Medical Science, Vol. 21, No. 84, March-April, 2017

CASE REPORT

ISSN
2321–7359
EISSN
2321–7367

Medical Science

An International Journal

Clear Cell Sarcoma-like Tumour of the Gastrointestinal Tract in a Leukemia Patient in Remission

Farsi A☼

General surgeon, Department of Surgery, Faculty of Medicine, King Abdulaziz University, Jeddah, Saudi Arabia

☼Correspondence to: Dr Ali Farsi, Department of Surgery, Faculty of Medicine, King Abdulaziz University Hospital, Pobox 80215, Jeddah 21589, Saudi Arabia

Publication History
Received: 24 December 2016
Accepted: 20 January 2017
Published: March-April 2017

Citation
Farsi A. Clear Cell Sarcoma-like Tumour of the Gastrointestinal Tract in a Leukemia Patient in Remission. *Medical Science*, 2017, 21(84), 75-78

Publication License
This work is licensed under a Creative Commons Attribution 4.0 International License.

General Note
Article is recommended to print in recycled paper.

ABSTRACT

Clear cell sarcoma-like tumour of the gastrointestinal tract is a very rare tumour affecting young adults. This is the case of a nineteen year old acute lymphocytic leukemia patient in remission after chemotherapy who was found to have this tumour on mesenteric lymph node biopsy. She was found to have a primarytumour in her small bowel which was successfully excised, with postoperative

© 2017 Discovery Publication. All Rights Reserved

pathology showing the immunohistochemical and genetic features typical for this neoplasm. These tumours are very rare and its important to identify and report all cases to gain a better understanding of their behaviour and treatment.

Keywords: Clear cell sarcoma-like tumour of the gastrointestinal tract, leukemia, S100, malignant gastrointestinal neuroectodermal tumour, EWSR1

Abbreviations: CCSLGT – Clear cell sarcoma-like tumour of the gastrointestinal tract

1. INTRODUCTION

Clear cell Sarcoma-like tumourof the gastrointestinal tract (CCSLGT) is a very rare tumour affecting the stomach, small bowel and colon in children and young adults (1). Clear cell Sarcoma was initially described by Franz Enzinger who initially coined the term Clear cell sarcoma of tendons and aponeurosis when he described them in the extremities (2). The first case to be described in the gastrointestinal tract was by Efkhors et al (3) in the duodenum. This very rare tumour has also been referred to as malignant gastrointestinal neuroectodermal tumour (4), as malignant melanoma of soft parts (5), and as osteoclast rich tumours of the gastrointestinal tract (9). Since then, few cases have been reported, mainly as case reports and case series. Thus, it still is a very poorly understood and difficult to diagnose entity.What follows is the case of a young female patient who presented with this rare tumour.

2. CASE REPORT

A nineteen years old female patient presented with a twelve month history of fatigue and shortness of breath. She had a background history of acute lymphocytic leukemia at the age of 3 which was treated with chemotherapy and had been in remission for 13 years. She was found to have a hypochromic microcytic anemia. This was felt to be due to iron deficiency but failed to improve with iron therapy. She subsequently required four packed red blood cell transfusions to treat her anemia. A bone marrow biopsy was performed and found no evidence of a malignancy. A PET CT was performed and it revealed concentric mural thickening of a loop of small bowel in the left lower quadrant along with multiple enlarged lymph nodes in the adjacent mesentery. A laparoscopic excisional biopsy was performed and the largest lymph node in the mesentery was successfully excised completely. The pathology of this showed clear cell sarcoma like tumour of the gastrointestinal tract. The patient then went on to have a laparotomy and small bowel resection of the thickened segment of jejunum with stapled anastomosis of the small bowel. In addition, the superior mesenteric vein was skeletonized and the adjacent enlarged lymph nodes in the mesentery were excised. Her postoperative course was uneventful and she was discharged from the hospital. She was discussed at the multidisciplinary sarcoma meeting and as all the gross disease had been removed, the decision was made to observe her with serial imaging.

