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SYNOVIAL SARCOMA OF THE COLON WITH INTRACYTOPLASMIC MUCIN- A CASE REPORT

Pathology

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ABSTRACT

Synovial sarcomas are rare tumors, accounting for 5-10% of adult soft tissue sarcomas affecting young adults in para-articular regions. Primary intra-abdominal synovial sarcoma is rare, with about 47 cases reported in the literature. Only few cases of synovial sarcomas with cystic and myxoid change have been described. Intracytoplasmic mucin in synovial sarcomas with presence of glandular component is extremely rare. Lung is the most common site for metastasis and only few cases are reported with liver and omental metastasis. Intra-abdominal synovial sarcomas seem to have a better prognosis compared to the synovial sarcomas in para-articular regions.

We present an unusual case of a 35 year old female who presented with right lumbar pain. CT scan showed an exophytic growth on the serosal surface of colon. Histology showed predominantly spindle cells with occasional small glands having focal intracytoplasmic mucin which was diagnosed as synovial sarcoma with the aid of immunohistochemistry and molecular studies.

KEYWORDS

Synovial sarcoma, intra-abdominal, morphology, mucin, immunohistochemistry.

INTRODUCTION:

Synovial sarcoma (SS) is a rare tumor accounting for about 5-10 % cases of the soft tissue sarcomas. According to earlier studies, only 5–10% of synovial sarcomas occur in unusual sites like head and neck, mediastinum, abdominal wall, esophagus, and retroperitoneum (Sarkar, 2018). Isolated rare cases involving visceral organs are also reported which include pleura, prostate, intra-abdominal organs, blood vessels, skin, and nerves (Buiga-potcoavã, Crisan, & Olinici, 2005). Primary intra- abdominal synovial sarcoma is extremely rare. Till date, only 47 cases have been reported in literature(Kritsaneepaiboon, Sangkhathat, & Mitarnun, 2015). Intracytoplasmic mucin in synovial sarcomas is very unusual, with only few cases reported in literature. Hence, we report a rare case of synovial sarcoma in the colon because of its rarity in this site and the presence of intracytoplasmic mucin.

CASE SUMMARY:

A 35 year old female presented with complaints of right lumbar pain and a palpable mass. CT scan showed an exophytic growth on the transverse colon for which a right hemicolectomy was done. Intra-operatively, omental and hepatic nodules were also noted and excised.

Right extended hemicolectomy specimen was received which showed a large serosal mass on the colon measuring 8.9 x 6.9 x 2 cm with multiple cysts on the external surface. Serial sections across the nodule showed a partly solid and cystic tumor with solid areas showing grey white to grey brown, glistening and focal yellow cut surface. The cysts were filled with seromucinous fluid. The ileal and colonic mucosa appeared normal. The rest of the serosa and the omentum also showed multiple tumor deposits.

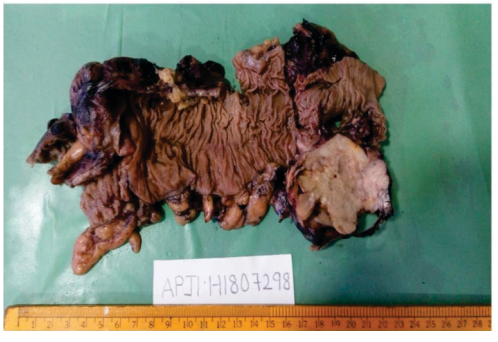

Figure 1: Right extended hemicolectomy specimen showing a serosal nodule. Cut surface showed a solid and cystic tumor with glistening appearance.

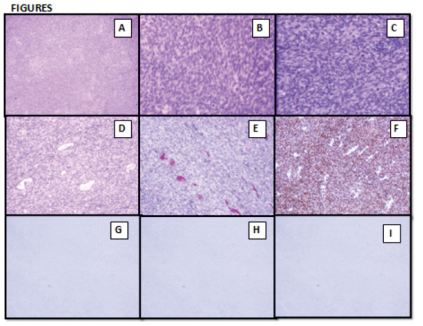

Figure 2 A - Cellular tumor with spindle cells in fascicles suggestive of a sarcoma. (H and E stain, 10X); B - Spindle to oval cells with plump hyperchromatic nuclei and mitosis suggestive of a sarcoma. (H and E stain, 40X); C- Intervening aggregates of polygonal cells with rhabdoid appearance. (H and E stain, 40X); D- Occasional gland formation. (H and E stain, 10X); E- Cytoplasmic and luminal mucin. (PAS stain, 10X); F - Tumor cells show nuclear positivity for TLE-1 confirming the diagnosis of synovial sarcoma (Immunohist ochemistry for TLE-1, 10X); G, H, I - Tumor cells are negative for DOG-1, CALRETININ, CK5/6 respectively (10X).

Histopathologic examination revealed a cellular tumor showing predominantly spindle cells in fascicles. Intervening aggregates of polygonal cells having a focal rhabdoid appearance with eccentric nuclei were seen. At places, hyaline like globules distending the cytoplasm were also noted. Brisk mitotic activity was seen averaging 4-5/ high power field, especially in the spindled areas. Large areas of cystic degeneration and necrosis were also seen. Cytoplasmic and luminal mucin was demonstrated using the periodic acid schiff stain with diastase and found to have resistant mucin. This, though unusual, has been reported in the literature. The ileal and colonic mucosa as well as the appendix appeared normal. Multiple serosal and omental deposits were noted. In view of the clinical and histomorphological features, the differential diagnoses considered were synovial sarcoma, gastrointestinal stromal tumor and mesothelioma.

