Latent adrenal Ewing sarcoma family of tumors: A case report

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Title: Latent adrenal Ewing sarcoma family of tumors: a case report

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Abstract

Ewing sarcoma family of tumors (ESFT) is derived from the neural crest, which originates from basal embryo cells in the primitive neural tube. ESFT often arises at the bone, chest wall, and soft tissues of the thoracic region. However, ESFT that arises from the adrenal gland is much rarer and it is usually revealed by clinical symptoms. We report an autopsy case of suicidal hanging, in which adrenal ESFT was incidentally revealed. To our knowledge, this is the first case of latent ESFT arising from the adrenal gland. Autopsy can sometimes reveal latent disease. Some of these latent diseases are very rare and we would not be able to detect them without a complete autopsy. As forensic pathologists, we should attempt to perform a complete autopsy and report new discoveries for the development of medicine.

Keywords: Ewing sarcoma family of tumors, Latent cancer, Adrenal gland tumor

1. Introduction

The Ewing sarcoma family of tumors (ESFT) is derived from the neural crest, which originates from basal embryo cells in the primitive neural tube. ESFT has a common set of
immunohistochemical and cytogenetic characteristics. ESFT is therefore classified as a family of molecularly and histopathologically related small, round, blue cell tumors. ESFT often arises in the bone, chest wall, and soft tissues of the thoracic region. However, ESFT that arises from the adrenal gland is rarer and is often associated with clinical symptoms [1].

We report an autopsy case of suicidal hanging, in which ESFT of the adrenal gland was incidentally revealed. To our knowledge, this is the first case of latent ESFT arising from the adrenal gland.

2. Case report

A 26-year-old man was found dead on the stairs of a hotel, hanging by the neck with his belt. The hanging was complete; the feet had not landed on the floor, with the knot of the ligature in the upper, posterior part of the head. He had depression and left a suicide note. With case investigation, the manner of death was thought to be suicidal.

3. Autopsy findings

The deceased was 169 cm tall and weighed 61.0 kg. There was no external injury
except for around the neck. On examination of the neck, a furrow was present, which did not completely encircle the neck. Focal cutaneous hemorrhages and blisters were present above and below the furrow. The skin, underlying fat, and soft tissue in the area of the ligature marks were dark brown and dry.

Subpleural petechial hemorrhages, acute lung emphysema, and liquid blood in the heart were found on internal examination, indicating general autopsy signs of asphyxia. Intra-muscle hemorrhages of the neck were revealed and the hyoid bone was fractured. The weight of the left and right lung was 532 and 636 g, respectively. We also found an 8-cm diameter, whitish-yellow tumor in the right retroperitoneum. The tumor was completely separated from right kidney (Fig. 1). No other pathological change was found.

On microscopic examination, the tumor was composed of malignant, small, round cells (Fig. 2A). As the tumor was located in the right adrenal area and had a cortical rim (Fig. 3), the tumor was presumed to have arisen from the adrenal gland. Immunohistologically, the tumor was positively stained for Cluster of Differentiation 99 (CD99) (Fig. 2B) and vimentin, but negatively for synaptophysin, desmin, WT1, CD3, CD20, and CD34. Therefore, the cause of death was hanging and a latent ESFT of the adrenal gland was diagnosed.

4. Discussion
ESFT includes Ewing sarcoma of the bone, extraskeletal Ewing sarcoma, peripheral primitive neuroectodermal tumor (PNET), and Askin tumor (thoracopulmonary PNET) [2]. PNET was first described in 1918 by Stout in a 42-year-old man with ulnar nerve tumors. Then, in 1921, Ewing sarcoma was described in a 14-year-old boy with osteoclasia in the ulna. In 1973, both tumors were defined as PNETs. In 1979, Askin tumors were defined as PNETs in the thoracic cavity [1]. They are, now, collectively termed as ESFT. ESFT is always derived from the primitive neural crest, which originates from basal embryo cells in the primitive neural tube.

In the present case, the tumor was located in the right retroperitoneum. It was completely separated from the right kidney and had a cortical rim, suggesting a primary adrenal tumor. Malignant, small, round cells include neuroblastoma, rhabdomyosarcoma, Wilms tumor, desmoplastic small round-cell tumor, malignant lymphoma solitary fibrous tumor, or ESFT during differential diagnosis. Immunohistochemical examination in our case revealed that the tumor was positively stained with CD99, which is consistent with ESFT. The other tumors are ruled out by negative staining for synaptophysin, desmin, WT1, or CD34. We therefore diagnosed primary adrenal ESFT on the basis of pathological examination. ESFT of adrenal gland is a very rare disease and only a few cases have been
reported so far [1,3-11]. Molecular analysis of ESFT reveals a reciprocal translocation of t
(11; 22) (q24; q12) and t (21; 22) (q22; q12) [12]. In the present case, however, we could not
perform molecular analysis because of postmortem changes. We should have preserved a
frozen sample of the tumor.

