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Primary Intra-Abdominal Synovial Sarcoma in A Young Adolescent with Delayed Presentation : A Rare Entity

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Introduction

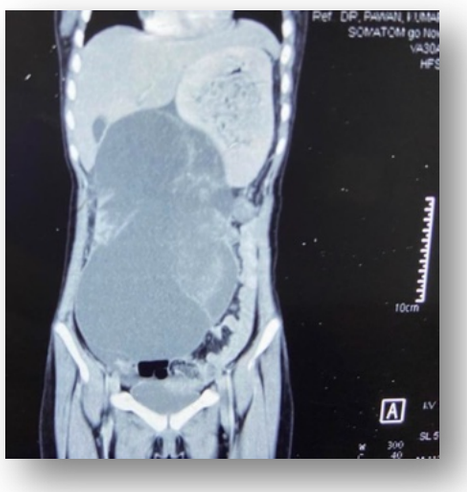
- Synovial sarcoma is a rare subtype (4th most common) of soft tissue sarcoma in children and adults.
- Primary intra-abdominal synovial sarcoma is uncommon and mainly retroperitoneal.
- In a child with an abdominal lump primary germ cell tumor, lymphoma, and ovarian cyst is excluded before reaching a diagnosis of soft tissue sarcoma arising from the abdomen.
- The authors present a rare case of intraabdominal Synovial sarcoma with delayed presentation.

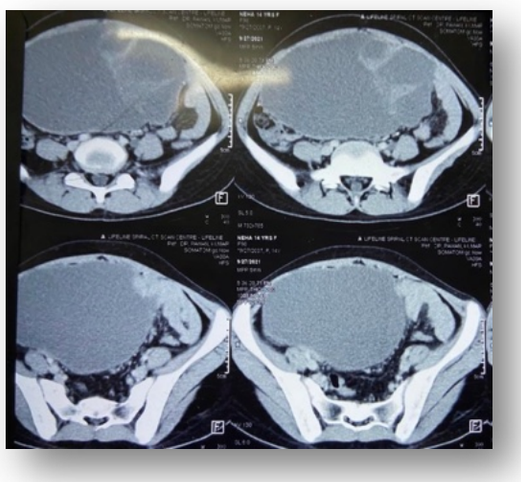
Case Summary

A 15-year girl presented to ED with complaints of respiratory discomfort, abdominal distension & pain abdomen with recurrent vomiting for 10 days, associated with a 15x20 cm lump, firm with restricted mobility, occupying whole of the abdomen from epigastrium to pelvis. There was no history of jaundice, hematemesis, melena, fever, ATT intake or previous surgery. Imaging was s/o solid -cystic intraabdominal mass lesion? GIST/ neuroendocrine tumor/mesenchymal origin mass. US guided FNAC (Outside): Neuroendocrine tumor- Carcinoid. Repeat USG guided biopsy at KGMU: ? STS/? GIST

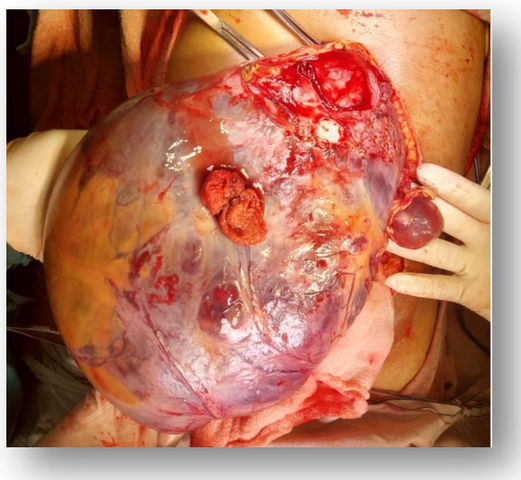
Treatment

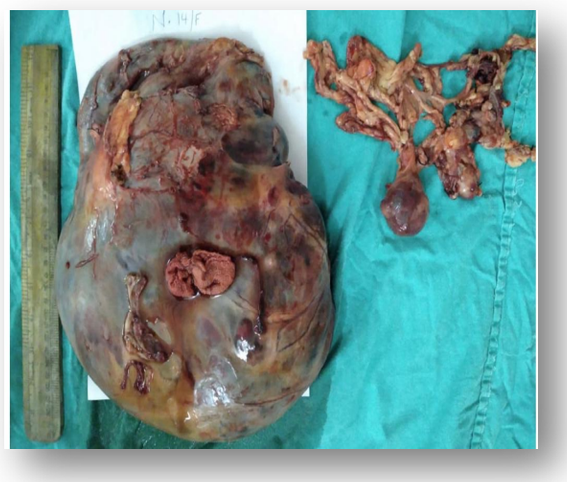
- Exploratory laparotomy → large cystic lump extending from epigastrium to pelvis causing GOO with multiple omental nodules and peritoneal deposits.
- Excision of lump and radical omentectomy with peritonectomy under GA with left ICD
- HPE: Synovial Sarcoma FNCLCC grade 3
- IHC: Positive for Vimentin, Bcl-2, SMA, CD99, CD56 and negative for CD34, CD117, Chromagranin, Synaptophysin.
- Post op recovery uneventful , planned for adjuvant chemotherapy



