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Gut Intrigue: Navigating a Diagnostic Challenge and Uncommon Presentation of Bowel Wall Calcification

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Case Background

- Demographics: 54 year-old man from the Philippines who moved to the UK in 2003
- Presenting complaint: confusion, deranged liver function tests, hyponatraemia (124 mmol/L) and hypercalcaemia (2.5 mmol/L)
- PMH: insulin dependent T2DM
- Neurological examination: No meningism, bilateral past pointing and dysdiadochokinesia, power 5/5 all areas, reflexes 2+ bilaterally
- Respiratory examination: normal
- Initial imaging investigation: MRI brain with contrast

MRI Head

Cerebral and cerebellar hemisphere lesions with subcortical and leptomeningeal distribution. Post contrast enhancement with larger lesions rim enhancing. Lesions demonstrate restricted diffusion.

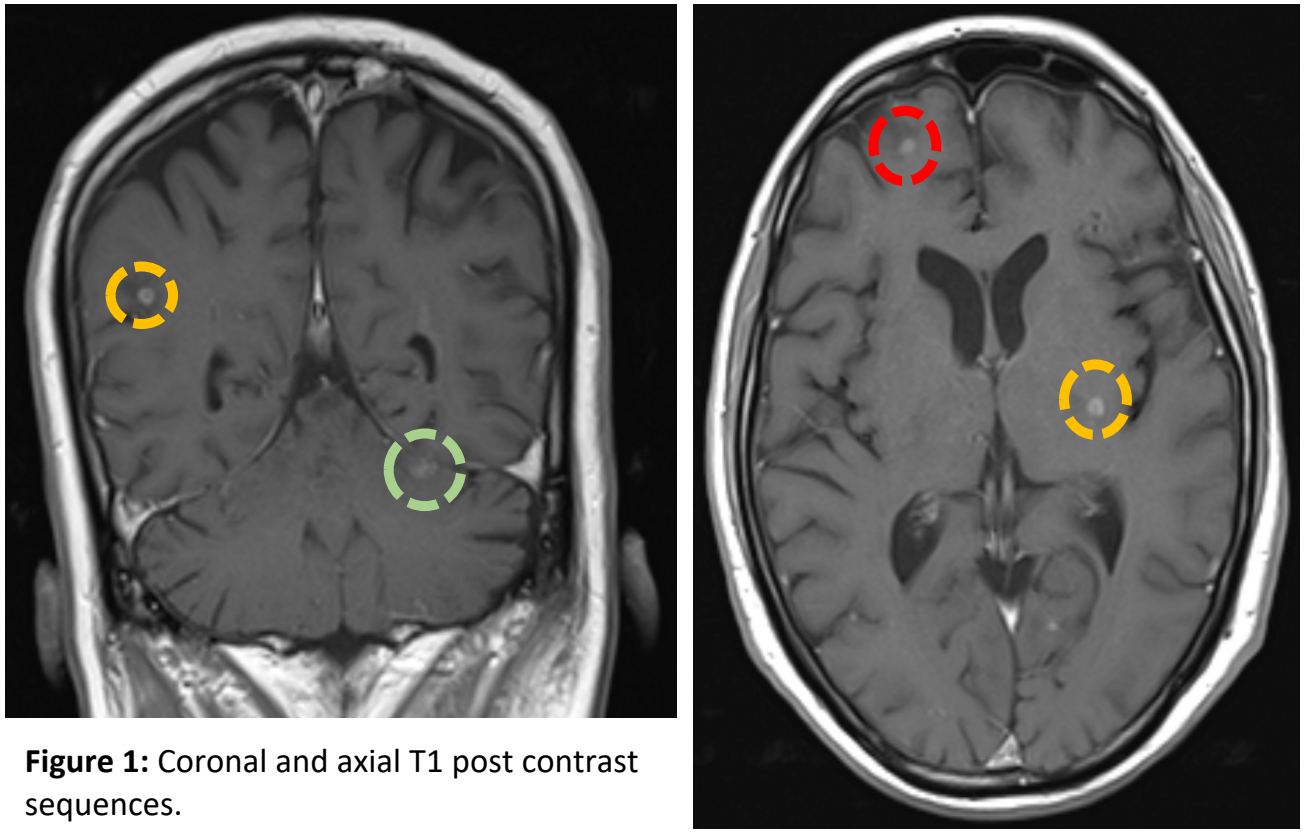

Figure 1: Coronal and axial T1 post contrast sequences.

