

Monophasic synovial sarcoma as a cause of obstructive ileus: A case report

ARGYRIOS IOANNIDIS¹, CHRISTOS KOUTSERIMPAS², ATHANASIOS PAPATSORIS³, ALEXANDRA ARGYROU⁴, HARALAMPOS DELIVELIOTIS³, GEORGIOS VELIMEZIS¹ and ATHANASIOS-MELETIOS DIMOPOULOS⁵

¹Second Department of Surgery, Sismanoglio General Hospital of Athens, 15126 Athens;

²Department of Orthopedics and Traumatology, '251' Hellenic Air Force General Hospital of Athens, 11525 Athens;

³Second University Department of Urology, National and Kapodistrian University of Athens School of Medicine, Sismanoglio General Hospital of Athens, 15126 Athens; ⁴Department of Basic Medical Sciences, Laboratory of Biology, School of Medicine, National and Kapodistrian University of Athens, 11527 Athens; ⁵Department of Clinical Therapeutics, National and Kapodistrian University of Athens School of Medicine, Alexandra Hospital, 11528 Athens, Greece

Received September 30, 2018; Accepted October 30, 2018

DOI: 10.3892/mco.2018.1768

Abstract. Synovial sarcoma is a high-grade soft tissue sarcoma, divided histologically into 3 types: Monophasic, biphasic and poorly differentiated. An extremely rare case of obstructive ileus due to monophasic synovial sarcoma in a 54-year-old male patient is reported in the present study. The patient presented to the emergency department with signs and symptoms of obstructive ileus. Computer Tomography (CT) scan showed a pelvic mass (20x17x12 cm) causing obstruction at the sigmoid colon. An exploratory laparotomy through a midline incision was performed. The described mass was unresectable as it was infiltrating major vessels. Therefore, a diverting loop descending colostomy and biopsy of the solid mass were performed. Pathological examination revealed a monophasic synovial sarcoma. The patient was referred to the Department of Oncology and started treatment with doxorubicin and ifosfamide.

Introduction

Synovial sarcoma represents a high-grade soft tissue sarcoma, divided histologically into 3 types: Monophasic, biphasic and poorly differentiated. It has also been associated with a specific chromosomal translocation t(X:18) (p11: q11) (1,2). Synovial sarcoma appears most commonly in the lower extremities, while it is less common in the upper extremities, head, neck and trunk (3). Synovial sarcoma may occur in any region and at any age; however, the third decade represents the peak incidence (4). Older age, tumor size >5 cm, as well as trunk

location have been considered as risk factors for late stage disease. Patients younger than 25 years, with tumor size <5 cm and well-differentiated, may be considered low-risk, whereas patients older than 25 years, with tumor size >5 cm and poorly differentiated are considered high-risk (5). Patients suffering from trunk synovial sarcoma may present with abdominal pain, bloated sensation, as well as dyspepsia (3-5).

An extremely rare case of obstructive ileus due to monophasic synovial sarcoma in a 54-year-old male patient is reported in the present study.

Case presentation

A 54-year-old male presented to the emergency department of the Sismanoglio General Hospital of Athens complaining of abdominal pain and vomiting. His symptoms started 2 days prior to admission. His medical history was remarkable for psoriasis (under adalimumab/2 weeks) and a transurethral resection (TUR) of the prostate due to benign hypertrophy. The patient was stable (blood pressure was 130/80 mmHg, heart rate was 83 beats per min, Spo2 was 97%) and afebrile (37°C). Medical examination revealed distended abdomen and hypoactive bowel sounds in auscultation, while the plain X-ray of the abdomen showed gas-fluid levels. Initial laboratory investigation exhibited hemoglobin of 12.1 g/dl. A computer tomography (CT) scan was then performed with oral contrast. The CT scan showed signs of obstructive ileus at the sigmoid colon caused by a solid pelvic mass whose origin could not be determined (20x17x12 cm). In order to further investigate the mass and its origin, a magnetic resonance imaging (MRI) scan was also performed (Fig. 1).

On identification of the obstructive ileus, the patient was led to the operating theater and an exploratory laparotomy through a midline incision was performed. The described mass was considered unresectable due to contact with major vessels. Therefore, a loop descending colostomy and biopsy of the solid mass were performed. The patient had an uneventful recovery. He was discharged on the 8th postoperative day.