Immunohistochemically, the tumor cells showed diffuse strong nuclear positivity for TLE-1 (Figure F). CD117, DOG-1, Calretinin, CK7, cyclin D1, CD10 and CK20 were negative (Figure G,H,I). This immunohistochemical profile confirmed a diagnosis of synovial sarcoma, having excluded the other differential diagnosis.

Hence, molecular studies for t(X; 18) or the SYT/SSX fusion gene was advised.

DISCUSSION:

Synovial sarcoma is a deep seated tumor accounting for about 5-10 % cases of the soft tissue sarcomas affecting young children and adolescents. SS usually arises in the extremities, most commonly in the lower limbs affecting the periarticular areas.

SS arises from primitive mesenchymal cells which could explain its extra-articular and unusual location (Kritsaneepaiboon et al., 2015). According to earlier studies, only 5–10% of synovial sarcomas occur in the head and neck, mediastinum, abdominal wall, esophagus, and retroperitoneum (Sarkar, 2018). Isolated rare cases involving visceral organs have also been reported including sites like the pleura, the prostate, intra-abdominal organs, blood vessels, skin, and nerves (Buiga-potcoavã et al., 2005).

In 2005, Potcoava B et al. reported a case of a 41-year-old female with an intra-abdominal synovial sarcoma in an unusual location, the ascending mesocolon (Buiga-potcoavã et al., 2005). A similar case was reported by Sarkar S et al. in 2018 in which a 46-year male presented with an intra-abdominal mass filling the entire pelvis and extending upwards till the subhepatic region (Sarkar, 2018). Both the cases were confirmed as synovial sarcoma by immunohistochemistry and molecular studies.

Primary intra- abdominal synovial sarcoma is rare with about 47 cases reported in literature (Kritsaneepaiboon et al., 2015). Our case presented with an exophytic growth on the surface of the transverse colon.

Histologically, SS has 3 patterns- monophasic, biphasic and poorly differentiated. The biphasic pattern can have glands showing intracytoplasmic mucin which can sometimes be a predominant component. These glands show intraluminal mucin but, presence of intracytoplasmic mucin is rare (Weinreb, Perez-Ordonez, Guha, & Kiehl, 2008) as was seen in our case. (Figure E)

Weinreb I et al. in 2018 described an unusual case of mucinous gland predominant SS of a large peripheral nerve with less than 1% spindle cell component. Metastatic mucinous carcinoma was considered as a differential diagnosis in view of the abundant luminal and intracellular mucin. IHC and ancillary testing for the SYT gene (18q11.2) rearrangement helped in confirming it as synovial sarcoma (Weinreb et al., 2008).

Our case showed predominant spindle cells with few mucinous glands and focal intracytoplasmic mucin which was confirmed by PAS with diastase. 85-90% cases are positive for TLE-1, as was seen in the present case.

With the aid of immunohistochemistry, PCR and FISH based detection of SYT-SSX fusion because of t(X;18), synovial sarcoma can be detected in unusual sites like head and neck areas, abdominal wall, retroperitoneum (Tsuji & Hisaoka, 1998). Our case showed an intra-abdominal tumor, which is a rare site for the lesion.

Liver and omentum are rare sites of tumor metastases, which were seen in our case (de Necochea-Campion et al., 2017) (Sinniah, Roche, & Cameron, 2012). Synovial sarcomas are aggressive tumors with a high chance of local recurrence (Buiga-potcoavã et al., 2005).

CONCLUSION:

SS is a rare entity and can occur in unusual sites like intra-abdominal region. Tumors in these unusual sites seem to have a better prognosis. Hence, correct diagnosis with the aid of histomorphology, IHC and molecular studies is essential for appropriate treatment and tumor prognostication.

REFERENCES:

1. Buiga-potcoavã, R., Crisan, D., & Olinici, C. D. (2005). Primary Intraabdominal Synovial Sarcoma : a Case Report, 14(1), 67–69.
2. de Necochea-Campion, R., Zuckerman, L. M., Mirshahidi, H. R., Khosrowpour, S., Chen, C.-S., & Mirshahidi, S. (2017). Metastatic biomarkers in synovial sarcoma. Biomarker Research, 5(1), 4.
3. Kritsaneepaiboon, S., Sangkhathat, S., & Mitarnun, W. (2015). Pediatric Radiology: Primary synovial sarcoma of the abdominal wall: a case report and literature review Kritsaneepaiboon et al. Primary synovial sarcoma of the abdominal wall: a case report and literature review. Radiology Case, 9(7), 47–52.
4. Sarkar, S. (2018). CASE REPORT A Case of Primary Intra-abdominal Synovial Sarcoma.
5. Sinniah, R. P., Roche, E., & Cameron, D. (2012). LETTER TO THE EDITOR, 3(4), e11-2.
6. Tsuji, S., & Hisaoka, M. (1998). Detection of SYT-SSX Fusion Transcripts in Synovial Sarcoma by Reverse Transcription-Polymerase Chain Reaction Using Archival Paraffin-Embedded Tissues. The American Journal of Pathology, 153(6), 1807–1812.
7. Weinreb, I., Perez-Ordonez, B., Guha, A., & Kiehl, T.-R. (2008). Mucinous, gland predominant synovial sarcoma of a large peripheral nerve: a rare case closely mimicking metastatic mucinous carcinoma. Journal of Clinical Pathology, 61(5), 672–676.