Synovial sarcoma of the spine: a case involving paraspinal muscle with extensive calcification and the surgical consideration in treatment

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Abstract

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Introduction Synovial sarcoma is a rare malignant tumor of the spine. This tumor may present as a painless mass of the spine or slowly enlarge, causing pain or neurologic deficits. As it is difficult to differentiate this lesion from other soft tissue tumors, synovial sarcoma requires histologic confirmation for definite diagnosis. Thus, the treatment strategy is often planned in the final step depending on the pathologic results. Despite its rare incidence, a few cases of primary or metastatic synovial sarcoma involving the spinal cord, foramen, vertebral body, or paraspinal muscles have been reported in the literature.

Materials and methods We present the case of a 29-year-old man with a synovial sarcoma in the paraspinal muscle of the cervical spine. The patient was evaluated radiologically and histologically. Plain radiography, computed tomography, and magnetic resonance imaging were performed as part of the preoperative workup, and immunohistochemical and cytogenetic studies were additionally performed to identify the histologic features of the tumor. The patient underwent marginal resection followed by adjuvant radiation therapy. The patient has been followed up for 2 years.

Conclusions This article highlights the features of synovial sarcoma of the spine via a comprehensive review. Synovial sarcoma of the spine is uncommon, but it is a challenging issue in both diagnostic and therapeutic aspects. The currently available evidence suggests the use of a multidisciplinary approach in the treatment of synovial sarcoma, which includes complete resection and radiation therapy.

Keywords Synovial sarcoma · Cervical spine · Paraspinal muscle

Case presentation

A 29-year-old man visited the emergency department of our hospital with a complaint of an enlarging mass in the right posterior neck accompanied by right arm pain and limited range of motion. He mentioned that the palpable neck mass developed 5 years ago with no symptoms. There was no significant history of trauma. On physical examination, the neck mass was hard and tense without pain or warmth. Neither specific signs nor abnormal findings were noted on neurologic examination. Results of a complete blood count and chemistry, electrolyte, and other laboratory tests were normal.

Diagnostic imaging section

On initial plain radiography of the neck taken in the anteroposterior (Fig. 1a) and lateral views (b), a large ossified round mass was found at the right occipitocervical junction (Fig. 1). On spine magnetic resonance imaging (MRI), a $6.6 \times 5.6 \times 4.0$ cm lobulated mass was located posterior to the spinous process

of the C2–3 spine, and the mass extended to the contralateral paravertebral space over the middle line (Fig. 2). The lesion had heterogeneous components with extensive, dense, and stippled calcification, highly suggestive of a bone or cartilage-forming tumor such as extra-skeletal Ewing sarcoma and other unusual soft tissue sarcomas. There was no definite evidence of bone invasion. The patient underwent CT-guided percutaneous needle biopsy, and the tumor was confirmed to be a calcified synovial sarcoma by permanent biopsy.

Historical review of synovial sarcoma

Synovial sarcoma, a highly malignant tumor, is a specific type of soft tissue sarcoma that accounts for 5–10 % of soft tissue sarcomas. Although synovial sarcoma does not originate from the synovial tissue, the term arose from its similar microscopic features to the developing synovium [3]. Synovial sarcoma can be found in any anatomic site of the body, but it mainly develops in deep soft tissues adjacent to joints or the tendon sheaths of the extremities. Synovial

Fig. 1 Plain radiograph of the neck taken in the anteroposterior (a) and lateral views (b). Preoperative computed tomography scans revealing a lobulated paravertebral space mass with massive

calcification at the right occipitocervical junction on axial (c) and sagittal reconstruction (d) imaging

Fig. 2 Preoperative magnetic resonance imaging of the cervical spine. A T2-weighted image in the sagittal view (a) and T1-enhanced weighted images in the sagittal (b) and axial (c) views

sarcoma is specifically predominant in young adults with a peak incidence in the third decade of life [3, 7].

The radiological features of synovial sarcoma are usually nonspecific. On plain radiographs, calcification is found in up to 30 % of synovial sarcomas, of which those with extensive calcification have a favorable prognosis [9]. On MRI, synovial sarcoma is often seen as a heterogeneous mass presenting both solid and cystic components of hemorrhage and fibrous tissues [6]. As synovial sarcomas always have well-defined margins, tumors in the head and neck region are difficult to distinguish from other benign or malignant tumors [13].

Histologically, synovial sarcoma is classified into three groups: biphasic type, monophasic type, and poorly differentiated type. The tumor consists of round epithelial cells, fibrous spindle cells, or both. Because of its various histologic findings, synovial sarcoma is often confused with other mesenchymal tumors. Immunohistochemical and cytogenetic studies are needed to distinguish synovial sarcoma from other malignant soft tissue tumors. In the present case, the tumor contained biphasic cell components on microscopy and exhibited high cellularity and a high Ki-67 labeling index (13.59 %). On immunohistochemistry, the tumor cells were positive for a specific epithelial marker (cytokeratin AE1/AE3) and Ewing sarcoma marker (CD99). On cytogenetic analysis, SYT gene rearrangement was detected, supporting the diagnosis of synovial sarcoma (Fig. 3). The recurrent translocation $t(X:18)(p11.2;q11.2)$ is observed in ~90 % of synovial sarcomas. However, the association between the fusion subtype and clinical outcome is unclear, and the histologic grade of tumors might be a more important factor for prognosis [3, 4, 8].

