mandated and the possibility of fatal pulmonary tumor embolism should also be addressed and fully discussed preoperatively.

KEY WORDS : Extragonadal (retroperitoneal) · Pulmonary tumor embolism · Spinal cord compression · Yolk sac tumor.

Introduction

Yolk sac tumor(YST) is the most common primary testicular malignancy of germinal origin in childhood and infancy. It usually arises in the gonads, but can occasionally occur in extragonadal sites such ascentral nervous system, mediastinum, retroperitoneum, and sacral region[11,12]. Epidural metastases of the YST were rarely reported to cause spinal cord compression, although found in children. Only one case of such clinical manifestation has been described in adult patient in English literatures[4,5,7,8].

Pulmonary tumor embolism(PTE), unusual cause of dyspnea in a patient with cancer, is characterized by pulmonary hypertension and right heart failure, and is caused by multiple tumor embolic shedding to the pulmonary vasculature[1,3,8]. Up to the present, only two cases of fatal PTE due to the YST have been reported, and development of such PTE during neurosurgical procedure was described in only one case[1,10].

We present a rare case of vertebral metastasis from the retroperitoneal YST in an oophorectomized adult female who eventually succumbed to an intraoperative PTE. Possible pathogenic mechanisms of this fatal intraoperative PTE were discussed.

Case Report

A 35-year-old woman was consulted to the Neurosurgical department for operative management of recalcitrant spinal epidural mass. The patient complained of progressive painful weakness of lower limbs for 2 weeks. On examination, she showed paraparesis (Gr. 2), painful dysesthesia in both legs and urinary incontinence. She had been suffering from intermittent dyspnea during exertion for several months. She also felt tenderness at palpating on the left flank region. But, she denied abdominal pain and tenderness despite noticeable bulging on external appearance and palpation. Plain lumbar X-rays showed compression fracture at L-1 to L-3 vertebral bodies with consequent kyphotic deformity. Magnetic resonance images of the lumbar spine showed severe conus-cauda compression by the epidural mass chiefly located at L-3, signal change within L-1 to L-4 vertebral bodies and huge left suprarenal mass (62 × 60 × 132mm) that encased the left common iliac artery and inferior vena cava(IVC). The retroperitoneal mass involved from L-1 to L-5 vertebral levels, ventrally displaced abdominal aorta and communicated with the epidural mass. Radionuclide bone scan also revealed foci of hot uptake

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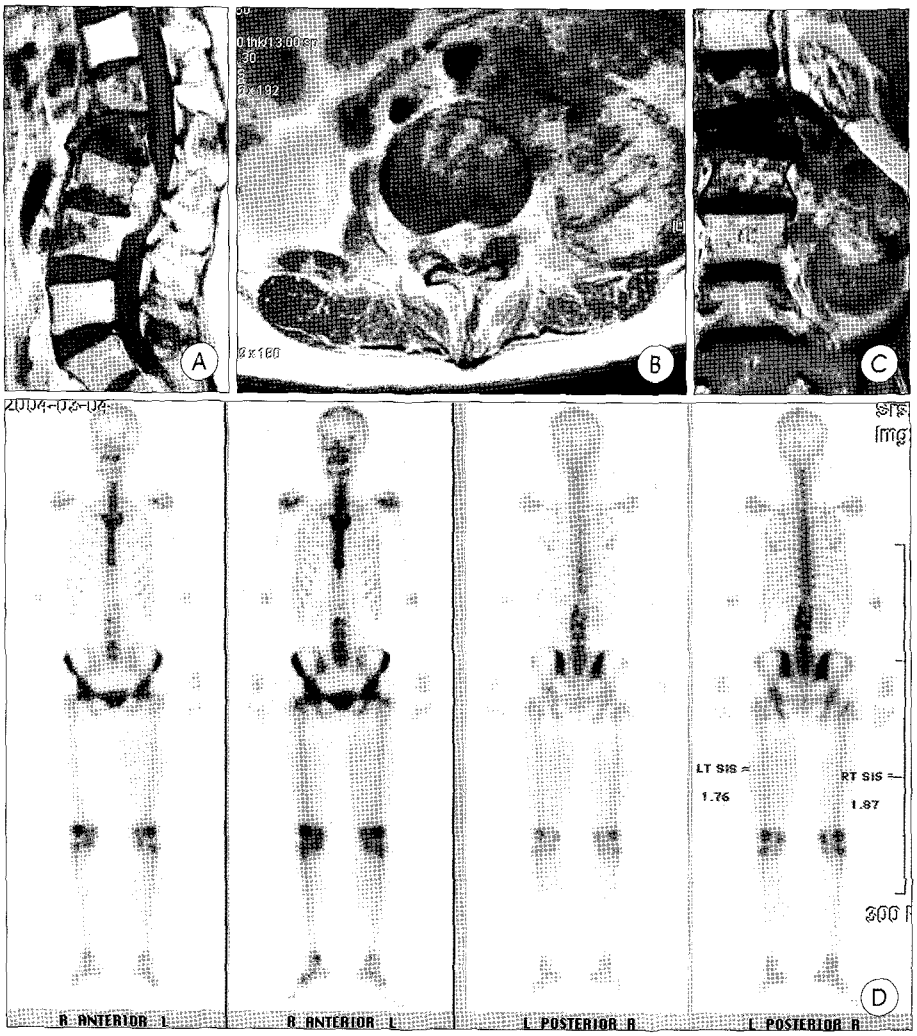

