Pancytopenia and an abdominal mass

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Cases of duplicitas asymmetros should be regularly followed up as there is a risk of fetal remnants developing into teratomatous growths.

Case report

A 35-year-old woman presented with a one-day history of stabbing, central chest pain radiating to her left upper abdomen and back. This followed two weeks of general malaise associated with cough, fevers, joint pain and night sweats. Her significant past medical history was of beta-thalassaemia minor with associated splenomegaly and five miscarriages. She was born in Jamaica as a sole-surviving conjoined twin. On inspection of the abdomen, there were two horizontal midline scars where the remnants of the smaller infant had been resected.

Examination revealed that she was febrile and had a tachycardia of 120 bpm with an ejection systolic murmur, loudest over the aortic region. There was marked hepatosplenomegaly, cervical lymphadenopathy and swelling of the joints of the hands bilaterally. Her blood results showed pancytopenia (haemoglobin 9.3 g/dL, white cell count 1.8 × 10⁹/L, platelet count 38 × 10⁹/L) with a notable neutropenia (1.3 × 10⁹/L), raised C-reactive protein (72 mg/L), and a negative HIV test. She was treated empirically with doxycycline.

The patient’s neutrophil count gradually fell to 0.8 (× 10⁹/L), at which point granulocyte colony-stimulating factor was administered. She refused bone marrow aspiration. In addition to the haematological consequences of hypersplenism, it was thought likely that both ethnic neutropenia and administration of doxycycline contributed to her declining neutrophil count.

An echocardiogram showed a moderate-sized pericardial effusion localized posteriorly and hyperdynamic systolic function. Pericarditis in the presence of presumed neutropenic sepsis was diagnosed and was treated with non-steroidal anti-inflammatory drugs. After recovery, the patient underwent further investigations for autoimmune disease as an outpatient.

An abdominal ultrasound scan with portal vein Doppler imaging showed abnormal vessels in the porta hepatitis extending into the hilum of an enlarged 18 cm spleen. There was arterial and venous signal with flow in the direction of the portal vein. Due to the unusual appearance of the vessels, the differential diagnosis was between atypical varices or residuals from the patient’s co-twin.

Computerized tomography confirmed the presence of abnormal blood vessels and showed an area within the liver containing fat, calcification and an air-filled cavity with a soft tissue polyp.
extending into it, lying between the hepatic portal vein and the inferior vena cava (Figures 1 and 2). The mass was consistent in appearance with a teratoma and was believed to comprise remnants of the co-twin. The multidisciplinary team opted not to resect the hepatic mass at this stage due to the risk of damage to the portal veins.

The patient’s splenomegaly was attributed to a combination of beta-thalassaemia minor, which was likely to form the underlying cause of her pancytopaenia, and to extrahepatic portal hypertension, caused by compression of the portal vein by the teratoma.

Discussion

Parasitic twinning or duplicitas asymmetros is an extremely rare condition in which fission of the ovum not only fails to complete, producing conjoined monochorionic monoamniotic embryos, but the partial division is asymmetrical, leading to the development of a smaller, malformed co-twin attached to its more anatomically normal sibling.

Although the incidence of duplicitas asymmetros is low, this case illustrates the importance of following up such infants as there is a risk of fetal remnants developing into teratomatous growths. We believe this to be the first reported case of a hepatic teratoma developing secondary to duplicitas asymmetros.

References

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