ATYPICAL PRESENTATION OF SOFT TISSUE SARCOMA

Editor,

Soft-tissue sarcomas are rare malignancies of mesodermal origin and constitute <1% of new malignancies. There are multiple histological sub-types and only about 10% of all sarcomas are synovial. They usually present as a painless limb mass. Metastases can be as common as 40% with high-grade tumours and occur most commonly in the lung, but may also affect liver, brain and bone

A 27 year old man presenting with left sided chest pain was found to have multiple soft tissue lung lesions on chest x-ray (Figure1). A CT guided biopsy was unsuccessful and referral for VATS (video assisted thoracoscopic surgery) was made to obtain tissue samples.

At review 1 month after the initial chest X-ray, the presence of long-standing inflammation of the right foot became evident. The right foot had been inflamed for 6 months and was significantly larger than the left with mild erythema (Figure2). MRI of the foot revealed a large lesion at the medial aspect with bony erosion consistent with a malignant tumour.

VATS identified large abnormal lung nodules (Figure3) confirmed to be malignant on frozen section. The histology was not typical of lung carcinoma, melanoma or common pleural/lung lesions. Immunohistochemistry showed positive staining of biphasic spindle cells with CD99, EMA and bcl-2, as well as focal staining with Cam 5.2 and AE1/3 in the epithelioid areas. This favoured a diagnosis of synovial sarcoma confirmed with the presence of t(X;18) translocation identified by molecular studies. No further biopsy of the primary lesion was deemed necessary to confirm the diagnosis of metastatic synovial sarcoma.

The t(X;18) chromosomal translocation identified in the sample sent for molecular pathology is typical for synovial sarcoma and essentially describes a fusion between SSX on chromosome X and SS18 on chromosome 18 2,3. This translocation is virtually pathognomonic for human synovial sarcoma 3. The fusion process is similar to the oncogenesis of certain leukaemias 3.

The stable SS18-SSX fusion protein created as a result of the translocation is subsequently incorporated into the BAF complex. This prevents the BAF47 subunit, usually part of the complex, from inclusion. BAF47 is a tumour suppressor gene and its absence may play a significant role in the development of synovial sarcoma 2. Interestingly, proliferation of synovial sarcoma may be potentially reversible if normal complexes are reassembled. This may guide the development of future therapeutic agents 3.

This case highlights the importance of a thorough patient review. This was an atypical presentation of synovial sarcoma with the pulmonary metastases identified prior to the primary

Fig 1 Chest X-ray showing multiple lung nodules

Fig 2 Swollen right foot as compared to left foot, initially thought to be unrelated

Fig 3 Macroscopic images of lung nodules during VATS procedure
malignancy. It is unusual for synovial sarcoma to be identified from incidental findings of lung metastases. The patient was treated with trabectedin instead of doxorubicin in view of compromised left ventricular ejection fraction. Trabectedin is at least as efficacious as doxorubicin for the treatment of translocation-related sarcomas. One year after initial diagnosis he is receiving palliative chemotherapy, with metastases mostly stable in size. The mass in the right foot has continued to grow in size, as have some of the pulmonary nodules.

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NO BABY BOOM OR SEX RATIO CHANGES FOLLOWING FIFTY SHADES OF GREY IN ENGLAND AND WALES

Editor,

_Fifty Shades of Grey_ (FSOG) was a 7/2011 erotic romance that traces the deepening and complex relationship between a college graduate and a young business magnate, with soft porn elements that include bondage/discipline, dominance/submission, and sadism/masochism. The media hyped this, claiming FSOG “sparked a “mommy porn” revolution”. The second and third volumes of FSOG were published in 4/2012. A film adaptation of the first book was released on 13 February 2015.

FSOG was touted as inciting increased coital activity, thereby potentially resulting in a baby boom. “It’s one of the hottest and best-selling book series of all time. It has made millions of readers swoon”. August authorities were cited as confirming this effect. Robin Milhausen, an Associate Professor of Family Relations and Human Sexuality at the University of Guelph, was quoted: “the material is arousing… Many women respond to the book and don’t even know it. It’s leading to more sex”.

The male to female ratio at birth (male divided by total births: M/T) approximates 0.515 (slight males excess). Numerous factors may affect M/T. Increased coital activity may not only increase the birth rate but also increases M/T as sex ratio at conception follows a U-shaped regression curve on cycle day of insemination. Thus, increased coital activity will increase the likelihood of conception early in the cycle, increasing male conceptions.

This study sought spikes in total births or M/T in England and Wales circa nine months following FSOG books.

METHODS

Monthly male and female births for England and Wales were obtained from the Office for National Statistics for 1/99-8/99 (Ms. Athena Ray – personal communication). The null hypothesis was that FSOG releases in 7/2011 and 4/2012 did not influence total births and M/T circa nine months later, i.e. 4/2012 and 1/2013.

RESULTS

This study analysed 11831728 live births (M/T 0.5128, 95% CI 0.5125-0.5130). Annual births and M/T shows no discernible spikes (figure 1). A monthly breakdown for 1/2010-8/2016 shows no discernible spikes in total births or M/T at/around 4/2012 and 1/2013 (figure 2).

DISCUSSION

Linda Murray, Global Editor-in-Chief of BabyCenter.com stated that “reading ‘50 Shades of Grey’ is acting like an aphrodisiac for women…It’s putting them in the mood more frequently and they’re having more sex and they’re ultimately getting pregnant faster”. And the Daily Mail averred that “the meteoric rise of Fifty Shades of Grey is set to spark a new wave of births, according to pregnancy and parenting websites”. FSOG was therefore anticipated to result in a “revolution … coming to the delivery room, where a baby boom sparked by the “Fifty Shades of Grey” phenomenon is predicted”.

![Fig 1. Annual births and M/T for England and Wales, 1999-2015](image1.png)

![Fig 2. Monthly births and M/T for England and Wales, January 2010-August 2016](image2.png)