Fig. 1. *Preoperative Images. Contrast-enhanced sagittal T1-weighted magnetic resonance image* (A) and axial T1-weighted image at the L-3 level (B) show destruction of L-1 and L-2 vertebral bodies, and concomitant signal changes within bodies of L1 to L-4. Epidural mass exhibits strong but inhomogeneous enhancement and compresses cauda equina principally at the L-3 level. Enhanced coronal T1-weighted image (C) shows left paraspinal suprarenal mass (62×60×132mm) with inhomogeneous signal intensities connecting with the destroyed L-2 body. Radionuclide bone scan (D) shows increased uptake at L-1 to L-3, while decreased uptake at T-12 level. The latter finding is suggestive of post-irradiation change of bone marrow.

at corresponding vertebrae (Fig. 1). Chest X-rays and computed tomography scans showed multiple metastatic nodules, and serum tumor marker study revealed notably high level of α-fetoprotein(AFP) of 597ng/ml, suggestive of metastatic YST. For three years, she has been on the second cycle chemotherapy (etoposide, vincristine) and palliative external irradiation (3,500 rad). She had a significant medical history of oophorectomy for left ovarian cyst at the age of 6.

Because of strong evidence of left iliac artery and IVC involvement with the retroperitoneal mass and no abdominal symptoms, direct decompression via a posterior approach was planned, instead of retroperitoneal route. The patient was placed into a prone position under a general anesthesia. Anterolaterally placed, yellow-gray epidural mass with friable but rubbery consistency was found following full decompression of L-1 to L-4 and retracting thecal sac. Tumor was initially removed manually by piecemeal fashion, and remaining portion around the intervertebral foramina of L-2 and L-3 was resected by using Ultrasonic aspirator, Dissectron (Satelec, Bordeaux, France). During the procedure to this point, blood loss was approximately 1,500ml, and all intraoperative anesthetic parameters were within normal, being continuously monitored by an intra-arterial catheter. When preparing for instrumentation, blood in the operative bed suddenly turned into black. Attending anesthesiologist found the patient's cyanotic hands and face, and her arterial oxygen saturation dropped to 60~70% and central venous pressure elevated markedly (28mmHg). Despite succeeding anesthetic manipulation, her blood pressure was abruptly fallen down to 50mmHg, quickly followed by ventricular fibrillation and asystole. Cardiopulmonary resuscitation was immediately commenced after placing her toward the supine position, and continued for 40minutes with every effort available, but the patient passed away.

Autopsy was recommended to disclose the exact cause of sudden death, but her family refused. Histopathological examination of the surgical specimen confirmed findings consistent with YST of extragonadal origin (Fig. 2).

Discussion

The YST is an uncommon malignancy that usually occurs in the testis or ovary of children or young adults. However, the YST was occasionally reported to occur in the mediastinum, sacrococcygeal region, retroperitoneum, liver, vagina and brain. In addition to the primary involvement of the central nervous system, these tumors infrequently metastasize to the brain and spine. Majority of symptomatic spinal cord compression has been reported in juvenile patients[4,7,8], and only

