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CASE REPORT

Conus–Cauda Syndrome as a Presenting Symptom of Endodermal Sinus Tumor of the Ovary

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We report on a case of a 46-year-old woman with a conus–cauda syndrome due to an endodermal sinus tumor of the right ovary with multiple metastases in the spine and pelvic bone. Before removing the tumor surgically, combination chemotherapy was given to treat the metastases, which threatened to compromise the spinal cord. © 1995 Academic Press, Inc.

INTRODUCTION

Endodermal sinus tumor (EST) of the ovary is an extraembryonal nonseminatous germ cell tumor. It accounts for about 1% of all ovarian malignancies. It generally occurs in late childhood and early adulthood. The mean age at which the diagnosis is set is about 19 [1, 2], but although uncommon at advanged age, it has been reported up to the age of 62 [3].

The tumor is highly malignant and grows rapidly, so the delay between the onset of symptoms and the diagnosis is usually short with an average of about 1 month [1, 2].

Therapy depends on the stage of the tumor and the wish to preserve fertility. Since in most cases the tumor is diagnosed at stage I [1, 2, 4], removing the tumor through unilateral salpingo-oophorectomy followed by combination chemotherapy is the treatment of choice. In the lower stages the prognosis is quite good and fertility can be preserved [4]. Nevertheless, the prognosis in stage IV EST is still very poor [2]. In stage IV total hysterectomy with bilateral salpingo-oophorectomy and cytoreductive surgery in combination with multiple agent chemotherapy is, in general, the chosen therapy.

A wide range of combinations of chemotherapeutic agents has been used in the past [1, 4]. Most widely used are VAC (vincristine, actinomycin D, and cyclofosfamide), PVB (cisplatin, vinblastine, and bleomycin), BEP (bleomycin, etoposide, and cisplatin), and VcBP (vincristine, bleomycin, and cisplatin).

A conus–cauda syndrome is a neurological syndrome due to a combined lesion of the cauda equina and of the lower sacral segments of the spinal cord (conus medullaris) at the level of the first lumbar vertebra. A lesion of the conus medullaris is characterized by early disturbance of bladder and bowel function, sensory loss (saddle anesthesia), and dissociated sensory disturbance in the sacral and cocygeal segments. A lesion of the cauda equina leads to a flaccid paralysis of the lower extremities, sensory loss, loss of sphincter control, and absent reflexes. The syndrome varies to the site and the extend of the lesion.

We describe a patient with an unusual presentation due to metastases from EST of the ovary and we applied an unusual sequence of treatment.

CASE HISTORY

A 46-year-old Dutch woman suffering from a progressive backache and a heavy feeling in both thighs for 1 month was referred to the Department of Neurosurgery. She was a nulligravida and had no relevant medical history. Walking had become progressively impaired during the preceding weeks. She recently suffered from urinary retention and incontinence for feces.

On physical examination a mass in the abdomen was palpated extending from the pelvis to the epigastric region. On neurological examination she had a marked flaccid paresis of both legs, proximal grade 1/5, and distal grade 0/5. All sensory modalities were absent down from the level L4. The knee reflexes were negative on both sides, whereas both ankle reflexes were increased. The plantar responses were indifferent. The anal reflex was intact. Under the diagnosis of an almost complete conus–cauda syndrome an MRI scan of the lumbosacral spine was performed. A tumor in L1, expanding into the soft tissues and compressing the spinal cord and cauda was seen (Fig. 1). Also a process in L4 and

