

A rare pediatric malignant neck mass: Synovial sarcoma

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Synovial sarcomas are rare soft tissue sarcomas that typically seen in the peri-articular region and predominantly located in the extremities. It most commonly affects young adults of the second to fourth decade. Head and neck synovial sarcomas are uncommon and carry a poor prognosis. In the head and neck region, tumor is localized laterally in the parapharyngeal space often. The tumor can spread loco-regionally and systemically easily, so it makes management challenging. Herein, we report a case of a 12-year-old child with a synovial sarcoma located in the right parapharyngeal space of the neck. The lesion extended from parapharyngeal space to subglottic level of the neck and it fills the parapharyngeal space and compresses the major vascular, laryngeal and neural structures.

Keywords:

neck mass, parapharyngeal, synovial sarcoma

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Introduction

Sarcomas are rare mesenchymal neoplasms accounting for 1% of all malignant tumors in the human body [1]. Soft tissue sarcomas have many subtypes with different histological structures and biological behaviours. Nearly 10% of all sarcomas arise in the head and neck region. Local aggressive growing pattern and distant metastases are common [2,3]. Synovial sarcoma comprises nearly 10% of soft tissue sarcomas [3]. Infrequency of synovial sarcomas in the head and neck results in difficulties in the diagnosis and management. Complete surgical excision followed by chemoradiation is the suggested treatment modality [4]. In this paper, we present a 12-year-old boy with a rapidly growing synovial sarcoma in the right parapharyngeal space.

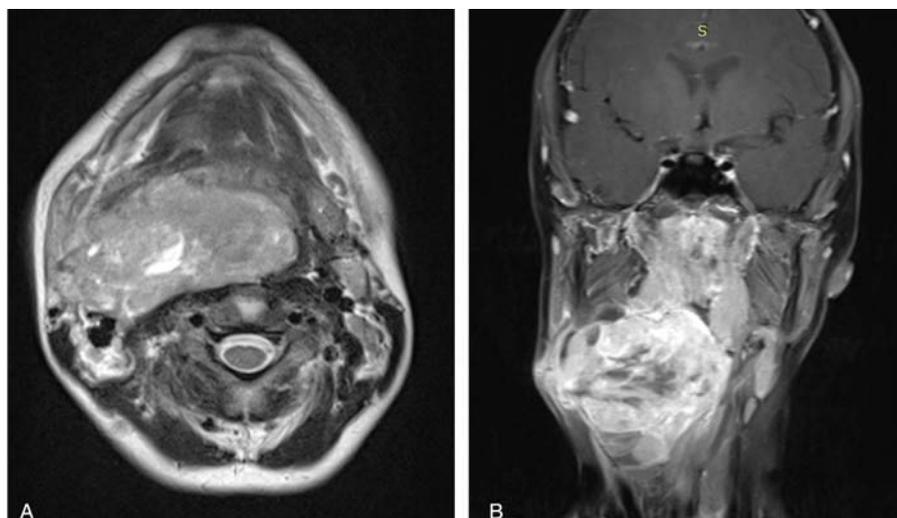
Case report

A twelve year old boy presented to the pediatric emergency unit with a large neck mass and acute respiratory distress. He has been suffering a rapidly growing painless right cervical neck mass, dysphagia and dyspnea for two months. He had a 10kg weight lost within two months. He had consulted to another clinic previously. An incisional biopsy had been performed lacking a definitive histopathological diagnosis. In physical examination there was a fixed, solid cervical mass on the right side. Examination of the oral cavity and oropharynx revealed a huge tumor with a smooth surface compromising the airway. An emergent tracheotomy procedure was performed because of acute respiratory distress. Subsequently

the patient was referred to ENT department for further evaluation and definitive treatment. Systemic examinations and routine blood tests were all in normal limits. An incisional biopsy procedure was repeated and synovial sarcoma diagnosis was confirmed after histopathological examination. Immunohistochemically, tumor cells were positive for CD99 and Bcl -2; epithelial cells of tissue were positive for Pan - keratin and Keratin -7. Vimentin were positive for epithelial and stromal component of tissue. Magnetic resonance imaging (MRI) of the head and neck revealed a 72*85*42mm mass arising from the right palatine tonsil extending inferiorly to the subglottic level. The right parapharyngeal space was completely occupied with the tumor. The mass was compressing major vascular and laryngeal structures without invasion (Fig. 1). A whole body positron emission tomography/computed tomography (PET-CT) examination was performed to rule out any distant metastasis. There was not any increased maximum standardized uptake values (SUVmax) other than the primary site, thus a complete excision of the tumor was planned. A trans-cervical incision was performed and the previous biopsy scar was included in the excisional material (Fig. 2). During the operation the tumor was observed to be adherent to deep fascia of the neck, pharynx and larynx. It was meticulously dissected from