In the previously descriptions of adrenal ESFT in the literature (Table 1), most
patients had complained of abdominal pain and the tumors were subsequently discovered.
ESFT progresses rapidly and some patients already had metastasis by the time the primary
tumor was found. However, in the present case, the deceased had no symptoms and did not
notice that he had the tumor. Autopsy incidentally revealed the tumor and such a case is
very uncommon. To our knowledge, this is the first report of latent adrenal ESFT.

With respect to determination of the cause of death, especially in cases of trauma,
we may be able to detect the cause of death using only external examination, as in the
present case of hanging. However, autopsy can sometimes reveal latent disease [13]. Some
of these latent diseases are very rare and we would not be able to detect them without a
complete autopsy. As forensic pathologists, we should attempt to perform a complete
autopsy and report new discoveries for the development of medicine.

5. Conflict of interest
The authors have declared no conflict of interest.

References


**Figure legends**

Fig. 1. Macroscopic examination of the kidney. An 8-cm diameter, whitish-yellow tumor was found in the right retroperitoneum (A). The tumor was completely separated from the right kidney (B).

Fig. 2. Microscopic examination of the tumor. The tumor was composed of malignant, small, round cells (A, H&E staining) and stained positively for Cluster of Differentiation 99 (CD99) (B, CD99 staining).

Fig. 3. Microscopic examination of the tumor. The tumor was located in the right adrenal area and had a cortical rim. (H&E staining)
Figure 2

A

B
<table>
<thead>
<tr>
<th>Authors [reference]</th>
<th>Age (yr)/sex</th>
<th>Outcome*</th>
<th>CD99</th>
<th>Clinical symptoms</th>
<th>Tumor size</th>
</tr>
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<tr>
<td>Marina et al. [3]</td>
<td>17/F</td>
<td>Dead</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td></td>
<td>7/M</td>
<td>Dead</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
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<tr>
<td></td>
<td>4/M</td>
<td>Dead</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Renshaw et al. [4]</td>
<td>46/F</td>
<td>NR</td>
<td>+</td>
<td>NR</td>
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<tr>
<td></td>
<td>20/F</td>
<td>NR</td>
<td>+</td>
<td>NR</td>
<td>NR</td>
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<tr>
<td></td>
<td>48/F</td>
<td>NR</td>
<td>+</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Matsuoka et al. [5]</td>
<td>32/F</td>
<td>Dead at 5 mo.</td>
<td>+</td>
<td>Abdominal pain</td>
<td>10 cm</td>
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<tr>
<td>Pirani et al. [6]</td>
<td>57/M</td>
<td>NR</td>
<td>+</td>
<td>Lower extremity pain</td>
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<tr>
<td>Kato et al. [7]</td>
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<td>Dead at 15 mo.</td>
<td>+</td>
<td>NR</td>
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<tr>
<td>Komatsu et al. [8]</td>
<td>53/F</td>
<td>Alive at 10 mo.</td>
<td>+</td>
<td>Health screening</td>
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<td>NR</td>
<td>+</td>
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<td>10 cm</td>
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<td>Age</td>
<td>Gender</td>
<td>Additional Condition</td>
<td>Cause of Death</td>
<td>Tumor Size</td>
</tr>
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<tr>
<td>Kim et al. [10]</td>
<td></td>
<td></td>
<td></td>
<td>NR + Abdominal pain</td>
<td>15 cm</td>
</tr>
<tr>
<td></td>
<td>25</td>
<td>F</td>
<td>+</td>
<td></td>
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</tr>
<tr>
<td></td>
<td>24</td>
<td>F</td>
<td>+</td>
<td>Flank pain</td>
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</tr>
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<td>Zhang &amp; Li [1]</td>
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</tr>
<tr>
<td></td>
<td>30</td>
<td>M</td>
<td>+</td>
<td></td>
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<tr>
<td></td>
<td>21</td>
<td>F</td>
<td></td>
<td>Dead at 6 mo.</td>
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</tr>
<tr>
<td></td>
<td>24</td>
<td>F</td>
<td></td>
<td>Alive at 13 mo.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>22</td>
<td>M</td>
<td></td>
<td>Alive at 1 mo.</td>
<td>17 cm</td>
</tr>
<tr>
<td>Stephenson et al. [11]</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>17</td>
<td>F</td>
<td>+</td>
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<td></td>
<td></td>
<td>Dead</td>
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<tr>
<td></td>
<td>26</td>
<td>M</td>
<td>+</td>
<td>Latent cancer</td>
<td></td>
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</table>

*Time after initial presentation

**Abbreviations:** F, female; M, male; mo., months NR, not recorded