2.1. Gross and microscopic features

Gross examination revealed an ulcerated tumour plaque in the small bowel, measuring 45 X 32mm and involving 95% of the small bowel circumference. It was 10mm in maximum thickness and appeared to involve the full thickness of the small bowel wall. It appeared to invade the mesenteric fat with about 5mm of extramuscular spread. The tumour did not appear to involve the serosa macroscopically. Microscopically, the tumour involved the mucosa, submucosa and muscularispropria. It had a predominantly nodular and solid growth pattern. Tumour cells were mildly to moderately pleomorphic, plump polygonal to sometimes spindled. Nuclei tended to be round to ovoid. Chromatin varied from granular to vesicular. Nucleoli tended to be small or inconspicuous. Cytoplasm varied from pale to eosinophilic or amphophilic, and cell borders were not well defined. Mitoses and apopotic bodies were readily identified. Scattered osteoclast-like giant cells were dispersed amongst the tumour cells. There was no lymphovascular, venous or perineural invasion identified.

With regards to the lymph nodes. There were metastatic deposits in five of the 25 harvested lymph nodes.

Farsi A,
Clear Cell Sarcoma-like Tumour of the Gastrointestinal Tract in a Leukemia Patient in Remission,
Medical Science, 2017, 21(84), 75-78,
www.discoveryjournals.com

© 2017 Discovery Publication. All Rights Reserved

2.2. Immunohistochemical and molecular genetic features

The tumour was found to be positive for S100. It was positive for AE1/AE3. It was positive for synaptophysin, with patchy staining of weak to strong intensity. The tumour was positive for EMA. It was positive for SMA, with staining patchy and granular. The tumour was negative for HMB45, Melan A, CAM 5.2, chromogranin, CD56, c-kit, DOG-1, CD34 and desmin. Ki67 proliferative index was rather high, being up to about 50%. Interphase FISH for EWSR1 (22q12) was performed with a significant proportion of tumour nuclei showing evidence of rearrangement.

3. DISCUSSION

Clear cell sarcoma-like tumour of the gastrointestinal tract is a very rare tumour with a poor prognosis that can be difficult to diagnose. Possible reasons for this include the rarity of CCSLGT with failure to suspect the diagnosis early, presenting symptoms are often vague and non-specific, the difficulty in distinguishing CCSLGT from melanoma due to the often similar morphology and immunohistochemistry and due to the fact that CCSLGT is an aggressive tumour with a high recurrence and metastasis risk (8).Our patient's history was significant for a previous history of acute lymphocytic leukemia treated with chemotherapy. Two other cases of this rare pathology have also been described in patients who previously had leukemia at an earlier age (6,7), suggesting that the association between of clear cell sarcoma like tumours of the gastrointestinal tract and chemotherapy should be looked at more closely. CCSLGT has also been reported to have occurred after childhood hepatoblastoma treated with surgery, radiation and chemotherapy (10) and in a patient with neuroblastoma during infancy treated with radiation therapy (11). As in our case, these tumours primarily affect the small bowel (70% of cases), and in over two-thirds, there is lymph node involvement at the time of surgery (8). Unlike our patient, many of these patients will have liver metastasis present at the time of diagnosis (8). This tumour is most common in young adults in the third and fourth decade of life, and there is no gender predilection (12).

CCSLGT tend to have a median size of about 5cm, and often will cause mucosal ulceration (13). Under the microscope, they tend to grow in a nested, solid or fascicular pattern (12). The cells tend to be polygonal, oval or spindle with clear or eosinophilic cytoplast and vesicular nuclei with prominent nucleoli, and multinucleated neoplastic giant cells can often be seen which suggests the diagnosis of CCSLGT (12, 13). Immunohistochemistry typically will be positive for S100 protein, SOX10 and vimentin while also being negative for HMB45, Melan-A, tyrosinase, microphthalmia transcription factor, CD117, DOG1 and CD34 (12, 14). Genetic studies will show either EWSR1-CREB1 or EWSR1-ATF1, which helps differentiate this rare tumour from melanoma (15).