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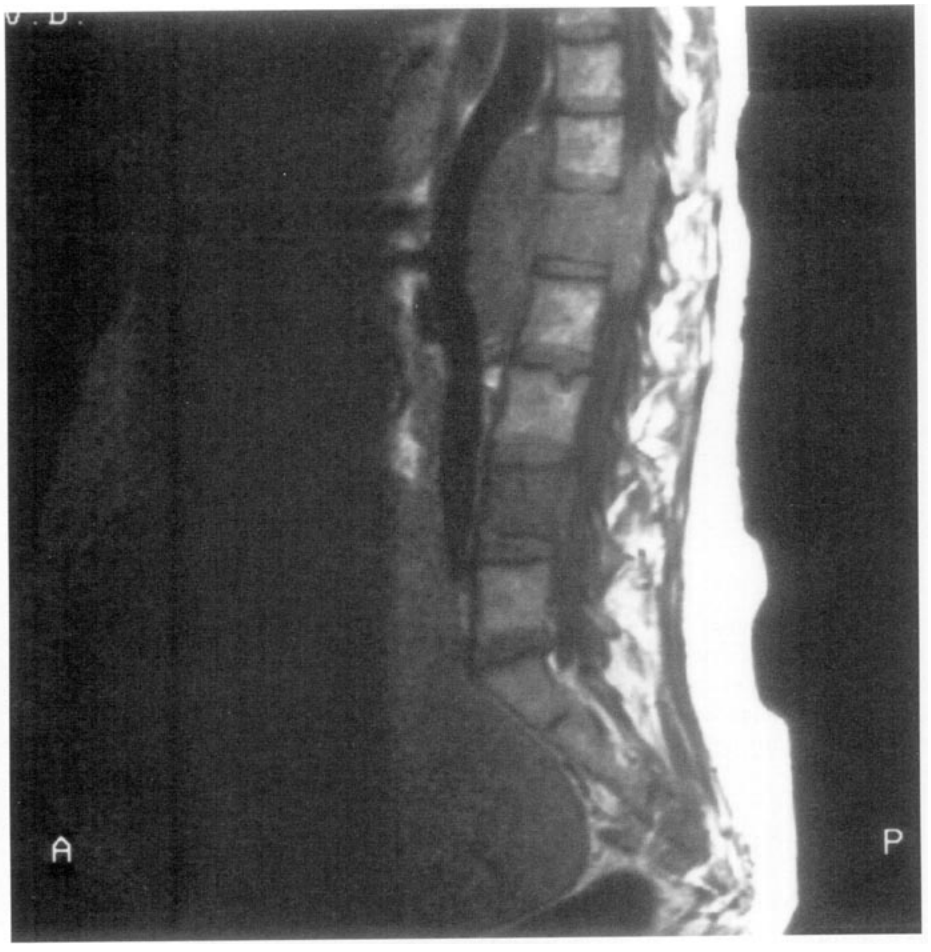

FIG. 1. A T1-weighted SE image (TR500; TE25 at 0.5 T) shows a metastasis in L1 with extensive involvement of surrounding tissues including displacement of the aorta and epidural tumor. A second metastasis is seen in L4. (The lower sacral metastasis was better seen on different slices.)

in the lower sacral region was identified. In case of an acute compression of the spinal cord due to a vertebral metastasis, in our clinic the policy is to perform a laminectomy to decompress the cord followed by radiation therapy. A laminectomy Th12–L1 was performed. The pathologic examination of the biopsies taken during the operation showed an endodermal sinus tumor. Because of this diagnosis we chose to start chemotherapy instead of radiotherapy. The patient was then referred to the Department of Gynecology for further treatment. On a second MRI scan of the lower abdomen we saw a cystic process in the pelvis with metastases in L1, L4, and in the lower sacral region (Fig. 2).

Because of a FIGO stage IV endodermal sinus tumor of the ovary with metastases, which led to severe neurological complications, we chose to start combination chemotherapy prior to cytoreductive surgery. Chemotherapy consisted of 3 courses of cisplatin 50 mg/m^2 iv, Days 1 and 2, vincristine 1.4 mg/m^2 iv, Day 1, and bleomycin 30 mg as an 12-hr infusion iv, Day 1 (BOP) every 10 days, followed by 3 courses consisting of cisplatin 20 mg/m^2 iv, Days 1 to 5, ifosfamide 1.0 g/m^2 iv, Days 1 to 5, and etoposide 100 mg/m^2 iv, Days 1, 3, and 5 (VIP) every 21 days. Serum α-fetoprotein (AFP), as a tumor marker, was assessed on a regular basis and its level decreased rapidly after starting chemotherapy (Fig. 3).