Correspondence to: Dr Argyrios Ioannidis, Second Department of Surgery, Sismanoglio General Hospital of Athens, Sismanogliou 1, Marousi, 15126 Athens, Greece
E-mail: agis.ioannidis@gmail.com

Key words: synovial sarcoma, ileus, monophasic synovial sarcoma

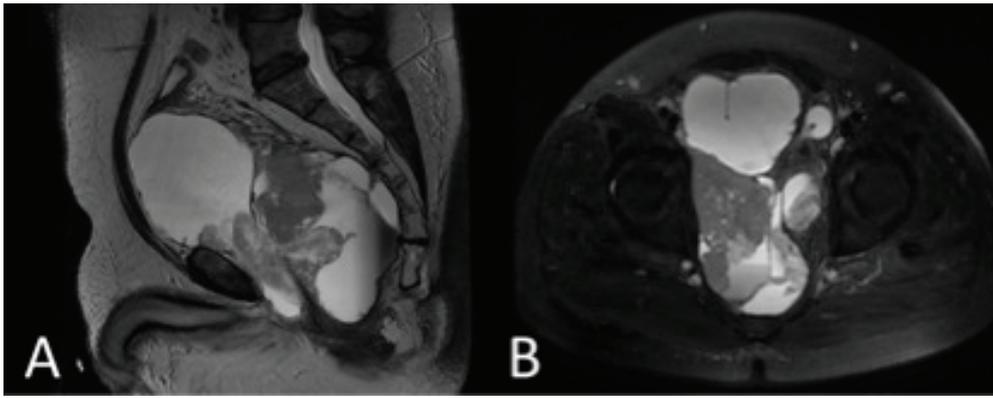


Figure 1. Abdomen pre-operative MRI scan. (A) Sagittal and (B) axial views of the abdomen, revealing the mass. MRI, magnetic resonance imaging.

The pathological examination revealed the presence of spindle and epithelioid cells, with marked nuclear pleomorphism and high mitotic index. Immunohistochemical investigation exhibited positivity for CD99, BCL2, S-100 and negativity for CD34, HHV8, podoplanin, CK8/18 myoglobin, SOX-10, SMA, caldesmon, desmin and MDM-2. Characteristics were compatible with the diagnosis of monophasic synovial sarcoma.

The patient was then referred to the Department of Medical Oncology. He was commenced on chemotherapy with doxorubicin and ifosfamide. He is currently on the third cycle and restaging is planned.

The present study has been approved by the Sismanoglio General Hospital of Athens bioethics committee. Patient consent was received.

Discussion

Synovial sarcoma is a rare and high grade soft-tissue sarcoma and has 3 types: Biphasic, monophasic and poorly differentiated, while it is characterized by a specific chromosomal translocation $t(X:18)(p11:q11)$ (1,2). Biphasic synovial sarcomas are characterized by the presence of spindle and epithelioid cells, while monophasic synovial sarcomas involve only one cell type (4). The most common site of synovial sarcoma are the lower extremities, followed by upper extremities, head and neck and trunk. It is of note that synovial sarcoma can occur in any region and at any age, but the peak incidence is the third decade and the most common site, the extremities (5). The incidence of synovial sarcomas was 0.9 in 1984, while in 2012 it was found to be 1.548/100,000 population (6). Older age, tumor size >5 cm, as well as trunk location have been characterized as risk factors for late stage disease. Patients younger than 25 years, with tumor size <5 cm and well-differentiated, are considered low-risk, whereas patients older than 25 years, with tumor size >5 cm and poorly differentiated are considered high-risk (7,8). Patients with trunk synovial sarcoma, such as the reported one, usually present with abdominal pain, bloated sensation and dyspepsia, while bowel obstruction has been described in cases of gastrointestinal tumor (9-11).

A case of a 54-year-old male with a monophasic synovial sarcoma (20x17x12 cm) causing obstructive ileus was presented. Ileus caused by synovial sarcoma is extremely

rare. There have been a few reports of obstructive ileus due to gastrointestinal synovial sarcomas, whereas in the present case the tumor originated from the pelvis, making it even rarer (9-11).

For cases with localized, low-risk disease treatment involves surgical resection and, in some cases, (neo)adjuvant radiotherapy (12). Previous studies reported metastases in 50% of those cases, while the 5-year survival is 60.5% (6,12,13). Additionally, some studies have shown a 10-year survival rate ranging between 24 and 68% (6,12-14). The most common metastases are found in the lungs, followed by lymph nodes, bone and liver. In metastatic cases, treatment is not considered curative (12).

In contrast to other soft tissue tumors, synovial sarcomas have proven to be relatively chemo-sensitive (12). Ifosfamide, as well as ifosfamide combinations have tested active in different treatment stages. In high-risk extremity and chest wall synovial sarcomas, neoadjuvant doxorubicin and ifosfamide has exhibited the same activity as high-dose stand-alone ifosfamide. Combination chemotherapy with doxorubicin and ifosfamide seems to improve outcome (12,15). In the first-line metastatic disease, combination with doxorubicin and ifosfamide is the preferred choice in relatively fit cases, while in the remaining patients sequential doxorubicin and ifosfamide may be administered. In second and subsequent lines, pazopanib and trabectedin have proven effective (12,15,16). Many new approaches to treat metastatic synovial sarcomas are currently under investigation, such as other receptor tyrosine kinase inhibitors, epigenetic modulators, compounds interfering with DNA damage response, and immunotherapy (12). The reported case had an unresectable tumor, due to its contact with major vessels. Following biopsy the patient was commenced on chemotherapy with doxorubicin and ifosfamide. He is now on the third cycle and restaging is planned.