- Subsequent sputum AFB positive
- Diagnosed with miliary tuberculosis with CNS involvement and commenced quadruple therapy
- Hypercalcaemia felt to be secondary to granulomatous disease

Further Workup

- Dysuria and elevated urine WCC prompted an ultrasound Urinary Tract which revealed an atrophic right kidney with pelvicalyceal dilatation.
- Mural calcification of the urinary bladder was noted on ultrasound.
- He was noted to clinically have a palpable bladder and a CT Urogram was performed.

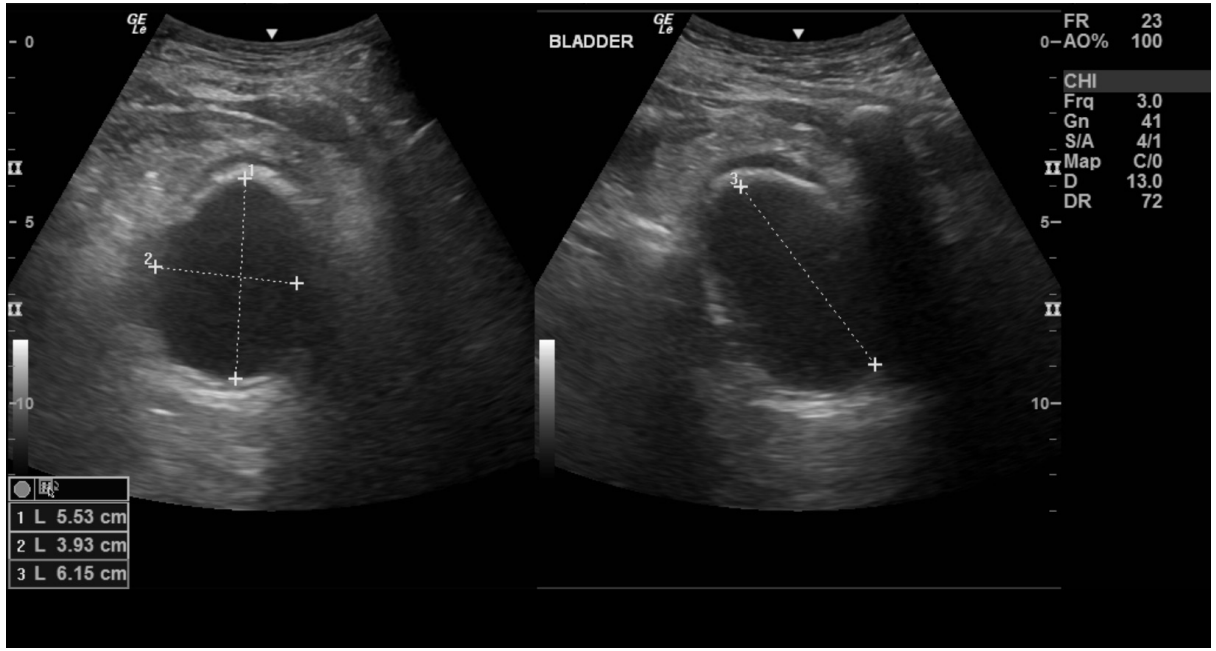

Figure 2: B-mode ultrasound images showing mural calcification of the urinary bladder.

Further Workup

- CT Urinary Tract pre and post contrast demonstrated a characteristic right "putty kidney“ seen with tuberculosis, left distal ureter focal narrowing, and new eggshell calcification of the bladder and urethra.
- The aetiology was unclear, either reflecting further change related to genitourinary tuberculosis or schistosomiasis. Flexible cystoscopy was initially challenging and nondiagnostic due to the urethral calcification.
- Management solely with tuberculosis treatment was continued as serology and urine were negative for Schistosome.