4. CONCLUSION

Clear cell sarcoma-like tumour of the gastrointestinal tract is a very rare tumour affecting young adults that in this case presented as a second neoplasm after a childhood tumour treated with chemotherapy. The reason for this remains unclear. The rarity of this neoplasm and the poor understanding of its nature and risk factors makes it important to record each case diagnosed so the disease maybe better understood. It often has lymph node and liver metastasis and the prognosis is poor. It can be difficult to diagnose microscopically and immunohistochemical and genetic studies are an important part of establishing the diagnosis.

DISCLOSURE STATEMENT

There is no special financial support for this paper and the author has no conflicts of interest.

REFERENCES

1. Thway K, Judson I, Fisher C "Clear Cell Sarcoma-Like Tumor of the Gastrointestinal Tract, Presenting as a Second Malignancy after Childhood Hepatoblastoma", Case Reports in Medicine, Volume 2014 (2014), Article ID 984369
2. Enzinger FM " Clear-cell sarcoma of tendons and aponeuroses an analysis of 21 cases" Cancer 18: 1163-1174, 1965
3. Ekfors TO, Kujari H, Isomaki M. "Clear cell sarcoma of tendons and aponeuroses (malignant melanoma of soft parts) in the duodenum: the first visceral case" Histopathology. 1993;22(3):255–259
4. Kong J, Li N, Wu S, Guo X, Gu C, Feng Z "Malignant gastrointestinal neuroectodermal tumour: a case report and review of the literature" Oncol Lett. 2014 Dec;8(6):2687-2690
5. Chung EB and Enzinger FM "Malignant melanoma of soft parts. A reassessment of clear cell sarcoma" Am J SurgPathol 7: 405-413, 1983
6. Zambrano E, Reyes-Mugica M, Franchi A, Rosai J, "An osteoclast-rich tumor of the gastrointestinal tract with

© 2017 Discovery Publication. All Rights Reserved

features resembling clear cell sarcoma of soft parts: reports of 6 cases of a GIST simulator," International Journal of Surgical Pathology, vol. 11, no. 2, pp. 75–81, 2003

7. Venkataraman G, Quinn AM, Williams J, Hammadeh R, "Clear cell sarcoma of the small bowel: a potential pitfall. Case report," Acta Pathologica, Microbiologica, et ImmunologicaScandinavica, vol. 113, no. 10, pp. 716–719, 2005.
8. Mehta A, Brownson K, Massarani R, Esposito T ,Abood G, Shoup M "Clear cell sarcoma of the jejunum—Surgical management in two patients with review of the literature" Case reports in clinical medicine, Vol.2, No.4, 272-276, 2013
9. GahanbaniArdakani A, Boyle DJ, Elton C "Gastrointestinal clear cell sarcoma-like tumour of the ascending colon" Ann R Coll SurgEngl 2016; 98: e37–e39
10. Thway K, Judson I, Fisher C. "Clear cell sarcoma-like tumor of the gastrointestinal tract, presenting as a second malignancy after childhood hepatoblastoma" Case Rep Med. 2014;2014:984369
11. Balkaransingh P, Saad SA, Govil SC, Thind PK, Balance CM, Weiss AR. Clear cell sarcoma of the gastrointestinal tract presenting as a second malignant neoplasm following neuroblastoma in infancy. Pediatr Blood Cancer. 2012;58:481-482
12. Yegen G, Güllüoğlu M, Mete Ö, Önder S, Kapran Y"Clear cell sarcoma-like tumor of the gastrointestinal tract: a case report and review of the literature." Int J SurgPathol. 2015 Feb;23(1):61-7
13. Zhao Z, Zhang D, Li W, Zhang L, Li Z, Zhou J. "Primary malignant neuroectodermal tumor of the ileum with predominantly uncommon pseudopapillary architecture." International Journal of Clinical and Experimental Pathology. 2014;7(12):8967-8971.
14. Wang J, Thway K "Clear Cell Sarcoma–like Tumor of the Gastrointestinal Tract: An Evolving Entity" Archives of Pathology & Laboratory Medicine: March 2015, Vol. 139, No. 3, pp. 407-412.
15. Thway K, Fisher C (2012) "Tumors with EWSR1-CREB1 and EWSR1-ATF1 fusions: the current status". Am J SurgPathol, April 2012, 36:e1– e11

© 2017 Discovery Publication. All Rights Reserved