MONOPHASIC SYNOVIAL SARCOMA AT UNUSUAL LOCATIONS

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ABSTRACT
A synovial sarcoma (SS) is an aggressive soft tissue tumor, which mainly occurs in the para-articular region of extremities with a predilection of lower limb (Xu et al., 2010). Synovial sarcoma (SS) though rare, is a morphologically, clinically and genetically distinct entity. With the advent of immunohistochemistry and molecular techniques, cases of synovial sarcomas have been reported in unusual location including the head and neck (Tilakaratna, 2006), mediastinum (Witkin et al., 1989), lung (Zeren et al., 1995), abdominal wall (Fetsch et al., 1993), intraabdominal (Fisher et al., 2004), kidney (Jun et al., 2004) and retroperitoneum (Adam et al., 1984). The monophasic SS consisting solely of sarcomatous components is often diagnostically challenging. Careful attention to histopathological features, high degree of suspicion and a battery of IHC markers allow us to exclude the wide range of tumours in the differential and arrive at the correct diagnosis.

Key Words: Monophasic Synovial Sarcoma, Inguinoscrotal Swelling, Cytokeratin, Vimentin

INTRODUCTION
A synovial sarcoma (SS) is an aggressive soft tissue tumor, which mainly occurs in the para-articular region of extremities with a predilection of lower limb. It usually develops in adolescents and young adults between the age of 15 and 40 years (Weiss and Goldblum, 2001). The detection of a reciprocal translocation between chromosomes X and 18t (X: 18) has led to the identification of an SS18 gene (also known as SYT) rearrangement being involved in the formation of a SYT-SSX fusion protein in synovial sarcomas (Clark et al., 1994). Synovial sarcoma (SS) though rare, is a morphologically, clinically and genetically distinct entity.

With the advent of immunohistochemistry and molecular techniques, cases of synovial sarcomas have been reported in unusual location including the head and neck (Tilakaratna, 2006), mediastinum (Witkin et al., 1989), lung (Zeren et al., 1995), abdominal wall (Fetsch and Meis, 1993), intraabdominal (Fisher et al., 2004), kidney (Jun et al., 2004) and retroperitoneum (Adam et al., 1984). SS of the head and neck region was first described by Jernstrom (1954), reporting on a case of malignant synovioma of the pharynx. Synovial sarcoma as a para-testicular mass involving inguinal or scrotal region is an uncommon condition (Naito et al., 2000). The monophasic SS consisting solely of sarcomatous components is often diagnostically challenging. Here we present two unusual presentations of mono-phasic synovial sarcoma.

CASES
CASE 1: An 11-year old boy presented with a slow growing swelling in the scalp. It was painless and been growing since 2 years. No significant past history. General physical examination was normal. His hematological profile was within normal limits. X-ray/CT scan showed a small soft tissue swelling in the scalp posteriorly on the left. No underlying abnormalities were seen. Chest X-ray was also within normal limits. A wide excision was done and intra-operatively the lump was found to be highly vascular. On gross examination an irregular grayish white mass (3 x 3 x 2) cm in size, firm in consistency. On cut section mass was grayish, well circumscribed and solid with areas of haemorrhage. Microscopically, the tumour was very cellular, with extensive areas showing haemangiopericytoma like patterns with slit like vascular spaces surrounded by spindle cells with uniform and relatively small, tapering nuclei and inconspicuous nucleoli and poorly defined cytoplasm in a variably collagenous stroma. Frequent mitoses were also common. Immunohistochemistry showed the tumour to be positive for cytokeratin and Bcl2.
Case Report

CASE 2: A 70 years old male presented with a slow growing, painless left sided inguinoscrotal swelling for four years. On examination the swelling was ill-defined, bosselated with variegated feeling about (8x10) cm in size, non-tender and immobile. Bilateral testes were normal in size, shape and sensation. High Resolution ultrasonography of the inguinoscrotal region with Doppler revealed a very large ill defined heterogenous SOL with moderate vascularity. CT scan of abdomen & pelvis was suggestive of extra testicular soft tissue sarcoma with infiltration to neighbouring structures.

A grayish white mass of (8x10x10) cm in size with firm consistency. On cut section the mass was well circumscribed, consisting of solid and cystic parts with haemorrhage and necrosis and was covered by an ill-defined capsule. Microscopically, the tumour was cellular with oval to spindle shaped cells with uniform and relatively small tapering nuclei and inconspicuous nucleoli in a scanty stroma. Few mitotic figures were noted. IHC was positive for vimentin, cytokeratin & bcl-2.

DISCUSSION

Synovial sarcomas are uncommon soft tissue tumors accounting for 5-10% of the soft tissue sarcomas. They usually develop in children and young adults and approximately 95% of SSs occur in the extremities. They can metastasize distantly, especially to the lung and lymph. Two major histologic subtypes exist: monophasic and biphasic synovial sarcomas. The classical SS has a biphasic appearance with a mixture of epithelial and spindle cells in varying proportions. The monophasic SS consisting solely of sarcomatous components is often diagnostically challenging. Synovial sarcomas typically occur in patients between the age of 15 and 40 years, rarely in the elderly (Weiss and Goldblum, 2001). Patients commonly present with a palpable mass or swelling, associated with pain in almost half of the patients. The most common sites include the lower extremities (60%), with a special predilection for the knee area; the upper extremities (23%); the head and neck region (9%), particularly in the retropharyngeal and parapharyngeal areas; and the trunk (8.1%), most commonly involving the abdominal wall or retroperitoneum. The monophasic variant may easily simulate fibrosarcoma, MPNST, HPC, and mesenchymal chondrosarcoma. In other words, because of the high degree of cellularity, a herringbone pattern or an HPC pattern may predominate. However, clues to the real nature of the tumor include the presence of small, oval overlapping nuclei, the identification of rare clusters of more plump and eosinophilic cells, the lack of the nerve like wavy nucleus, the lack of any cartilaginous differentiation, and the location (Mills et al., 2004). The spindle cells were positive for cytokeratin (CK), epithelial membrane antigen (EMA), calretinin, vimentin (VIM), bcl-2 and CD-99, but negative for smooth muscle actin(SMA), muscle specific actin (MSA), CD117 and S100 (Mills et al., 2004). Surgery is the mainstay treatment. Chemotherapy and radiation therapy should be considered as additional treatment options or may be utilized in cases of relapse. Prognosis is poor with local recurrence and metastases in almost 50% of the patient (Weiss and Goldblum, 2001).

Conclusion

Monophasic synovial sarcoma in an unusual location can present as a diagnostic dilemma. Careful attention to histopathological features, high degree of suspicion and a battery of IHC markers allow us to exclude the wide range of tumours in the differential and arrive at the correct diagnosis.

REFERENCES


Case Report