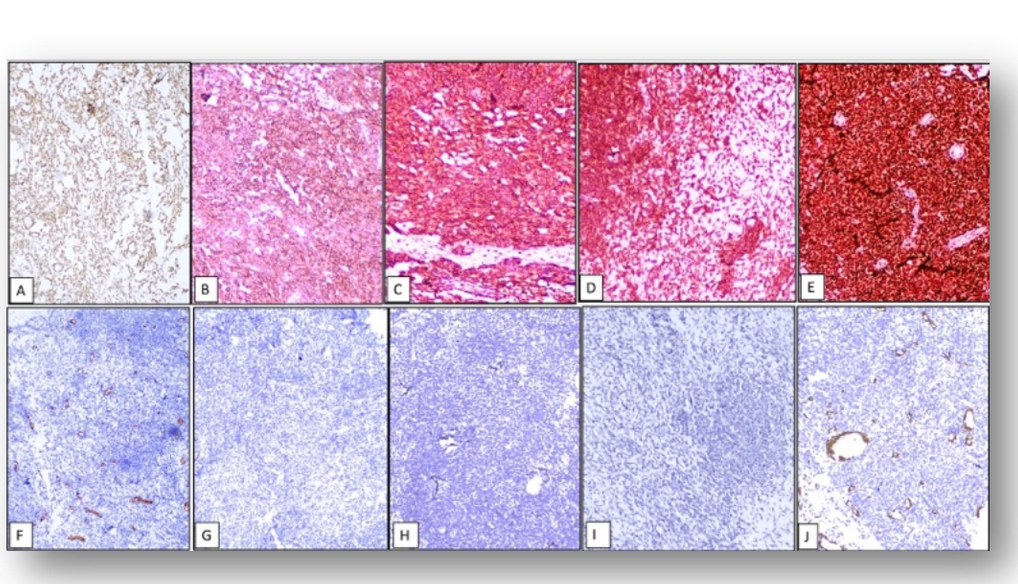


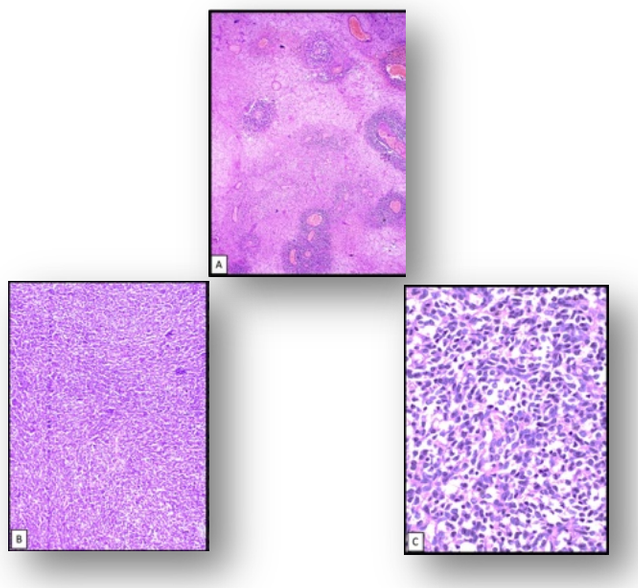
Large heterogenous mass extending to pelvis





Excised 20x15 mass (8.5 Kg) with Radical omentectomy





Tumour cells are diffusely positive for Vimentin (A), CD56 (B), CD99 (C), BL2 (D), TLE1 (E), while negative for CD34 (F), Synaptophysin (G), CD117 (H), Chromagranin (I), and SMA (J) (100X)

Conclusion

- Primary intraperitoneal synovial sarcoma is rare.
- Surgery remains the mainstay of treatment of soft tissue sarcoma, adjunctive radiation and chemotherapy, alone or in combination, has substantial role in recurrence and metastatic control.
- Treatment should be individualised according to patient and disease related factors .
- Early referral to a tertiary care centre impacts outcome.

References

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