

Case Report

Relapse of synovial sarcoma in head and neck after a six-year disease-free period: a case report and literature review

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Abstract: Synovial sarcoma is a malignant mesenchymal tumor that is rarely found in head and neck region. Although recurrence of synovial sarcoma has been reported, relapse of tumor after a long time of disease-free period is less common in head and neck. We present a case of a seventeen-year old male who revisited after a 6-year of disease-free period following resection of synovial sarcoma for neck mass. The tumor grew beneath the sternocleidomastoid muscle, and pushing the carotid sheath outward that made it present as an “olive-like” appearance. Tumor resection and neck dissection were performed. And the carotid vessel affected by tumor was preserved during tumor resection. The patient received closely follow-ups in eighteen months after surgery and no evidence of further relapse were yet found. In addition, the clinical features, histopathology and treatment of synovial sarcoma were reviewed with a focus on their manifestation and management in head and neck.

Keywords: Relapse, synovial sarcoma, head and neck, immunostaining, resection

Introduction

Synovial sarcoma (SS) is a malignant mesenchymal neoplasm, accounting for 8-10% of all soft tissue sarcoma, which is most commonly found in the extremities. Occurrence, in head and neck, a poor location for synovial tissue, is uncommon and it is estimated no more than 200 cases has been reported worldwide since the first one was reported in pharyngeal in 1954 [1-5]. Reported sites of synovial sarcoma in head and neck include oropharynx, larynx, hypopharynx, oral cavity, parotid gland, lateral neck, jaws and scalp [2, 3, 5-7]. The most reported common site is the hypopharynx [1, 3, 8].

Although synovial sarcoma is considered as an aggressive tumor, the overall survival in five-year can reach >90% in children and adolescents [9]. The prognosis of SS varies on tumor histological subtype, location, size, and more important on surgical margin [1, 9, 10]. The

relapse or metastasis of synovial sarcoma is also seen in head and neck, but it usually occurs within the first two years after initial treatment [1, 6, 9, 11]. Here we report a relapse case 6-year after initial surgical excision in left lateral cervical region, and relapse in the contralateral neck with extremely extrusion of the carotid artery and vertebral artery.

Case report

The patient is now a 17-year-old teenager who was seen at our institution in 2008 with left lateral neck mass. He received a surgical excision and the postsurgical pathologic diagnosis was synovial sarcoma. Although receiving some traditional herb treatment after surgery, he didn't undergo any adjuvant chemo or radiotherapy. The patient was deemed disease-free until six years later when he presented with a gradually-increasing mass in right lateral neck. Ten month after he noted a mass in right neck, it was gradually growing into a fist-like size when he revis-

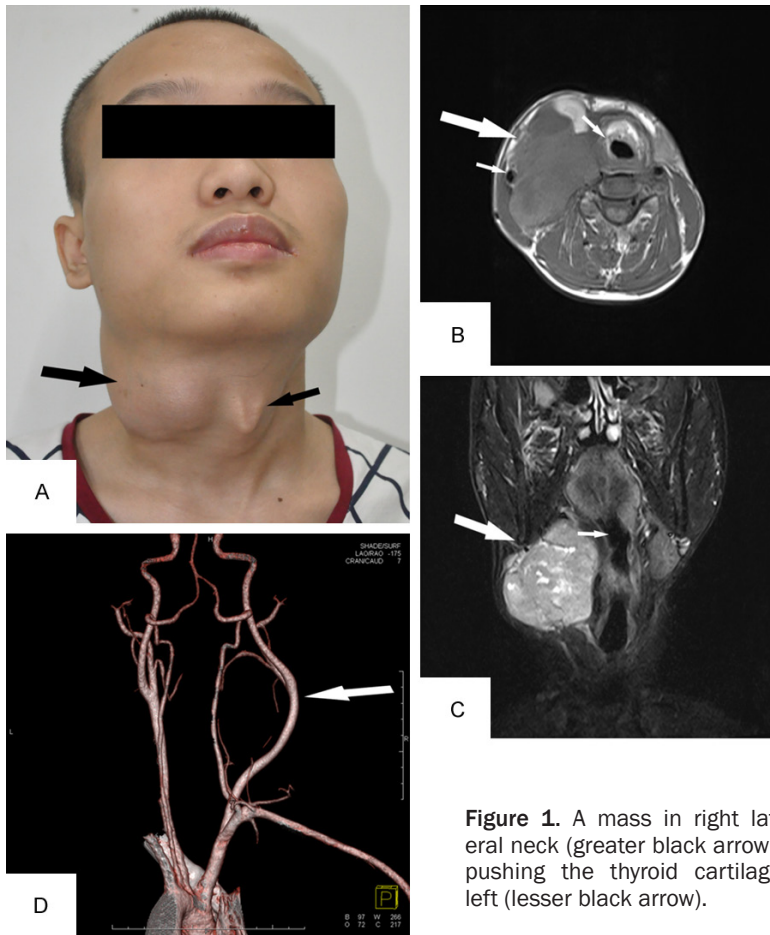


Figure 1. A mass in right lateral neck (greater black arrow), pushing the thyroid cartilage left (lesser black arrow).

