Spontaneous Splenic Rupture Secondary to Phaeohyphomycosis and Splenic Abscesses

N Sithasanan, FRCS, L A Chong, MRCPCH, H Ariffin, MRCP
Division of Paediatric Surgery, University Malaya Medical Centre, 50603 Kuala Lumpur

SUMMARY
Phaeohyphomycosis consists of a group of mycotic infections characterized by the presence of dematiaceous (dark walled) septate hyphae. Splenic abscess and spontaneous rupture is an infrequent complication in children with haematological malignancies and can be life threatening. To the best of our knowledge this is the first report of a case of splenic rupture following the development of multiple abscesses secondary to infestation by this rare fungal species.

KEYWORDS:
Phaeohyphomycosis, Splenic abscess, Splenic rupture

CASE REPORT
A four year old girl diagnosed with precursor-B cell acute lymphoblastic leukaemia (ALL), five days after commencement of induction therapy, developed pyrexia that did not subside despite the use of broad spectrum antibiotics. The chemotherapy was continued as she was not overtly toxic and blood cultures were negative. The fever persisted warranting antifungal cover, and Amphotericin B was added. A surveillance Computerized Tomogram (CT) scan of the thorax and abdomen in search of a focus for the infection revealed no focal lesions. Nine days into antifungal therapy the pyrexia subsided only to re-emerge four days later, prompting a second CT scan of the abdomen. The CT scan of her abdomen revealed multiple hypo dense lesions in the spleen with enhancing peripheries (Fig. 1) the largest lesion located in the centre of the body of spleen closer to the diaphragmatic surface. These lesions were highly suggestive of abscesses although blood cultures were still negative. Antifungal therapy was therefore changed to Itraconazole. Two days following the commencement of the newer antifungals she developed a spiking temperature and in addition developed left shoulder tip pain and an acute abdomen.

An emergent laparotomy revealed 500 mls of a haemoperitoneum secondary to a ruptured spleen that was studded with abscesses (Fig. 2). The diaphragmatic surface of the spleen in the area of the large abscess had ruptured. Certain other smaller abscesses were seen on the diaphragmatic surface of the spleen and were closely adherent to the diaphragm, with a few perforating it into the left hemithorax.

A splenectomy was performed and the abdomen thoroughly lavaged. Following a very stormy post operative period complicated by sepsis and right lower lobar pneumonia she finally recovered and was able to recommence her chemotherapy for the ALL 14 days after the surgery.

The histology of the spleen revealed multiple macro and micro abscesses infested with dematiaceous fungi. The exact species could not be identified due to gross necrosis of the tissues.

DISCUSSION
Splenic rupture is an infrequent complication in acute leukaemia. Although the incidence of detection has
improved with our better understanding and imaging techniques it still remains very rare with a reported incidence of 0.18% in children with various haematological malignancies. Factors that account for spontaneous splenic rupture are a high leukocyte count, thrombocytopenia, and coagulaopathy. Coexisting fungal infection may in addition contribute to the rupture.

Phaeohyphomycosis is a group of more than a hundred species of fungi that reside in the soil and cause a variety of infections involving skin and subcutaneous tissues, brain, solid viscera like liver, spleen and kidneys and disseminated infections too. Their dark colour is attributable to the presence of melanin in their cell walls. The melanin apart from imparting the dark colour also contributes to the virulence of these organisms by enabling them scavenge free radicals and hypochlorite produced by the host phagocytes, thus preventing their demise in battle. In addition the fungus is capable of producing a structure that penetrates host cells. These factors therefore also explain the potency in immunocompetent hosts. A high index of suspicion is required to diagnose this extremely rare infection and prompt and appropriate therapy is of paramount importance given their resistance to conventional antifungals like Amphotericin B. They appear to only be sensitive to the newer antifungal agents like Itraconazole and Terbafine. Although discrete abscesses may be amenable to treatment with intervention radiological techniques, multiple splenic abscesses and certainly a ruptured spleen require a splenectomy. To the best of our knowledge this is the first case of a spontaneous splenic rupture secondary to a splenic abscess caused by a dematiaceous fungus.

Splenic abscesses are rare and carry a high morbidity. The traditional treatment is splenectomy although with improved imaging and techniques discrete abscesses may be amenable to percutaneous drainage. The detection rate of these abscesses on ultrasound and CT scans is enhanced by normalization of the neutrophil counts since the inflammatory reaction of the tissues then enhances their detection on CT scans.

CONCLUSION
Chemophrophylaxis against fungal infections is fairly standard in immunocompromised children. Unrelenting sepsis and abdominal signs of peritonitis in such patients should raise the suspicion of abscess formation in solid viscera and subsequent rupture. A higher index of suspicion is required to rule out phaeohyphomycosis which is resistant to traditional anti fungal therapy and commence treatment with appropriate newer anti fungals like Itraconazole and Terbafine. Although discrete abscesses may be amenable to treatment with intervention radiological techniques, multiple splenic abscesses and certainly a ruptured spleen require a splenectomy. To the best of our knowledge this is the first case of a spontaneous splenic rupture secondary to a splenic abscess caused by a dematiaceous fungus.

REFERENCES