Treatment of synovial sarcomas remains challenging and their incidence is on the increase. We presented an extremely rare case of obstructive ileus due to pelvic synovial sarcoma. This tumor has a poor prognosis, while considering this tumor in the diagnosis of non-specific symptomatology, such as abdominal pain and dyspepsia, would be an exaggeration. It is of paramount importance to report such cases in order to better understand their treatment options, as well as their pathophysiology. More studies are needed to identify improved chemotherapy agents with better outcomes.

Acknowledgements

Not applicable.

Funding

No funding was received.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Authors' contributions

CK, AI, AA contributed to data collection and the first manuscript draft. CK and AI conducted the literature search and analysis. AP, HD, GV, AMD contributed to the initial concept and final manuscript proofing. All authors have read and approved the manuscript.

Ethics approval and consent to participate

The Sismanoglio General Hospital of Athens bioethics committee has approved the present study.

Patient consent for publication

Patient consent was received.

Competing interests

The authors declare no conflicts of interest.

References

1. Fisher C: Synovial sarcoma. *Ann Diagn Pathol* 2: 401-421, 1998.
2. Clark J, Rocques PJ, Crew AJ, Gill S, Shipley J, Chan AM, Gusterson BA and Cooper CS: Identification of novel genes, SYT and SSX, involved in the t(X;18)(p11.2;q11.2) translocation found in human synovial sarcoma. *Nat Genet* 7: 502-508, 1994.
3. Corey RM, Swett K and Ward WG: Epidemiology and survivorship of soft tissue sarcomas in adults: A national cancer database report. *Cancer Med* 3: 1404-1415, 2014.
4. Spillane AJ, A'Hern R, Judson IR, Fisher C and Thomas JM: Synovial sarcoma: A clinicopathologic, staging, and prognostic assessment. *J Clin Oncol* 18: 3794-3803, 2000.
5. Vlenterie M, Ho VK, Kaal SE, Vlenterie R, Haas R, van der Graaf WT: Age as an independent prognostic factor for survival of localized synovial sarcoma patients. *Br J Cancer* 113: 1602-1606, 2015.
6. Wang S, Song R, Sun T, Hou B, Hong G, Mallampati S, Sun H, Zhou X, Zhou C, Zhang H, *et al*: Survival changes in patients with synovial sarcoma, 1983-2012. *J Cancer* 8: 1759-1768, 2017.
7. Deshmukh R, Mankin HJ and Singer S: Synovial sarcoma: The importance of size and location for survival. *Clin Orthop Relat Res* 419: 155-161, 2004.
8. Pan M and Merchant M: Risk factors including age, stage and anatomic location that impact the outcomes of patients with synovial sarcoma. *Med Sci (Basel)* 6: E21, 2018.
9. Chan GS, Yuen ST and Chan KW: Synovial sarcoma presenting as a polypoid jejunal mass. *Histopathology* 44: 191-193, 2004.
10. Alsharief AN, Fageeh M and Alabdulkarim Y: Monophasic synovial sarcoma presenting as a primary ileal mass: A case report and review of the literature. *J Med Case Reports* 6: 83, 2012.
11. Parfitt JR, Xu J, Kontozoglou T, Oluwafemi AR and Driman DK: Primary monophasic synovial sarcoma of the colon. *Histopathology* 50: 521-523, 2007.
12. Desai IME, Fleuren EDG and van der Graaf WTA: Systemic treatment for adults with synovial sarcoma. *Curr Treat Options Oncol* 19: 13, 2018.
13. Krieg AH, Hefti F, Speth BM, Jundt G, Guillou L, Exner UG, von Hochstetter AR, Cserhati MD, Fuchs B, Mouhsine E, *et al*: Synovial sarcomas usually metastasize after >5 years: A multicenter retrospective analysis with minimum follow-up of 10 years for survivors. *Ann Oncol* 22: 458-467, 2011.
14. Butori C, Hofman V, Attias R, Mouroux J, Pedetour F and Hofman P: Diagnosis of primary esophageal synovial sarcoma by demonstration of t(X;18) translocation: A case report. *Virchows Arch* 449: 262-267, 2006.
15. Vlenterie M, Litière S, Rizzo E, Marréaud S, Judson I, Gelderblom H, Le Cesne A, Wardelmann E, Messiou C, Gronchi A, *et al*: Outcome of chemotherapy in advanced synovial sarcoma patients: Review of 15 clinical trials from the European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group: setting a new landmark for studies in this entity. *Eur J Cancer* 58: 62-72, 2016.
16. Judson I, Verweij J, Gelderblom H, Hartmann JT, Schöffski P, Blay JY, Kerst JM, Sufliarisky J, Whelan J, Hohenberger P, *et al*: European Organisation and Treatment of Cancer Soft Tissue and Bone Sarcoma Group: Doxorubicin alone versus intensified doxorubicin plus ifosfamide for first-line treatment of advanced or metastatic soft-tissue sarcoma: A randomised controlled phase 3 trial. *Lancet Oncol* 15: 415-423, 2014.