ited us. Physical examination revealed a palpable mass in right lateral neck with a size of about 8*6 cm at its extreme axis. The mass underlies the sternocleidomastoid, with medial to anterior cervical median line, pushing the trachea left about 1.0 cm; up to mastoid and mandibular gland level, downward to the middle and lower third of sternocleidomastoid, lateral to posterior margin of sternocleidomastoid (**Figure 1**). It palpates a middle-hard texture, a little tenderness, non-pushable, unchangeable with location test, and unmovable with deglutition. On the surface of mass, vascular pulsation was palpated. He denied dyspnea, dysphagia, and hoarseness.

Magnetic resonance imaging (MRI) showed an 8*6*5 cm prevertebral mass, with well-defined boundary, underlying the sternocleidomastoid in right neck, compressing carotid sheath on the mass surface and pushing the airway to left shift (**Figure 2A**). Contrast-enhanced scanning revealed a heterogeneous enhancement in the

majority of the lesion (**Figure 2B**). Computed tomographic angiography (CTA) showed the right carotid artery was pushed outward by the lesion, presenting an “olive-like” change with the vertebral artery (**Figure 2C**). According to his physical presentation, characteristics of MRI, and his previous neck lesion, a relapse of synovial sarcoma was highly suspected.

Based on the highly suspected relapse of synovial sarcoma, surgical resection was subsequently performed. Due to the well-bordered and the limited space of neck, excision of the tumor with local neck dissection (II, III, and IV) was enough. However, operation was not too easy, how to deal with the great vessel of carotid sheath was extremely important. Based on the fact that the tumor was surrounded by a capsule reflected by MR

and CTA imaging, we believed it was doable to peel off the carotid sheath from tumor. Thus, more attention was paid to separate the great vessel from tumor in surgery, and we made it. Besides the palpable mass, several small lymph nodes were seen in the upper right neck region, the greater was nearly 1.5 cm in size. The tumor beneath the sternocleidomastoid and carotid sheath; the latter was pushed outward and was not conglutinated with tumor. Measured in size of 7*5*5 cm, the tumor was gray and soft (**Figure 3A**), with an off-white section and fish-meat like appearance (**Figure 3B**).

Routine hematoxylin eosin (H&E) stained microscopic sections showed the tumor was composed of round cells and spindle cells (**Figure 4A, 4B**), and no positive lymph node. Spindle-cells are arranged in fascicles or sheets with moderate nuclear pleomorphism (**Figure 4A**), while epithelial round-cells in glandular structures (**Figure 4B**). Immunohistochemical analysis showed a positive expression of CK-pan,



Figure 2. Preoperative MRI, axial view (A) shows a mass underlying sternocleidomastoid (greater arrow), pushing the airway (lesser arrow in both A and B) and compressing the carotid artery (lesser arrow); Coronal view (B) after contrast enhancement shows heterogeneous in the majority of mass (greater arrow), and the wall of airway was pushed (lesser arrow). Preoperative CTA (C) shows the carotid artery was pushed outward, presenting “olive-like” appearance with the vertebral artery (arrow).

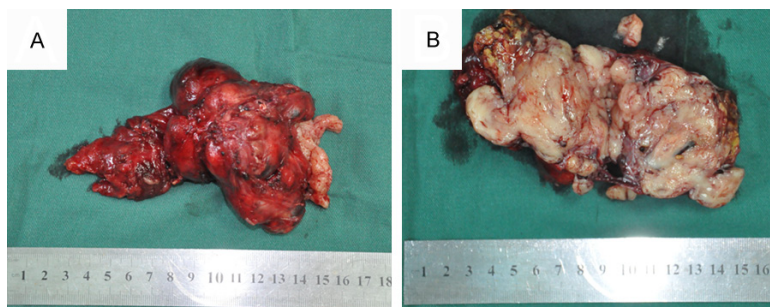


Figure 3. A. The photograph of resected tumor. B. The photograph of bisected tumor, presenting a “fish meat-like” appearance.

vimentin, EMA, CD99 and Ki-67 (5%-10%+), while S-100 was entirely negative (Figure 4C-H).

