Intraventricular Synovial Sarcoma: A Case Report and Literature Review

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Abstract Synovial sarcoma (SS) is a high-grade malignant neoplasm. SS is a rare cancer type, which is primarily derived from the soft tissues of the lower extremities. The head and neck region is quite an extremely rare location, particularly the ventricle. The origin of SS remains a challenge, which along with its propensity to present clinical features mimicking other neoplasms within the common site, can result in significant diagnostic difficulty. Herein, we present a case of 40-year-old male SS located in the left ventricle with information including manifestation, imaging and histopathological features. On CT, approximately 30% of cases appear detectable calcification, which may be focal or dispersed throughout the tumor, often with a fine, stippled, or opaque appearance. MRI revealed a heterogeneous expansive and multi-lobular mass involving left ventricle, with intense and heterogeneous enhancement. Tentorium was thickened and enhanced. Small cystic changes were found in the peripheral part of the tumor with no enhancement. Radiologically, discriminating SS from other types of cancer is very difficult. Nonetheless, they should be considered in the differential diagnosis. Owing to the rarity of SS in the ventricle, misdiagnosis is common. In summary, we report one SS in the ventricle, the unusual location, to remind the radiologist to make it come to our mind for future work.

Keywords Synovial sarcoma; MRI; Diagnosis; Intraventricular neoplasm

Background Synovial sarcoma (SS) is a high-grade malignant neoplasm, representing 5%-10% of all soft tissue sarcomas. SS occurs over a wide age range, predominantly distributed in adolescents and young adults. There is a slight male preponderance, with a male: female ratio of approximately 1.2:1. Accounting for over 85% occurring in the extremities, especially encountered in large joint, SS was originally noted for its propensity for soft tissues derived from synovial cell. Actually, SS could be found throughout the body. The head and neck region is quite an extremely rare location, particularly the ventricle. The origin of SS remains a challenge, which along with its propensity to present clinical features mimicking other neoplasms within the common site, can result in significant diagnostic difficulty. Herein, we present a case of SS located in the left ventricle with information including manifestation, imaging and histopathological features. To our knowledge, this is the first reported case of SS involving ventricle associated with tentorium invasion. We discuss the related diagnostic aspects.

1 Case Report

1.1 History and examination
This 40-year-old male complaining of a 2-month history of headache was referred to our Neurosurgery Department. Neurological examination revealed no other deficits. Preoperative MR imaging demonstrated the presence of a large heterogeneously enhancing mass centered in the left ventricle. The lesion has evolved toward the tentorium; intracranial extension was present. There was marked mass effect on the left temporal lobe. A malignant meningioma was suspected as the pre-operative diagnosis.

1.2 Imaging features
Our case is a middle-aged male presenting a comparatively short detectable medical history. MRI identified a large mass in the left ventricle with significant enhancement. The tumor was expansive but with well-defined edge, and had the typical imaging features of meningioma with tentorium involvement. Owing to minimal cystic lesions in the peripheral part of the tumor, and the great mass effect affecting the left temporal lobe, a malignant meningioma was supposed to be taken into optimal consideration (Figure 1).
Figure 1 Preoperative MR imaging
Note: Axial noncontrast T1-weighted (A), T2-weighted (B), FLAIR (C) and DWI (D) revealed a heterogeneous expansive and multi-lobular mass involving left ventricle. Axial and sagittal Gd-enhanced T1-weighted (E, F) demonstrated intense and heterogeneous enhancement. Tentorium was thickened and enhanced. Small cystic changes were found in the peripheral part of the tumor with no enhancement

1.3 Operation
We therefore performed surgical excision of the tumor. Intracranial hypertension was demonstrated during the operation. Grossly, the mass was found hypervascular, solid and well defined, but infiltrated into the tentorium. The cut surface was firm.

