Synovial sarcoma: a rare presentation of parapharyngeal mass

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Abstract Synovial sarcoma is a rare soft tissue sarcoma of the head and neck region involving the parapharyngeal space. The diagnosis of synovial sarcoma can be very challenging to the pathologists. We present a rare case of parapharyngeal synovial sarcoma in a young female patient who had a two-month history of left cervical intumescent mass at level II. The fine needle aspiration cytology of the mass was proved inconclusive. Transcervical excision of the mass was performed and the first case of parapharyngeal sarcoma was identified in our center by fluorescence in situ hybridization (FISH) technique. Repeat imaging revealed residual tumor. The patient successfully underwent a second excision of the residual tumor and received adjuvant radiotherapy.

Keywords: Synovial sarcoma, parapharyngeal space, fluorescence in situ hybridization (FISH)

Synovial sarcoma is a malignant tumor that frequently affects the extremities, especially around the joints, bursae and tendon sheath[1,2]. It has also been described in other locations including the head and neck region, mediastinum, heart, abdominal wall, mesentery, retroperitoneum and peritoneal cavity. It accounts for about 6.8% of head and neck tumors[3] with hypopharynx being the commonest site[4]. However, it also can occur in the pharynx, tongue, tonsils and orofacial soft tissues[5]. The diagnosis of synovial sarcoma requires an integrated multidisciplinary approach. Complete surgical excision followed by radiotherapy is advocated to achieve better prognosis[6].

Case Description

A 27-year-old woman presented with 2-month history of painless progressive left cervical intumescent mass. Local examination revealed a firm, well defined, and mobile level II mass measuring 3.5 cm × 4.5 cm with smooth surface (Figure 1). Ear, nose, throat and systemic examinations revealed normal findings. Contrast enhanced computed tomography (CT) scan showed a level II mass with heterogeneous enhancement in the left parapharyngeal space with medial displacement of the cornu of the hyoid bone, anterior displacement of the submandibular gland and medialisation of the adjacent hypopharyngeal wall (Figure 2). The fine needle aspiration cytology obtained cellular aspirate which was difficult to interpret.

Figure 1 Left lateral cervical intumescent mass at level II measuring 3.5 cm × 4.5 cm

Two weeks later, a transcervical excision of the tumor was performed. Intraoperative findings revealed an encapsulated mass located lateral to the carotid sheath, extending from the...
submandibular area down to the level of the superior thyroid cartilage. The tumor was completely removed. Histopathologic examination showed a biphasic pattern of glands and spindle cells (Figure 3). The final diagnosis was confirmed by fluorescence in situ hybridization (FISH) which showed translocation of the SYG (ss18) gene (dual color break apart probe 18q11.2) after further testing at another institute which we had consulted.

On CT scan or magnetic resonance imaging (MRI) scan, synovial sarcoma usually demonstrates features of homogenous solid mass. However, it may present heterogenous signals based on the degree of hemorrhage, necrosis or calcification[9]. Recent study by Tateishi et al.[2] showed that CT and MRI scans allow prognostic prediction in patients with synovial sarcoma. Proximal distribution, tumor size larger than 5 cm, absence of calcification, presence of hemorrhage and presence of triple signal pattern are significantly associated with disease-free survival rate. Penel et al.[8] reported that poor prognostic factors of synovial sarcoma include initial metastasis or presence of lymphadenopathy, absence of surgery and number of surgical procedures. Other studies had shown that tumor size, histological type, status of surgical margins and anatomical location also play important roles in predicting local recurrence[2,4]. In our case, histopathologic examination showed the epithelial component was more poorly differentiated than usual, possibly suggesting a poorer prognosis which led to recurrence of the disease.

The differential diagnosis between synovial sarcoma with other benign or malignant sarcomas such as rhabdomyosarcoma, fibrosarcoma or lymphoma is difficult to make clinically or radiologically. Tissue biopsy is still necessary for confirmative diagnosis of synovial sarcoma. Histopathologic diagnosis by fine needle aspiration is difficult because most malignant parapharyngeal tumors have varied histopathologic types[8]. Therefore, it is crucial to obtain adequate tissue sampling from parapharyngeal space adjacent to foramen ovale with enlarged level III nodes. She underwent further tumor excision with modified radical dissection on the left cervical lymph nodes. The tumor had infiltrated the sternocleidomastoid muscle and adhered to the carotid sheath with multiple nodes at the levels I, II and III. Four out of 20 lymph nodes at different levels were found to have tumor cell deposits. One of the surgical margins in level III was involved by the tumor. Therefore, the patient was subjected to adjuvant radiotherapy. She had completed 33 fractions of radiotherapy. During 18-month follow-up, there was no evidence of local recurrence or distant metastases.