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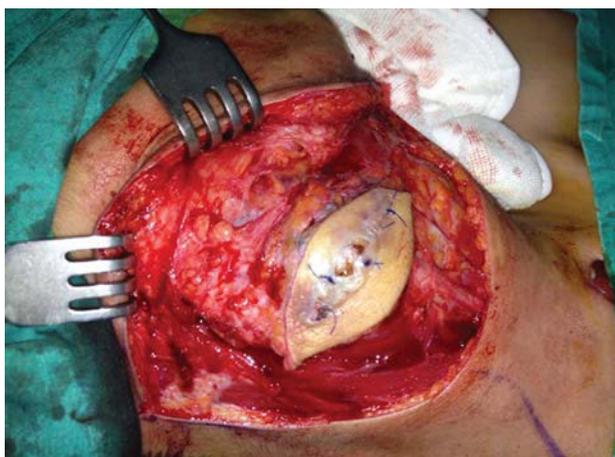
Figure 1



(a) Axial (a) and coronal (b) MRI sections of the tumor, respectively. The tumor is primarily arising from the oropharyngeal space It is occupying the whole parapharyngeal space and extending to the subglottic level involving the vital structures. (b) Axial (a) and coronal (b) MRI sections of the tumor, respectively. The tumor is primarily arising from the oropharyngeal space It is occupying the whole parapharyngeal space and extending to the subglottic level involving the vital structures.

internal carotid artery (ICA), internal jugular vein (IJV) and cranial nerves (CN) X, XI and XII, which all were not invaded by the tumor. Finally the tumor was totally excised and a functional neck dissection with the levels I, II and III was performed (Fig. 3). Histopathological examination confirmed the synovial sarcoma with a level I lymph node metastasis. Following the surgery the patient was subjected to a multiagent chemotherapy with vincristine, adriamycin, iphosphamide, cyclophosphamide and etoposide. Subsequently he was subjected to adjuvant radiotherapy. Presently he remains disease free 24 months after complete treatment.

Figure 2



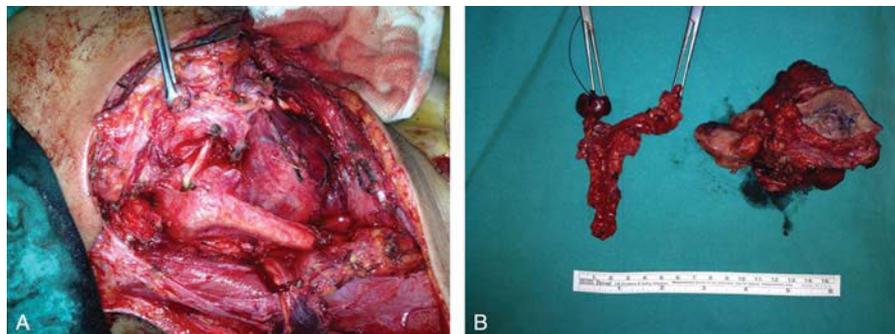
Intraoperative picture of the case. Past incisional scar included in the material.

Discussion

Synovial sarcoma is a mesenchymal malignancy, which primarily originates from the periarticular regions in the extremities [5]. It is entitled as 'synovial' because of its similarity to the tumors arising from synovial cells, however the neoplastic cells of this tumor originate from undifferentiated mesenchymal cells [6]. It is an uncommon sarcoma type, which less than 10% arise from head and neck region. Typically it originates from parapharyngeal space in the neck [5]. It may spread to regional lymph nodes and can easily metastasize distant organs resembling the other soft tissue sarcomas. Lung, bone, liver and central nervous system (CNS) are the most common regions bearing metastasis [7]. Majority of the head neck synovial sarcomas are located in the lateral neck, however it may also be located in the anterior neck [3].