1.4 Histopathological features
The definitive pathologic examination revealed SS with uniformly spindle cells. Immunohistochemical analysis showed VIM, p53, Ki67 (8%+) and bcl-2 positive staining. S-100, epithelial membrane antigen (EMA) and CD34 showed a negative result. The morphological and immunehistochemical features were characteristic of a monophasic synovial sarcoma.

2 Discussion
Synovial sarcoma (SS) is generally considered a high-grade malignant neoplasm, representing between 5% and 10% of all soft tissue sarcomas (Herzog et al., 2005; Sultan et al., 2009; Shi et al., 2013). SS of the head and neck region is quite rare, and accounts for approximately 3% to 10% of all synovial sarcomas. The most common site for SS in the head and neck region is the hypopharynx, because it is the seat of numerous synovial formations. To the best of our knowledge, this is the first reported case of SS of the intraventricle involving tentorium.

The origin of SS is still controversial. Virtually, it is named exclusively for its appearance, and as SS neither arise from nor differentiate toward synovium, the name is a historical error (Smith et al., 1995; Fisher et al., 1998; Thwayet al., 2014). Owning to its more common origination from the pluripotent mesenchymal cells nearby or even remotely from articular surfaces, tendons, tendon sheaths, juxta-articular membranes, and facial aponeuroses, rather than from mature synovial tissue. The ventricular SS, which has not been reported yet, is quite a unique site for the occurrence.
SS appears to be frequently derived from the lower extremities, predominately the knee, usually with close relation to tendon or tendon sheath and cystic structures, but rarely in joint cavity. Besides the common extremity site, the head and neck region is the next frequent site. Sarcomas of the head and neck represent approximately 1% of all head and neck malignancies. SS accounts for less than 10% of cases. Paravertebral site is the most common region for SS in the head and neck, with the presentation from skull base to the hypopharynx (including pharyngeal, parapharyngeal, and the prevertebral planes; skull base and temporomandibular joint fossa) (Tilakaratne et al., 2006; Horbinsk et al., 2008; Al-Daraji et al., 2009; Scheithauer et al., 2011; Keith et al., 2013; Lin et al., 2013; Xiao et al., 2014). Occurrences besides extremities or head and neck have also been documented, such as thorax (including heart and lungs), even rarely within abdomen and pelvis (Witkin et al., 1989; Gaertner et al., 1996; Nicholson et al., 1997; Nicholson et al., 1998; Fisher et al., 2004; Suster et al., 2005; Sakellaridis et al., 2006; Company et al., 2007; Schreiber et al., 2007; Makhlouf et al., 2008; Katakura et al., 2009; Cummings et al., 2010; Alsharief et al., 2012; Dhawan et al., 2012; Nomura et al., 2014; Crowson et al., 2015; Eravci et al., 2016).

The diagnosis of SS is generally made by its relatively distinctive, yet markedly variable histopathologic appearance, in conjunction with the histochemical, immunohistochemistry, electron microscopy, and cytogenetic analysis findings. Histologically, SS was originally considered a malignant tumor arising from synovial membrane. Studies have, however, shown that it may originate from pluripotent mesenchymal cells with no definitive association to articular surfaces or membranes. Therefore, it is notable that SS could occur in every anatomic site.

According to some investigators, SS apparently shows epithelial differentiation, but synovial membrane does not actually harbor epithelial cells. Thus, SS is named for its appearance, rather than the origin. Different from other sarcomas, SS is composed of carcinoma-like epithelial cells and fibrous sarcoma-like spindle cells. Although the 2 characteristic cell types of SS are morphologically distinct, they are histogenetically similar. Basically, SS comprises 2 major subtypes and rarer subtypes. The former include monophasic and biphasic spindle cell types; and the latter includes monophasic epithelial, poorly differentiated, calcifying/ossifying, and myxoid types, depending on their varying composition and degree of differentiation. The monophasic spindle cell type composed of plenty spindle cells is the commonest subtype of SS, and has a tendency towards indistinguishable from other tumors common to the region, such as adenocarcinoma.