Rationale for treatment

No consensus on the optimal treatment for synovial sarcoma has been established. Complete surgical resection with adjuvant radiotherapy and/or chemotherapy is the currently recommended treatment for synovial sarcoma [3, 8]. Although wide resection with negative surgical margins is a primary principle of surgery for high-grade tumors, marginal resection is occasionally preferred to preserve the adjacent structures with minimal injuries for a better functional outcome and improved patient quality of life. The spinal region in the posterior neck does not contain sufficient soft tissue, and hence, occasionally, requires local flaps after wide resection.

For cases with a high risk of local recurrence, postoperative radiotherapy is recommended. It has been documented that microscopically positive margins are acceptable for adjuvant radiation therapy, whereas additional gross resection has no obvious benefits [10]. Conversely, the role of

Fig. 3 Microscopic features of a permanent biopsy specimen. **a** The tissue section comprised a uniform population of short spindle cells with mild nuclear atypia, no necrosis, and a low mitotic rate; a focal hemangiopericytoma-like pattern was also present (hematoxylin-eosin staining, original magnification, $\times 100$). **b** Representative sample of break-apart fluorescence in situ hybridization assay for SYT(18q11.2) rearrangement. In a normal allele, the signals are fused and they appear to be yellow in color (yellow arrows); conversely, one orange and one green break-apart signal pattern per nucleus is indicative of a rearrangement in one copy of the SYT gene region (green and red arrows)

chemotherapy in the treatment of synovial sarcoma is controversial. Several chemotherapeutic agents including doxorubicin and ifosfamide are currently used in the treatment of soft tissue sarcomas [2, 3, 8]. Some studies consistently documented that the administration of chemotherapy improves survival in certain groups of candidates [1, 2].

Fig. 4 Follow-up magnetic resonance imaging of the cervical spine 2 years after surgery. A T2-weighted image in the sagittal view (a) and enhanced T1-weighted images in the sagittal (b) and axial (c) views

Despite treatment with various modalities, synovial sarcoma has the worst prognosis among soft tissue sarcomas. The 5- and 10-year survival rates for patients with synovial sarcoma are 75 and 50 %, respectively [11]. Local recurrence occurs in up to 30 % of patients, whereas distant metastasis is noted in 50 % of the patients. Patients with metastatic disease have a poor prognosis, with a survival of <2 years [3]. The good prognostic factors include age (≤ 20 years), tumor size (< 5 cm), a negative surgical margin after complete resection, and the administration of adjuvant radiotherapy [10, 12]. In the head and neck region, tumors located in the skull base or paraspinal muscles have poor prognoses [5]. Other factors such as histologic grade, cytogenetic subtype, and specific radiologic findings have also been referred to in several studies [1, 4, 13].

Procedure

The patient was placed in the prone position, and his head was fixed with the three-point pin system; a midline vertical skin incision was made from theinion to the C6 spinous process with the neck in a neutral posture. The tumor was located under the muscles of the posterior neck, which were very thin. We completely excised the tumor including the biopsy track. The plane between the semispinalis capitis and suboccipital muscles was dissected to expose the mass. The mass was pinkish white in color without significant adhesion to the surrounding muscle. There was a reactive thin membrane between the soft tissue and the tumor. Frozen-section analysis indicated a diagnosis of sarcoma. The mass was excised after removing the tip of the C2 spinous process with a chisel because adhesions between the mass and the C2 spinous process were observed.

Finally, a $9.0 \times 6.0 \times 4.5$ cm firm mass was carefully removed en bloc with negative margins. The operation was

completed without complications. There was no evidence of a residual lesion or unusual postoperative findings on MRI (Fig. 4). Several postoperative studies were performed for staging to plan further management. Whole-body 18F-fluorodeoxyglucose positron emission tomography-CT revealed no other active primary or metastatic lesions. No definite evidence of a metastatic lesion was found on chest radiography and CT. Finally, the tumor stage was classified as IIb (T2b, N0, M0, G2) according to the seventh American Joint Committee on Cancer staging system for soft tissue sarcoma. The patient underwent adjuvant radiation therapy with a total dose of 6,000 cGy. Chemotherapy was not administered in this case.

Outcome

Cervical spine MRI and chest CT were performed every 6 months. No local recurrence or metastatic lesion was detected. The patient has remained disease free for the last 2 years. In this case, a combination of marginal resection and adjuvant radiation therapy was successful in achieving local control. The favorable prognosis might be related to certain characteristics of the tumor, including extensive calcification.

Conflict of interest The authors declare that they have no competing interests.

Consent Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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