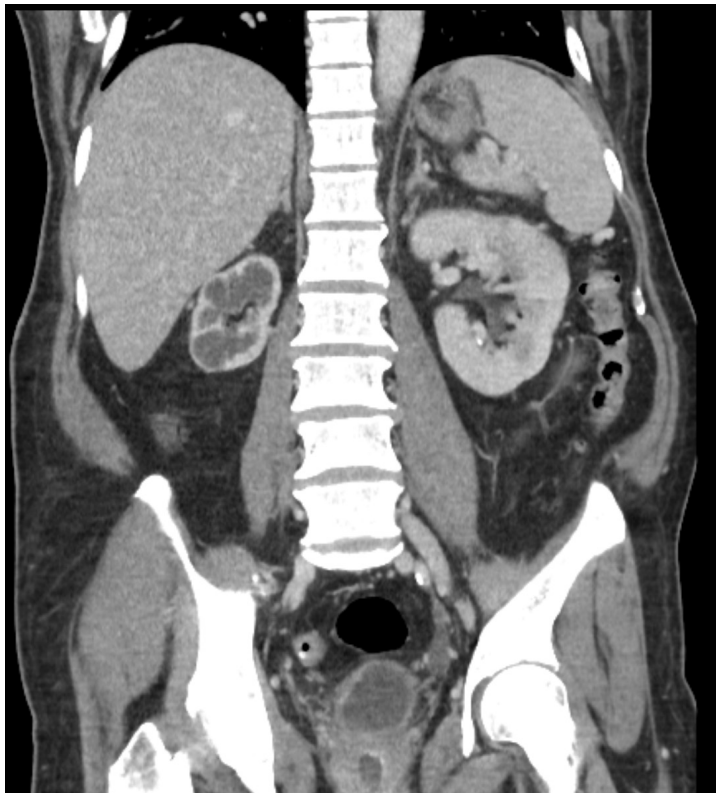

Figure 3: Coronal CT showing atrophic right kidney with cortical thinning and pelvicalyceal dilatation secondary to tuberculosis

Figure 4: Sagittal CT showing eggshell calcification of the urinary bladder and the urethra. Flecks of calcification in the rectum are also seen.

Follow up imaging

- Follow-up CT abdomen and pelvis with contrast to reassess the urinary tract showed stable urinary tract appearances.
- Additional note was made of mural calcification on the luminal aspect of the rectum and sigmoid colon including antidependent areas.

Figure 5: Axial CT showing more extensive linear mucosal/submucosal calcification of the sigmoid colon.

Figure 6: Sagittal CT showing bladder wall thickening and calcification.

Further investigation

- Due to persistent LUTS and inability to catheterise at the bedside despite multiple attempts, he was commenced on antimuscarinics.
- A rigid cystoscopy was performed under general anaesthetic, which confirmed calcification in the urethra and bladder, showed a non-occlusive prostate, and a false passage in urethra with narrow urethral lumen
- Urethral dilatation was performed, two biopsies were taken and a two-way catheter was inserted.
- Flexible sigmoidoscopy was performed with biopsies of the mural calcification taken the following day.

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Differentials for calcification of the bowel wall

Uncommon finding which may be secondary to benign, premalignant, or malignant lesions

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Relevant differentials in our case:

- Tuberculosis (concomitantly present in this patient): usually ileocaecal, strictures and fistulas may complicate.
- Metastatic calcification: deposition of calcium salt in normal tissue with an abnormal serum biochemical environment, such as chronic kidney disease, hyperparathyroidism, and hypercalcemia (our patient was initially hypercalcaemic).
- Schistosomiasis: tropical infection with wide endemic region.

Other differential diagnoses:

- Primary tumours of the GIT: gastrointestinal stromal tumour (GIST), mucinous adenocarcinoma (appendiceal or large bowel; associated with peritoneal and hepatic metastases).
- Haemangioma: usually present as occult or overt bleeding.
- Amyloidosis: gastric mucosa may display thickened folds, which may appear nodular or mass-like and which may contain calcifications.
- Phlebosclerotic colitis: unique form of ischemic colitis characterized by thread-like calcifications along affected colonic wall and mesenteric vein, right hemicolon, Asian patients typically, gradual and chronic presentation.

Final diagnosis and management

- Whilst the urine samples contained no ova cyst or parasites and Schistosoma serology was negative, the urethral biopsies grew Schistosomiasis japonicum.
- Biopsies from the sigmoid colon also grew Schistosome ova.
- Bowel wall calcification was retrospectively noted to predate the onset of bladder calcification on previous imaging.
- The patient was been treated with praziquantel. There was no subsequent development of bowel symptoms.
- He completed quadruple therapy for tuberculosis contemporaneously.