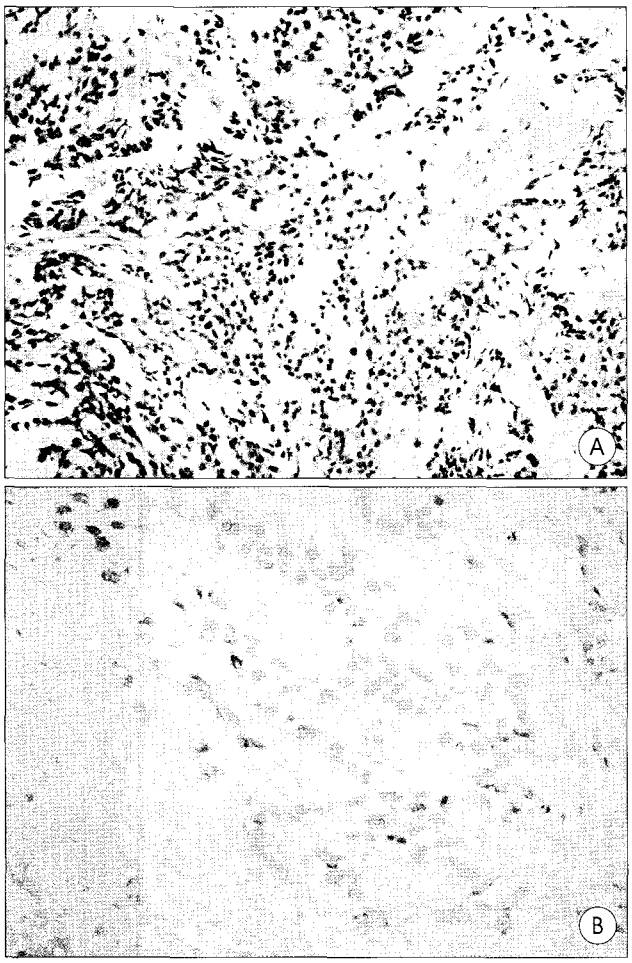

Fig. 2. Histopathology. (A) There are solid sheets of small round and polygonal tumor cells with intervening fibrovascular tissue, as well as reticular pattern of tumor cells in a loose myxoid stroma (H &×200 original magnification). (B) Some tumor cells exhibit strong positivity for α-fetoprotein (PAP, ×400 original magnification).

one case of metastatic ovarian YST in a 46-year-old woman with such conus-cauda syndrome has been described[5]. Comparing to the previous report, YST in the current case seemed to arise *de novo* in the left retroperitoneum because this patient had undertaken left unilateral oophorectomy about 30 years ago due to the ovarian cyst torsion. We are still uncertain about the pathogenesis of YST in the absence of ovarian tissue. Sustained hormonal stimulus to the microscopically remnant ovarian cells or dystrophic tissue deemed account for the development of extragonadal YST.

Therefore, this is the first case report of spinal cord compression due to vertebral metastasis from the primary retroperitoneal (extragonadal) YST. In this case, direct extension from a paraspinal retroperitoneal tumor through the intervertebral foramina seems to be likely. In addition, retroperitoneal lymphatics and lymph nodes also deem to play an ancillary role in its metastatic spread.

PTE is a well-recognized phenomenon in practically all types of carcinoma[2], but it is rarely diagnosed and also easily overlooked ante-mortem, because of critically ill condition of the patients[3,9]. Their incidence is, therefore, undoubtedly underestimated, and only two cases of fatal PTE were described in patients with the ovarian YST[1]. In PTE, large tumor emboli in the main pulmonary arteries or big segmental branches with subsequent mechanical obstruction of major pulmonary microvasculature accounts for the increased pulmonary resistance, which will lead to right heart failure and cor pulmonale[2,3,9]. After then, sudden onset of dyspnea, chest pain, and hemoptysis are quickly followed by circulatory collapse and death[3]. In cases with retroperitoneal malignancy, involvement of IVC might be another culprit for development of PTE : hematogenous dissemination via lumbar veins or direct pressure erosion of venous wall by bulky retroperitoneal lymph nodes[6]. In a patient with known cancer, PTE should be differentiated from pulmonary thromboembolism, lymphangitis, and pulmonary metastasis[3]. Pathologic confirmation of the same malignant cells within the pulmonary vasculature is the key factor in the differnetial diagnosis, if possible. In neurosurgical literatures, only one case of intraoperative fatal PTE has been documented, and this patient died of massive embolization of the pulmonary vasculature by fragmented tumor (choroid plexus papilloma) and secondary reactive pulmonary edema[10]. In this case, in spite of no postmortem pathologic verification of PTE due to refusal, following points can be addressed to assumeintraoperative PTE. First, the patient suffered intermittent dyspnea preoperatively, suggestive of PTE, although further diagnostic procedure such as ventilation-perfusion scanning was not performed[9]. Second, she underwent irradiation and systemic chemotherapy prior to the surgical intervention, and these therapies did weaken the vascular wall. Third, the operative procedure was conducted for 6 hours in prone position with intermittent external pressure loading, and this seemed to affect the retroperitoneal mass adversely. This possibility can be ascertained by reviewing MR findings of incorporation and close proximity to the iliac artery and IVC with the main retroperitoneal mass.

And last, both friable tumor nature and intraoperative use of Ultrasonic aspirator could facilitate microfragmentation and access to the pulmonary vasculature in a certain but not fully understood manner.

Conclusion

When confronting surgical intervention to the potential life-threatening high-risk situation, such as seen in this case, extreme intraoperative caution and preoperative informed consent prophylactically addressing this kind of sequels should be prepared.

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