Immunohistochemically, SS presents positive to cytokeratin (CK), epithelial membrane antigen (EMA), S-100, bcl-2, CD99 and calponin. Varying from other spindle cell tumors, SS is negative to CD34.

In terms of radiological findings, the imaging features of intraventricular SS remain needed to be accumulated. Generally, SS tends to occur in extremities, often as expansive lobulated masses, with well-defined margin, and most frequently close to joints. Although there is no consensus on imaging, calcification has been reported common and more easily discerned radiologically. On CT, approximately 30% of cases appear detectable calcification, which may be focal or dispersed throughout the tumor, often with a fine, stippled, or opaque appearance.

Magnetic resonance imaging is thought to be the imaging modality of choice for showing the internal features of the lesion and superior soft tissue contrast. In addition, MR has a priority in the assessment for invasive characteristics such as infiltration into the surrounding soft tissues (including muscle, adjacent bone) and adherent to tendon or neurovascular structures. Typically, SS range from 3 to 10 cm in diameter. The lesions tend to be small when they occur in hands or head and neck. The survival rate is regarded to be low when the lesion is more than 4 cm. Lesions less than 1 cm in diameter which are named as “Minute” SSs are clinically considered benign processes, such as ganglion cysts or glomus tumors. Occasionally, there is cyst formation, with smooth walled cysts, often multiple, containing mucoid fluid or blood. Hemorrhage and necrosis can be prominent in poorly differentiated SS, although are less prominent than in high-grade pleomorphic sarcomas.

SS is a rare cancer type, which is primarily derived from the soft tissues of the lower extremities. Ventricle is an extremely rare localization. Cases with the presentation affecting dura and cerebellum have been characterized. To
the best of our knowledge, involvement of ventricle has not been reported to date. The present case involving the lateral ventricle with the tentorium thickened may help us to better recognize the tumor. The types of tumors in the ventricle make up a long list, including choroid plexus papilloma, meningioma, central neurocytoma, astrocytoma and ependymoma. Radiologically, discriminating SS from other types of cancer is very difficult. Nonetheless, they should be considered in the differential diagnosis.

Intracranial synovial sarcoma is very rare. Tumors are from periventricular dura tissues rather than synovial cells. SS can appear as benign lesions with expansive smooth boundaries. SS is supposed to be taken into thought when septations, hemorrhages, calcification and cystic components shown in the imaging studies. The clinical presentation of head and neck SS correlates with the tumor region. Our patient presented with 2-month history of headache owing to extension of the tumor mass in the left ventricle and slight intracranial hypertension. Clinically, many patients are not indicated for surgery, because these ventricular tumors grow slowly, and are often asymptomatic until they reach a relatively large size sufficient to create pressure on the adjacent structures, when they cause symptoms or are identified incidentally. The most encountered symptom is headache, as seen in our case. The tumor showed the predominance to exert pushing margins and was circumscribed by a fibrous pseudocapsule, as they had slow growth history. With respect to our patient, the case presented with a well-encapsulated mass in the left ventricle that infiltrates tentorium. The growth was similar in appearance to a neoplasm originating from the dura within the lateral ventricle. Because of its smooth surface, along with marked affiliation with dura, the growth was diagnosed pre-operationally as meningioma. In addition, the pronounced mass effect on the surrounding temporal lobe along with the multiple cysts within the mass, the malignancy was considered. Owing to the rarity of SS in the ventricle, misdiagnosis is common. In summary, we report one SS in the ventricle, the unusual location, to remind the radiologist to make it come to our mind for future work.

Authors’ contributions
Liang Zong-hui conceived and designed the work that led to the submission. Dou Ya-fang and Liang Zong-hui evaluated images and analyzed data. Dou Ya-fang completed the writing of the article. Liang Zong-hui revised the paper. All authors participated in paper writing and approved the final manuscript.

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