A case of thoracic splenosis in a post-splenectomy patient following abdominal trauma: Hello Howell–Jolly

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Received 24 February 2014; revised 10 July 2014; accepted 15 July 2014

Seeding of splenic tissue to extra-abdominal sites is a relatively infrequent consequence of open abdominal trauma. Immunological function of these small foci of ectopic splenic tissue is unknown and their use in determining the splenic function may be limited. In this case report, a patient is described who had previously undergone an emergency splenectomy. The absence of Howell–Jolly bodies on the blood smear in a patient who had previously undergone surgical splenectomy raised the suspicion of splenosis. The immunological features as well as non-invasive evaluation of these ill-defined splenic tissue sites are discussed.

INTRODUCTION

Howell–Jolly bodies are nuclear DNA remnants of erythrocyte precursor cells that are usually expelled from the mature erythrocyte or phagocytosed by the normal spleen. In individuals who are completely asplenic (whether functionally or by surgical intervention), Howell–Jolly bodies appear in the peripheral blood due to lack of phagocytic function. The absence of these bodies in splenectomized patients potentially alerts clinicians to the presence of functional splenic tissue [1].

Splenosis is defined as the seeding of splenic tissue to ectopic sites, usually within the abdominal or thoracic cavity. Various atypical locations have been described which may cause diagnostic confusion with malignant tumours [2].

CASE REPORT

A 55-year-old Caucasian male was referred from another clinical department as he had undergone a routine chest radiograph that revealed multiple mediastinal and diaphragmatic nodules, raising concern over possible malignancy. The patient was employed as a Chemical Engineer at various chemical plants and exposed to a large array of chemical fumes, including arsenic. He was a smoker until 1 year before presentation and was previously exposed to asbestos related to his occupation as engineer in the mining and construction industry. His surgical history included abdominal gunshot wounds suffered in the 1970s while he was enrolled in military service, one which led to an emergency splenectomy. His medical history included admission for severe malaria in the mid-1980s, which required admission to an intensive care unit.

The absence of Howell–Jolly bodies on the peripheral blood smear at presentation (Fig. 1, left) was a possible clue to the presence of ectopic splenosis as their presence would be expected in a fully splenectomized patient. A CT scan of the chest (Fig. 2) was requested without contrast due to impaired baseline renal function. This revealed pleural plaque formation, as well as enlarged mediastinal lymph nodes together with multiple pleural- and mediastinal-based soft tissue nodules. Some of these nodules were calcified. Basal ground-glass opacification was also noted. Differential diagnosis of these lesions includes both primary and metastatic malignancies (e.g. malignant mesothelioma, lymphoma and metastatic adenocarcinoma) as well as non-malignant tumours (e.g. lipoma) and various infectious and non-infectious causes (e.g. asbestosis and sarcoidosis). Granulomatous infection due to M. tuberculosis was an important consideration as South Africa is a country burdened by a very high prevalence of disease due to this pathogen. Due to his increased occupational exposure asbestosis was a possibility and this would also explain the early ground-glass opacification. Sarcoidosis was considered, although no systemic or extra-pulmonary manifestations were present. The inability to perform a contrasted CT scan limited our ability to narrow the differential diagnosis in this patient.

The patient therefore underwent formal thoracotomy and excision biopsy at multiple sites. Histological examination of
the relevant sites revealed splenic tissue with white and red pulp, secondary germinal centres, lymphoid follicles and granulocytes, surrounded by a fibrotic ring. These findings were consistent with splenosis in all the relevant biopsies. No evidence of malignancy was observed (Figs 3 and 4).

At 1-year follow-up, the patient remained well and the appearance of Howell–Jolly bodies was noted on repeat blood smear (Fig. 1, right), following the previous excisions.

**DISCUSSION**

A common precipitating event in many case reports of thoracic splenosis is that of penetrating abdominal trauma together with diaphragmatic injury, with the usual sites of thoracic seeding being the sub-pleural mediastinal and diaphragmatic surfaces [3]. In this report, the splenic seeding at distant sites occurred after penetrating abdominal wounds. Histologically, these tissues contain both red and white pulp, and may be surrounded by a thick fibrous capsule that tends to be thicker than that of an accessory spleen. In addition, they are characterized by the absence of proper feeding vessels with distortion of structural architecture, when compared with accessory spleens. They may be radiologically indistinct, although accessory spleens are usually confined to the peri-splenic and peri-pancreatic areas [4]. Clinically, these lesions may cause chronic pain or remain asymptomatic for years and only be discovered incidentally. In this case study, they were serendipitously found on routine X-ray of the chest during evaluation for another problem.

From an immunological perspective, the presence of red cell remnants with regard to splenic function is not fully known. The correlation between haematological markers such as Howell–Jolly bodies has been evaluated by studies in patients with medical causes for hyposplenism, such as sickle cell disease and coeliac disease, as well as embolization...
utilized in portal hypertension [5]. The negative predictive value in the absence of Howell–Jolly bodies have been shown to be poor and could therefore not be used to argue for splenic function in this patient, although the presence of Howell–Jolly bodies do argue for a functional splenic volume <30% with a slightly better positive predictive value [6]. One study reporting on the use of Howell–Jolly bodies failed to offer a standard percentage at which hyposplenism could be diagnosed, and indeed cases of mildly decreased splenic function were not detected [7]. Currently, Tc99-labelled, heat-altered, autologous erythrocyte scintigraphy with SPECT/CT scanning is viewed as the ‘gold standard’ for the purpose of assessing immune status in hyposplenism, being able to quantify both structure and volume. Heat alteration artificially damages the surfaces of erythrocytes, which primes them for phagocytosis by normal splenic tissue. Both sensitivity and specificity are increased compared with 99mTc-labelled sulphur colloids, due to the absence of large hepatic clearance [8]. The quantification of Howell–Jolly bodies by flow cytometry correlates poorly with these scintigraphy scans, limiting their clinical use [9].

The first clinical issue raised by this case is the degree of long-term immune protection provided by ectopic splenic tissue and whether this should alter the approach to post-splenectomy vaccination. As marked splenic dysfunction leading to post-splenectomy sepsis or overwhelming post-splenectomy infection may occur even in the presence of signs indicating some limited splenic function (such as red cell remnants), patients should be advised to adhere to the standard post-splenectomy vaccination guidelines as if no functional spleen was present. Prudent to our case severe malaria has a high mortality rate and if possible avoidance of endemic areas is still recommended over anti-malarial prophylaxis only in asplenic individuals [10].

The second clinical issue is whether an invasive procedure is required for the diagnosis of thoracic nodules suspected to be splenosis. It is well established that radionuclide scanning is a useful and reliable confirmatory test that obviates the need for unnecessary surgical interventions and reduces patient anxiety, although this case report presents an exception to this rule due to the limitation on contrast scanning combined with a broad and potentially malignant differential diagnosis. The most frequent symptom relating to thoracic splenosis is that of chest pain and dyspnoea. In symptomatic patients surgical intervention might be indicated after exclusion of other causes. No treatment is recommended in asymptomatic cases as these lesions appear to have a benign long-term course.

ACKNOWLEDGEMENTS
The author thanks Prof. J. Ker and Prof. G. Tintinger for reviewing the original manuscript and Dr J. Du Plessis and G. Laubscher (Lancet Laboratories) for the provision of the histology slides.

FUNDING
No funding was obtained in the preparation of this case report.

CONFLICT OF INTEREST
No conflicts of interest to declare.

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