Patient denied any postoperative adjuvant therapy of radio/chemotherapy, excepting the regular follow-up at our institution. Until now, the patient is disease-free more than eighteen month.

Discussion

Sarcoma constitutes 4%-10% of malignancies in head and neck, higher than its total proportion of all malignancy in the body [2]; however, for synovial sarcoma, head and neck is a very uncommon site [2, 3, 9]. More than 70% of synovial sarcoma occurs in limbs, and only 1.9%-3.5% is developed in the head and neck [2, 12]. There are limited detailed reports for the relapse of synovial sarcoma with long-term follow-up in the neck. Our study here displayed a case of relapse to contralateral neck in adolescent with 6-year of disease-free after initial surgery.

Like other sarcomas, synovial sarcoma generally affects children and young adult population [9, 12]. It is the most common non-rhabdomyosarcoma soft tissue sarcoma in childhood [9, 13]. This case here was definitely consistent with the characteristic of predilection age: first diagnosed at his age of nine, and found recurrent at seventeen. The age of onset seems to be associated with tumor prognosis. Age >35 years with synovial sarcoma was reported to be an adverse prognostic factor by univariate analysis in one European study [10]. They also found most recurrence occurred within 2 years after the initial treatment and patients with recurrence occurring

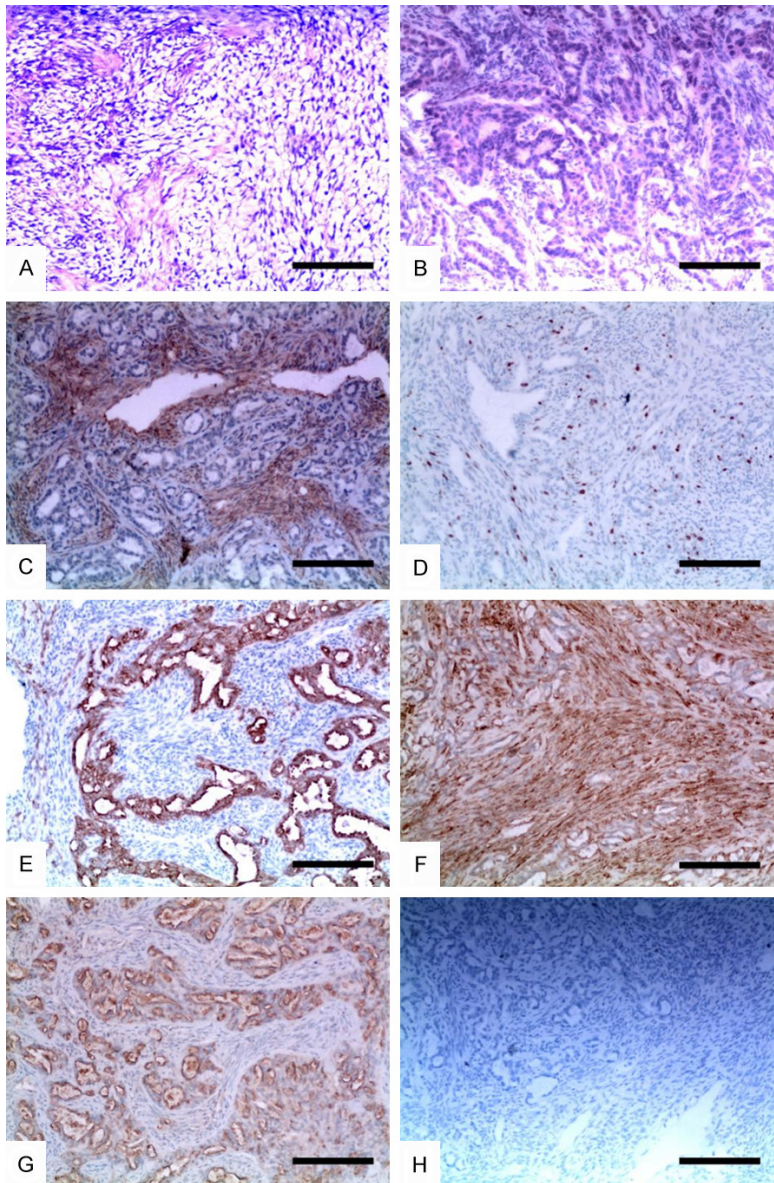


Figure 4. (A and B) Were H&E staining, showed spindle-cells arranged in variably sized fascicles or sheets (A), and epithelial round-cells arranged in glandular structures (B). (C-H) Was immunostaining. CD99 was positive in spindle-cells (C), Ki-67 was scattered expressed (5%-10% positive) in SS (D). CK-pan (E) and EMA (G) were strictly expressed in epithelial round-cells, and Vimentin was strong positive in spindle-cells (F). S-100 was entirely negative (H). Bar, 100 μm.