The differential diagnosis of cervical neck masses in children is extensive and multifocal, however malignant neck neoplasms are uncommon. Lymphoma is the leading group. Rhabdomyosarcoma, thyroid carcinoma and nasopharyngeal carcinoma are the following malignant neck mass reasons seen in children [8]. Synovial sarcoma should be considered especially if the tumor is rapidly growing and located laterally in the neck. There are no specific imaging criteria to make a preoperative diagnosis for synovial sarcoma of the neck. Vaid S *et al.* diagnosed a huge synovial sarcoma of the retropharyngeal space with lipomatous structure. Solid/cystic components, fluid-

Figure 3



(a) Intraoperative picture of the case after the complete resection of the tumor. (b) Excisional material of the tumor and functional neck dissection.

fluid levels, heterogenous contrast enhancement indicated a malignant potential and absolute diagnosis could be done with pathologic examination [9].

The patients usually present with a painless rapidly growing mass with associated compressive and infiltrative symptoms of the surrounding structures [10]. Our patient also presented with a giant right neck mass with compressive symptoms like dyspnea and dysphagia. Tracheotomy procedure was performed to secure airway.

Immunohistochemical examination is crucial in the diagnosis of synovial sarcoma. In this case; CD34, CD99, CD21, CD35, Keratin - 7, Keratin - 19, Pan - keratin, S -100, Bcl - 2, Vimentin, EMA and CD45. Tumor cells are positive for CD99 and Bcl -2 in diffuse pattern. Epithelial component are positive for Pan - keratin and Keratin - 7; Vimentin are positive for both epithelial and stromal component. Histochemically, secretions in in the tissue are positive for AB - PAS and resistant to diastase.

Multidisciplinary approach is advocated to yield better prognosis. For synovial sarcoma in the head and neck, conservative surgery repeated radiotherapy with chemotherapy was suggested for adequate control of tumor. Because of the past incisional biopsy scar, scar margins must be included in the excisional material. Synovial sarcoma usually fills the parapharyngeal space and spread through the nearly whole neck. It must be resected with clear margin and major vascular and visceral structures must be preserved unless affected. Limited excision is associated with a high incidence of local recurrence, so aggressive dissection to ensure negative margins is the most important aspect of the treatment. However, in some cases radical excision cannot be performed without sacrificing vital structures, thus adjuvant therapy should be discussed with the patient.

After the aggressive resection of tumor we added the functional neck dissection on the same side also. Histopathological report confirmed our accurate decision; level1 lymph nodes are metastatic for sarcoma. Cervical metastasis is reported to occur in 10–20% of the patients in literature [7]. Surely, neck dissection is recommended for the patients with positive cervical lymph nodes but elective neck dissection would also be valuable.

Children and adolescents with synovial sarcoma originating at non-extremity locations have a worse prognosis than those with limb synovial sarcomas. Most aggressive treatment strategies can be applied for reaching a better survival rates [11]. Because of the poor histologic differentiation and high proliferation rate of the tumor, it is considered to show a poor prognosis [4]. Synovial sarcomas carry an unpredictable clinical behaviour therefore identifying prognostic factors and survival outcomes are critical. Tumor size and absence of metastatic disease are the most significant factors for overall survival [12,13]. Besides that; Stanelle *et al.* submitted that tumor size, depth, invasion and primary location affect the survival in pediatric synovial sarcoma with 5- and 10- year overall survival rates of 73 and 65% [14]. Initial metastasis, presence of lymphadenopathy, tumor size, histological type, surgical margins and anatomical location play important roles in prognosis. Combined modality therapy yields better results; a 5 - and 10- year survival rates for these patients [15]. Therefore postoperatively, chemoradiation therapy must be given for better prognosis especially in children.

Definitive treatment guidelines have not been published yet; adjuvant treatment decreases the local recurrence and improvement in the survival. Tumor localization and size can be challenging for radical surgery to achieve total excision. After achieving histological confirmation, chemoradiation can be

used initially that results the tumor shrinkage which allowed us to perform a complete resection without any surgical morbidity [16]. Attempts at a clear margin resection seems to decrease the local recurrence and improves the survival. For achieving the clear margin primary re-resection can be considered. Radiation therapy, either external beam or brachytherapy, is considered in all patients with microscopic residual or positive nodes. It appears that it should also be considered in patients with complete resection, because local control and relapse are reduced in the completely resected patients given radiation. The role of chemotherapy remains controversial. Andrassy RJ *et al.* showed no improvement in synovial sarcomas of children treated with chemotherapy [17].

We conclude that aggressive tumor resection with neck dissection and postoperative chemoradiotherapy should be the choice of treatment for the synovial sarcomas of the head and neck. Therefore a multidisciplinary team, comprising a head and neck surgeon, a radiation oncologist and a medical oncologist is needed.

Conflicts of interest

There are no conflicts of interest.

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