Primary carcinosarcoma of the spleen

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Citation: Joy ER, Cornish JA, Bootun R, Datta S, Chandran P. Primary carcinoma of the spleen. JSCR 2012 7:3

ABSTRACT

Carcinosarcomas are rare, malignant tumours normally arising from the uterus, ovaries and fallopian tubes and having both carcinomatous (epithelial cells) and sarcomatous (connective tissue) components. This case describes the first published case of a carcinosarcoma of the spleen presenting as microcytic anaemia.

A 74 year old lady presented with a two stone weight loss and was found to have a microcytic, hypochromic anaemia. On examination there was a non-tender mass in the left upper quadrant and a CT scan demonstrated a mass arising from the spleen. She underwent a splenectomy, of which the histology confirmed carcinosarcoma. She is currently undergoing palliative chemotherapy.

Although the condition is rare, it is important to be aware of this condition due to the poor prognosis and the unusual ways in which it can present.

INTRODUCTION

Carcinosarcomas are rare, malignant tumours, which as the name suggests, contain elements of both carcinomas (epithelial cells) and sarcomas (connective tissue) (1). Although normally arising from the uterus, ovaries and fallopian tubes (2), this case was unusual in that the lesion was found to be arising from the spleen. Previous literature report only three cases regarding carcinosarcomas of the spleen, with this being the first case presenting as microcytic hypochromic anaemia (1-3).

CASE REPORT

A 74 year old lady with a past medical history of hypertension, hypercholesterolemia and arthritis was referred from her GP with an unexplained weight loss of two stone in the preceding three months and associated microcytic hypochromic anaemia without an obvious source of blood loss.

She had undergone an oesophago-gastro-duodenoscopy (OGD) two months previously which had been normal.

On initial review by the colorectal surgeon, she was noted to have a non-tender mass in the left upper quadrant and denied any gastrointestinal symptoms. Her blood results at the time

were: Haemoglobin (Hb) 8.5g/dL, mean corpuscular volume (MCV) 71fL, mean cell haemoglobin (MCH) 23.6pg, Ferritin 722.4?g/L, Platelets 436x10^9/L, Albumin 29g/L and C-reactive protein (CRP) 99.5mg/L.

In light of the possible diagnosis of colorectal cancer she was referred to Colorectal Multi-Disciplinary Team (MDT) meeting and a CT demonstrated a large malignant left sided abdominal mass with likely splinting of the left hemi diaphragm and a left sided effusion as a result. At the time it was thought that the likely organ of origin was the spleen.

A few weeks later she underwent a splenectomy, without a prior biopsy due to the high risk of bleeding involved. It was noted that there was some adherence to the diaphragm in addition to a few serosal deposits of tumour in the gastro-colic omentum. Histology with further molecular pathology confirmed a primary splenic carcinosarcoma. Her recovery post-operatively was hindered by a lower respiratory tract infection, and she was discharged home after 10 days. Two months after her surgery, some improvement was noted in her Hb (9.8g/dL) and MCV (82fL). The patient was reviewed by an oncologist who advised treatment with palliative chemotherapy used for metastatic carcinoma of unknown origin, Epirubicin, Oxaliplatin and Capecitabine. Over a year later she is still undergoing treatment.

DISCUSSION

Previous cases of carcinosarcoma of the spleen by Roa et al (1), Westra et al (2), and Shah et al (3), all describe a presentation of left upper quadrant pain. In comparison, the unique findings in this case were weight loss as patient's only symptom and the microcytic, hypochromic anaemia. Despite having a firm mass in the left upper quadrant on examination, it was non-tender on palpation.

It is rare for the spleen to be the primary site of malignant epithelial neoplasms (4,7), with the commonest malignancy arising from the spleen being lymphoreticular in nature (6). In addition, despite the spleen's role as a haematogenous filter of effete cells and blood borne antigens it is not a favoured target of metastatic cancer either (3,7). In fact, nearly all cases of primary carcinoma of the spleen were reported before 1923, before technologies had developed to allow distinction between carcinomas and sarcomas (5).

In contrast with the previously reported cases, our case also involves the oldest patient known to have carcinosarcoma at the age of 74, with previously reported cases ranging between 55 and 60 years, in addition to the lightest weight of any carcinosarcoma, which have ranged between 1680g and 2330g. As carcinosarcomas most commonly arise from the uterus, ovaries, fallopian tubes and vagina in descending order of frequency (3), it seems safe to assume that when they are located elsewhere in the body they are also more likely to be in women, as was the case here. This is also seen in the previously documented cases, with a gender split of 2:1 in favour of females.

Despite only one case documenting metastases at the time of splenectomy, all cases developed widespread metastatic disease in the months following surgery with the longest

survival 9 months post splenectomy. This enforces the general oncological rule that unusual cancers often behave aggressively with poor patient prognosis (1). However, our case is the longest survivor of this condition.

In conclusion, this case illustrates the painless presentation of a rare malignant condition usually associated with left upper quadrant pain. Despite the rarity of this condition, research is needed into the histogenesis and management of carcinosarcomas of the spleen, which currently carry an extremely poor prognosis.

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