within 2 years had a significant worse outcome than patients in whom it relapsed later. Molecular analysis of chromosomal alterations revealed additional genomic copy number changes were more common in adult (>18 years) than children, which were strongly associated with metastatic spread and unfavorable prognosis [14]. Another study also revealed

that 5-year overall survival in children/adolescents was better than adults [12].

Despite termed synovial sarcoma; they do not originate from synovium, nor differentiates into any synovial tissues. Its name is likely a misnomer resulted from early literature description of tumor cells histologically presenting synovial differentiation and its propensity to originate near the joint regions [14]. There are two histological forms in synovial sarcoma: monophasic and biphasic; the former lesions are fusocellular neoplasms with little atypia in a fascicular and staghorn array, whereas the later harbors epithelial-like elements including spindle cells disposed in glandular structures [1, 2]. Although there is no definitive association between the histological subtype and prognosis, some studies reported a subtle relationship between these two histological forms [1, 2]. They found monophasic tumors tended to present with the size of <5 cm, whereas the biphasic tended to reach the size of >5 cm. The tumor size is an independent factor for prognosis and the important parameter for the option of treatment, the size of >5 cm is often considered as high-risk for relapse and thus adjuvant treatment are usually recommended [3, 9, 10].

In addition, tumors with biphasic subtype are more often SYT-SSX1-positive, the latter showed a higher proliferation rate than any other various forms, and were associated with a poorer clinical outcome [2]. For this case, the histologic subtype of first lesion was biphasic synovial sarcoma, and relapsed after six-years of disease-free.

Rare tumors histologically resemble the biphasic SS, but for monophasic SS, there are many tumors which are difficult to differentiate, such as fibrosarcoma, malignant peripheral nerve sheath tumor and hemangiopericytoma [3, 15]. A panel of immunohistochemical staining is thus necessary to avoid pitfalls in the diagnosis. Among the immunoprofile of SS, positive expression of CD99, BCL-2, and Vimentin are usually seen in the spindle cells; Cytokeratin expression, EMA are seen in epithelial component of most of SS [6, 15]. Transducing-like enhancer proteins are also sensitive to SS. However, none of immune makers are specific; therefore, they need to combine with other appropriate negative markers to support the diagnosis of SS. The gold standard of SS is the detection of SYT-SSX gene fusion [16], which is necessary for the cases that are difficult to differentiate. In our case, we performed immunohistochemical analysis to establish the diagnosis of SS.

Due to the rarity of the disease and difficulty of conducting randomized clinical trial, the optimal treatment of synovial sarcoma is not yet well established, but surgical resection is the prime treatment, especially for the initial lesion [1, 9, 10]. In this case, the relapsed lesion originated the limited prevertebral space and gradually grew and increased, then consequently oppressed the nearby structures. Although the airway was compressed by tumor, the patient didn't have the dyspnea. The difficulty for the surgery was the carotid vessels, fortunately, the tumor didn't invade the vessels and due to the surrounded capsule, we succeeded in remove the huge tumor completely. The surgical margin is very important to evaluate whether need any other adjuvant therapy. According to the microscopic examination, surgical margin is classified as R0 (surgical margin free of tumor either macroscopically or microscopically), R1 (microscopically tumor presence), R2 (macroscopically tumor presence) [2, 10]. For the margin status of R1 and R2, adjuvant chemo or radiotherapy is usually recommended [1, 2, 10, 17], but whether the adjuvant chemo or radiotherapy really brings a positive effect on overall survival remains to be established [10]. In this case, its complete surrounded capsule made it possible to entirely resect the tumor. The pathological examination of resected tumor revealed the capsule was com-

posed of fiber-like connective tissue and no tumor cell involved. Pathological report didn't find any lymph node involvement in resected tissue (II, III, and IV group). In spite of that, due to the relapsed tumor and tumor size (7*5*5 cm), adjuvant therapy was recommended.

In conclusion, we present a relapsed case with 6-year of disease-free after initial surgery. Synovial sarcoma is rare lesion in head and neck, a detailed report for relapsed disease is too rarer. This case could provide more insight to our knowledge of synovial sarcoma for its invasion and manifestation in head and neck.

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Disclosure of conflicts of interest

None.

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