The patient received physical therapy and slowly regained strength in both legs, so she could walk a short distance between bars. After the first three courses there was a decrease in the expansion in the soft tissue of the metastasis in L1 seen on the MRI scan, but no changes in the cystic process in the pelvis. After the fourth course of chemotherapy the serum-AFP level no longer decreased log-linearly. The explanation for such a plateau in the concentration of serum-AFP could be because of either a reservoir of AFP in the cystic portion of the tumor or residual malignant disease. To solve this question a total hysterectomy with bilateral salpingo-oophorectomy was performed and a tumor weighing 3571 g, originating from the right adnex, was removed. The tumor contained 3060 g of a yellow–green fluid with a concentration of AFP of 3419 μg/liter. Serum-AFP before surgery was 67 μg/liter. Pathologic examination revealed an endodermal sinus tumor. At laparotomy, in addition to the adnexal tumor and a uterus with myomata weighting 1540 g, no abnormalities or signs of tumor were found.

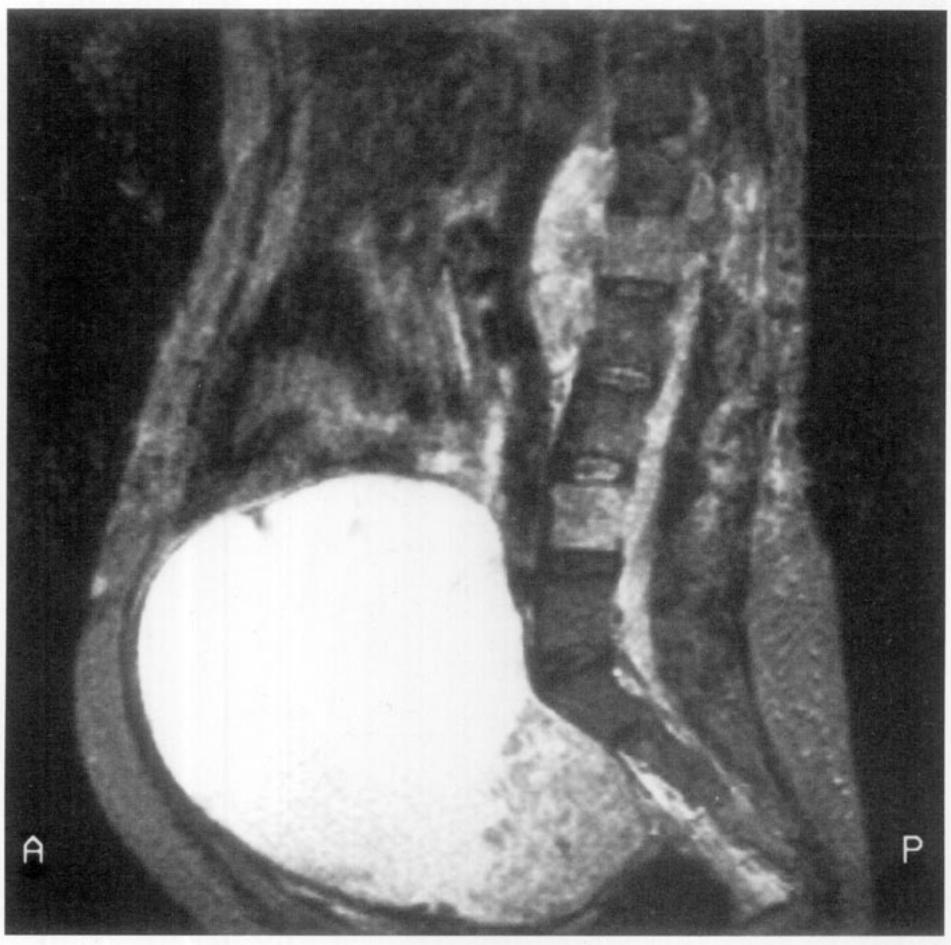

FIG. 2. Both the spinal metastases and the primary tumor are inhomogeneous on the T2-weighted SE sequence (TR 2760; TE 100). The primary tumor is largely cystic.

