A Subcutaneous Paraspinal Yolk Sac Tumor in a Child

Sibel A. Tekgündüz, MD,* Ceyhun Bozkurt, MD,* Gürses Şahin, MD,* Sema Apaydın, MD;† Ayşec. Ören, MD,* Eda Balkaya, MD,* and Ayşec.U. Ertem, MD*

Summary: A 3-year-old girl underwent a surgery at an external center on July 2011 for a swelling in the left lumbar paravertebral subcutaneous region. The mass was completely excised and the pathologic diagnosis was a yolk sac tumor (YST). Laboratory tests revealed a serum α-fetoprotein level of 278 IU/mL. Investigations using bone scintigraphy and magnetic resonance imaging revealed a scar tissue at the surgical site and lesions indicating metastasis at the lumbar first, second, third, and fifth vertebra. The patient was administered 5 cycles of PEB (cisplatin, etoposide, bleomycin) treatment. The serum α-fetoprotein level was 31 IU/mL after the pathology report. The lumbar magnetic resonance imaging and bone scintigraphy results were normal. The patient continues to be in remission since June 2012.

Key Words: yolk sac tumor, atypical localization, subcutaneous

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Yolk sac tumor (YST) is the most common malignant germ cell tumor of childhood.1 It is most commonly seen in the testis, ovary, and sacrococcygeal regions.2 It can also rarely be found in the pineal region, mediastinum, sinonasal region, cranial base, lungs, vagina, and penis. Our case is probably a YST with an atypical location derived from primary cells left under the skin because of a migration defect.

CASE REPORT

A girl aged 3 years and 2 months presented at an external center because of a swelling in the back in July 2011. Imaging studies revealed a left lumbar paravertebral subcutaneous mass (Fig. 1) and the mass was completely excised. The histopathologic report of the mass was hemangiomatous. The mass recurred at the same region a month later. The tumor mass was excised again in December 2011 and the pathologic report was a YST. As the 2 postoperative tumor specimens were not evaluated in our pathology department, we do not know whether margins were negative at resections. The patient was referred to our hospital for advanced investigation and treatment with this diagnosis. The findings on presentation were normal other than a 3-cm horizontal scar on the left paravertebral location in the lumbar region. The hemogram and biochemical analysis were normal at presentation, although evaluation at our clinic on January 2012 revealed that the serum LDH, AFP, and β-subunit of human chorionic gonadotropin levels were 195 IU/L, 278 IU/mL, and <0.5 mIU/mL, respectively. Imaging studies including posterior-anterior chest x-ray and abdominal ultrasonography were normal. Superficial tissue ultrasonography showed a 5 × 7 mm area with surrounding edema that was consistent with postoperative changes in the left lumbar paravertebral region. The bone scan revealed sclerosis of the S1-S2 vertebrae on the sagittal lateral view. We evaluated the pathologic specimens of both excisions. The most common histologic pattern was the glandular-alveolar pattern in the myxoid and a hypocellular background (Fig. 2A), with a few shiller-duval bodies. Immunohistochemical analysis revealed that there was diffuse staining with pankeratin and α-antitrypsin, focal staining with AFP (Fig. 2B) and staining with cytokeratin-7, consistent with the pure YST. The cranial, thoracic, and abdominal computed tomography imaging performed to find the primary mass and perform staging did not reveal any other mass. Bone scintigraphy revealed increased activity focally on both sides of the corpus in the L1 vertebra, with the left being more prominent, in the L2 and L3 vertebral corpuses, and the right half of the L5 vertebral corpus (Fig. 3). Moreover, there was increased activity in the joint femoral neck region compared with the other side. The right femur magnetic resonance (MR) image obtained to evaluate the enhanced regions on bone scintigraphy showed only minimal fluid increase. Lumbar MR image was compared with preoperative MR image and hypointense images were seen in the L1-L2-L3 and L5 vertebral bodies, indicating metastasis. After the investigations specified above, the patient received a diagnosis of metastatic extra-gonadal (primary lesion location: left lumbar paravertebral subcutaneous area over the muscle) YST due to the lumbar vertebra involvement. Although 4 to 6 cycles of PEB is regarded as the standard treatment of YST, according to the policy of our clinic, we use 5 cycles of PEB in patients presenting with metastatic YST. Therefore, chemotherapy treatment consisting of 5 cycles of PEB (cisplatin, etoposide, bleomycin) was planned. The MR imaging did not show a pathologic finding besides the scar tissue at the surgical site. Serum AFP level was 2.81 IU/mL (normal value, 0.5 to 5.51 IU/mL). The remission evaluation after 5 cycles of PEB treatment revealed normal bone scintigraphy. The findings using the MR imaging were also normal. The serum AFP level was 3 IU/mL (normal value, 0.5 to 5.51 IU/mL). The patient has been followed up in remission since June 2012.

DISCUSSION

Malignant germ cell tumors of the yolk sac type are most commonly seen in the testis, the ovary, and the sacrococcygeal region, but other cases with the primary lesion in the stomach, diaphragm, omentum, lungs, vagina, vulva, penis, heart, ears, mesentery, liver, thyroid, nasal region, eyes, floor of the mouth, masticator space, and cranial base have been reported.2–15 We did not come across any cases with an atypical subcutaneous tissue location in the literature. YSTs develop after malignant transformation of primordial germ cells in their midline locations during migration. We believe that the midline left paravertebral location of our case was subcutaneous because of a lack of involvement in this region.
migration defect. The vertebral lesions may have developed because of a local or lymphatic spread or the simultaneous localization of the primordial germ cells in the subcutaneous tissue and the vertebrae. Animal experiments have shown that this pathologic localization can be caused by abnormalities involving the soluble derived factor and the chemokine receptor CXCR-4 together with the stem cell receptor c-kit and the related ligand.¹ The origin of primary extragonadal germ cell tumors is unknown. Two possible explanations exist with regard to their source: (1) mis-integration of germ cells during embryologic development and (2) physiological distribution of germ cells to different organs of the body.²³ In conclusion, good results are obtained in YSTs with total excision and chemotherapy protocols containing platinum derivative–based drugs even when they are metastatic or in an atypical location.

REFERENCES

FIGURE 1. A and B, Subcutaneous mass with contrast enhancement on magnetic resonance imaging (white arrow) on axial plane and sagittal plane.


FIGURE 3. Image of lumbar vertebra involvement on technetium bone scan (white arrow).