**Discussion**

Synovial sarcoma is a rare but one of the most common types of malignant non-rhabdomyosarcomatous soft tissue sarcoma[7]. Guadagnolo et al.[1] reported a large series of 150 cases of synovial sarcoma and only 9 involving the head and neck region. In the head and neck region, synovial sarcoma originates from primitive mesenchyma with no synovial association[5,8]. Patients usually present a painless slow growing mass during their third and fourth decade of life with associated compressive or infiltrative symptom of surrounding structures[7]. Consistent with the literature, our patient presented with painless left cervical intumescent mass, however, without compressive or infiltrative symptoms. Some of the predisposing factors to development of sarcoma include genetic factors, viral infection in immunocompromised patients, irradiation, chronic lymphoedema, environmental carcinogens and trauma[8].

The patient was well until five months later when she presented with recurrent small cervical intumescent mass adjacent to the previous incision wound. She denied any other symptoms. Repeat scan showed residual mass within the left parapharyngeal space adjacent to foramen ovale with enlarged level III nodes. She underwent further tumor excision with modified radical dissection on the left cervical lymph nodes. The tumor had infiltrated the sternocleidomastoid muscle and adhered to the carotid sheath with multiple nodes at the levels I, II and III. Four out of 20 lymph nodes at different levels were found to have tumor cell deposits. One of the surgical margins in level III was involved by the tumor. Therefore, the patient was subjected to adjuvant radiotherapy. She had completed 33 fractions of radiotherapy. During 18-month follow-up, there was no evidence of local recurrence or distant metastases.

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**Figure 2** Contrast enhanced computed tomography scan demonstrates a heterogeneously enhanced mass (arrow) at level II within the left parapharyngeal space causing minimal medial displacement of the left cornu of the hyoid bone and anterior displacement of the submandibular gland.

**Figure 3** Histopathologic examination shows a biphasic pattern of glands and spindle cells (HE x100)
fine needle aspiration for accurate diagnosis which at the same time avoids delay in managing this malignant disease.

Histopathologically, synovial sarcoma is characterized by presence of epithelial and spindle cells with various different histopathologic subtypes. The most recognized subtypes are monophasic and biphasic variants. The classic monophasic subtype has a single cellular component whereas the biphasic subtype is composed of distinctive spindle and epithelial cells similar to our case.

The definitive diagnosis of synovial sarcoma is made by using FISH. The detection of a specific translocation between chromosome X and 18, t(X;18) (p11.2;q11.2) is important in confirming the diagnosis of synovial sarcoma especially when histological studies are ambiguous. FISH using a breakapart style probe, as performed in our case, is quick, sensitive and specific than commercially available FISH, or chromogenic in situ hybridization as reported by Terry et al. Sun et al. also reported that the efficiency of FISH is comparable to or even higher than that of reverse transcription-polymerase chain reaction for SYT-SSX detection.

Combined modality therapy is advocated to yield better prognosis. Guadagnolo et al. reported that conservative surgery and radiotherapy are adequate to control the primary tumor. Localized synovial sarcoma treated with conservative surgery and radiotherapy had overall 5- and 10-year survival rates of 76% and 57% , respectively. On the other hand, aggressive surgical excision followed by postoperative radiotherapy is believed to obtain better prognosis, as in our case. Inadequate surgical resection was most probably the reason for initial treatment failure causing her to require a second operation.

In synovial sarcoma of the head and neck region, cervical metastasis is reported to occur in 10%-20% of the patients. Cervical lymphadenectomy is recommended for the patients with positive cervical lymph nodes. However, it is still controversial whether elective treatment of the uninvolved neck would be valuable. Lung metastasis is the usual cause of death in synovial sarcoma patients as reported by Kartha et al. Therefore, regular plain chest radiograph is recommended to detect lung metastasis.

In conclusion, the initial diagnosis and management of synovial sarcoma is challenging due to rarity of this tumor and lack of large clinical experience. We conclude that complete surgical resection followed by radiotherapy should be performed in patients with localized synovial sarcoma.

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References