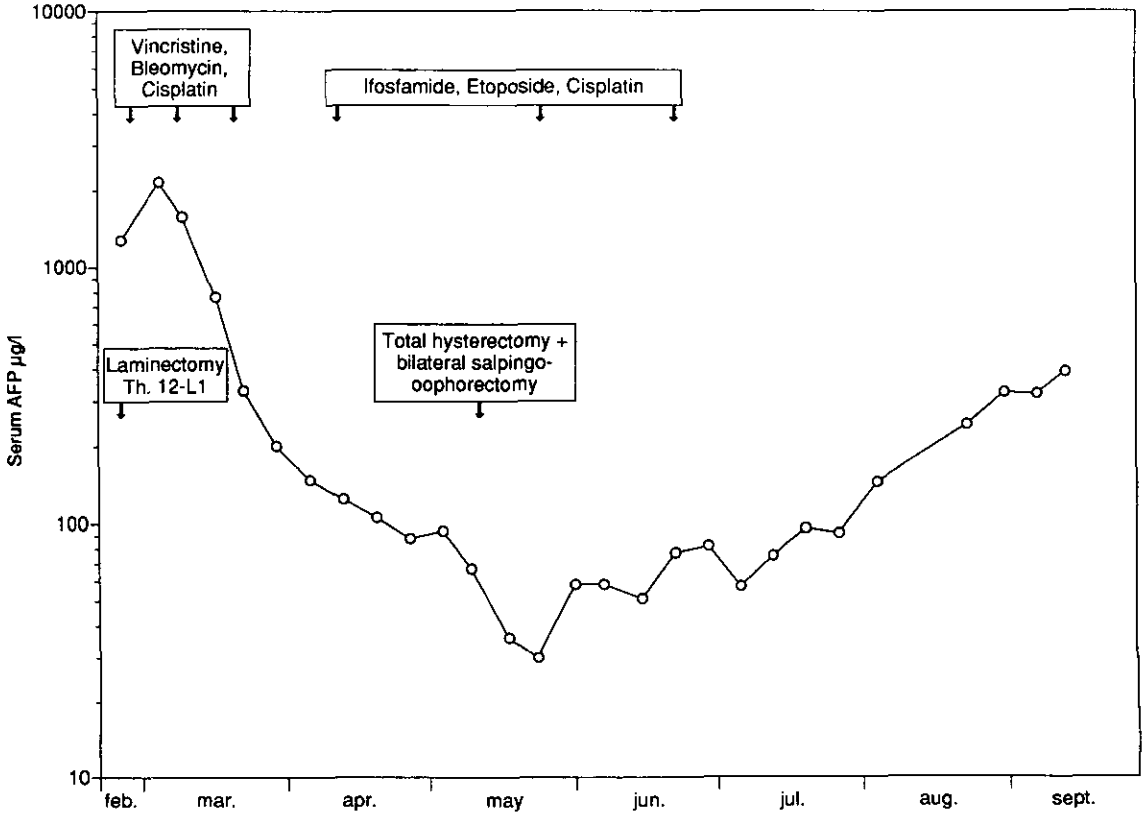

FIG. 3. Clinical course and serial serum-AFP values (logarithmic scale).

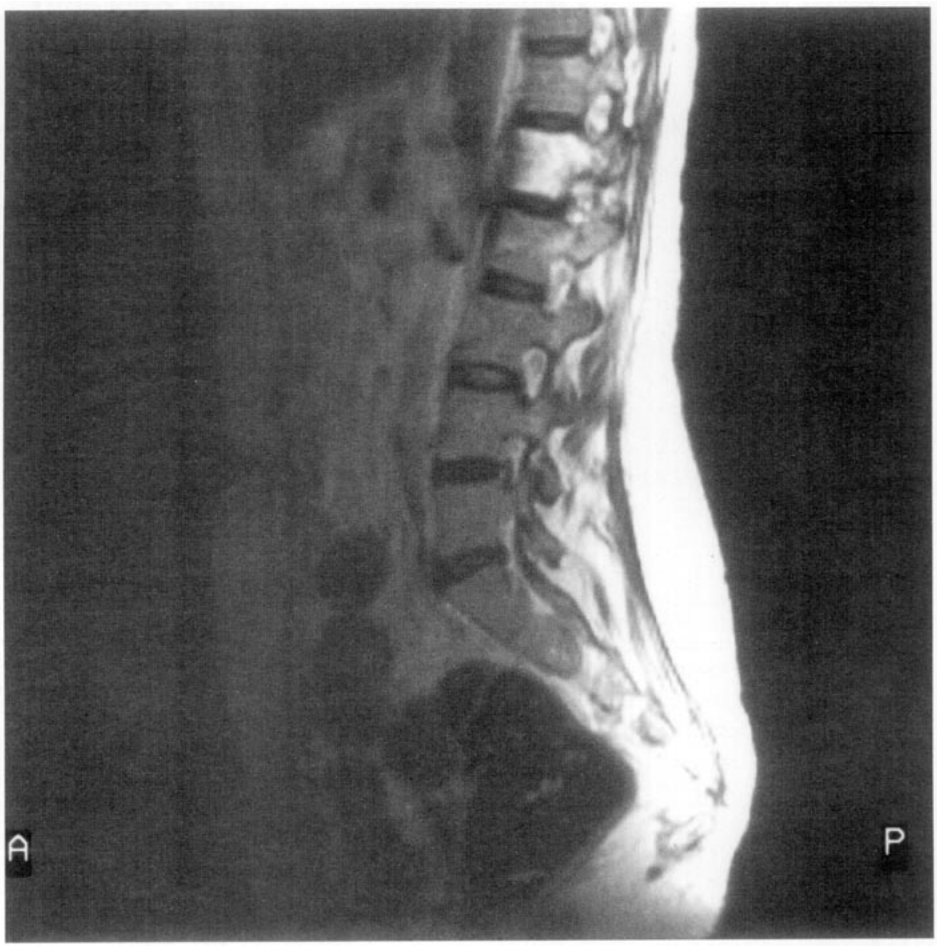

FIG. 4. After chemotherapy and surgical removal of the primary tumor the only abnormality is an inhomogeneous signal of L1 (TR 3120: TE 150; TSE image).

After the operation the same regimen of combination chemotherapy (cisplatin, ifosfamide, and etoposide) was continued. Despite therapy, serum-AFP resumed to increase after it had reached its lowest level at 30 μg/liter (Fig. 3).

A control MRI scan 1 month after surgery showed no evidence of remaining abdominal tumor or relapse (Fig. 4). The metastases in the vertebral column were unchanged, but a new metastasis in the acetabulum was seen.

The patient's condition deteriorated rapidly. The level of the serum-AFP continued to rise. Palliative radiotherapy on the vertebral column and pelvis was instituted for relief of pain. She died 7 months after onset of symptoms.

DISCUSSION

In about three-fourths of the patients with EST of the ovary, abdominal pain is a presenting symptom. One-third complain of an increase in abdominal girth due to a mass in the abdomen or pelvis. Fever is present in one-fourth and irregular vaginal bleeding in about 5% of the cases [1, 2, 5]. In more than half of the patients a palpable mass is present in the abdomen or pelvis which is asymptomatic in a few cases.

To the best of our knowledge, this is the first report of a patient presenting with a conus–cauda syndrome due to compression of the spinal cord by a metastasis of EST in a vertebral body. Metastases of EST of the ovary have been described predominantly in the liver, throughout the abdominal cavity, and in the retroperitoneal lymph nodes [1]. More distant metastases have been described in the lungs, in the supraclavicular lymph nodes, and in the brain. Bone metastases as in our case are very rare. Kurman and Norris reported a case with bone metastasis in the femur and in one case a metastasis in a vertebra [5].

In most cases of EST of the ovary the diagnosis is made by pathological examination of the primary tumor, which is available after laparotomy with uni- or bilateral salpingo-oophorectomy. In our case we acquired the pathological diagnosis through a biopsy of a metastasis. The metastases were not accessible for radical surgery and on the short term they were a serious threat to the well-being of the patient by compressing the spinal cord. We therefore chose to start combination chemotherapy immediately and postponed fur-

ther surgery until a reduction in tumor and metastases was achieved.

When the serum-AFP no longer decreased log-linearly, we considered resistance of the tumor to chemotherapy or a pool of AFP within the tumor which might cause a relatively high serum-AFP. On surgery we indeed found a high concentration of AFP within the tumor.

Notwithstanding the adverse outcome in our case we believe that in a case of EST stage IV with a metastasis compromising a vital function, it is preferable to postpone surgery and to start combination chemotherapy first because of the rapid growth of tumor and metastases.

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