Literature Review: Schistosomiasis geographic distribution and diagnosis

- Schistosomiasis, also known as bilharziasis is a multisystem infectious disease
- It is caused by a blood fluke (trematode) of genus Schistosoma.
- It is the second most common tropical/sub-tropical disease after malaria.
- The three most common species to affect humans are S. haematobium, S. mansoni and S. japonicum
- The endemic region is vast and dependent on species.
  - S. haematobium infection occurs in the Middle East and Africa,
  - S. mansoni is present in Middle East, Africa and South America.
  - S. japonicum is found in China, Indonesia and the Philippines

- There are acute and chronic phases of the disease.
- Chronic Schistosomiasis may be encountered in the UK in patients who have migrated from endemic areas. These patients tend not to present in the acute phase due to repeated exposure to the parasite in those endemic regions.
- Serological abnormalities include eosinophilia, mild to moderate anaemia, raised alkaline phosphatase and an increase in immunoglobulin levels, particularly immunoglobulin G.
- Diagnostic tests include urine microscopy, faecal smear testing and histological analysis of biopsy samples.

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Literature Review: Schistosomiasis gastrointestinal tract involvement (distribution)

- S. haematobium typically affects the urogenital tract, whereas S. mansoni and S. japonicum cause hepato-intestinal manifestations.
- Calcification is seen in urogenital schistosomiasis more often than gastrointestinal with S. haematobium more ova are laid, and the ova calcify more readily.
- Clinical features of intestinal Schistosomiasis include colic and diarrhoea, haematochezia and PR mucus, between two months and two years of the initial infection.
- The colon eventually becomes a fibrous tube, largely devoid of mucosa. Submucosal oedema, ulcers and fibrosis may result. Polyposis is also a recognised complication.
- The rectum and sigmoid colon are the most common intestinal involvement sites of schistosomiasis.
- The small bowel may be involved but this is much less common.
- The liver is commonly affected. Calcification may occur in the liver capsule and parenchyma. Parenchymal calcification tends to be oriented perpendicular to the capsule. This results in a tortoise-shell appearance on CT and. A lacy appearance on ultrasound. The hepatic lobules themselves are unaffected.
- Periportal fibrosis and resultant portal hypertension can mimic liver cirrhosis. It is important to accurately delineate the aetiology as management and prognosis differ, Important imaging differentiators include a lack of surface nodularity and regenerative nodules in hepatic schistosomiasis.

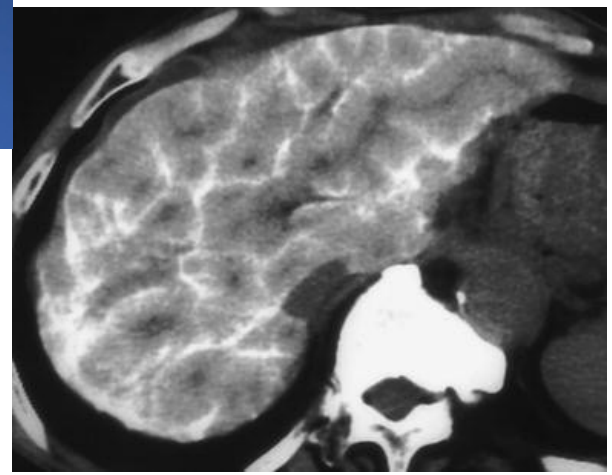
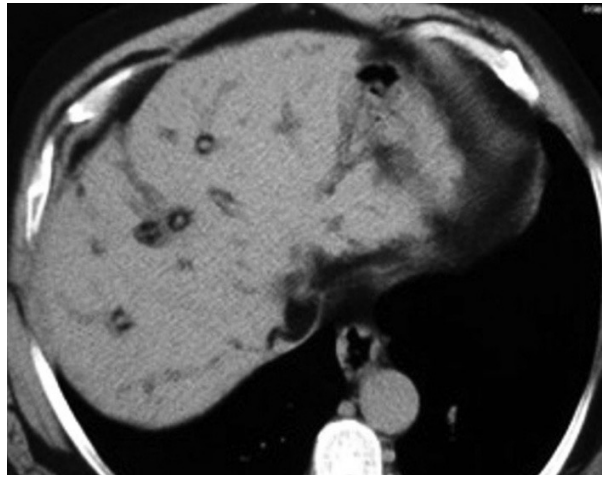


Image coutersy Manzella et al 'Schistosomiasis of the liver' DOI:10.1007/s00261-007-9329-7



Periportal fibrosis
Image courtesy https://liveratlas.org Case 164

Literature Review: Schistosomiasis gastrointestinal tract involvement (pathophysiology)

- The larvae, known as cercaiae, attach to the host's skin, typically the feet, and subsequently penetrate it.
- They develop a protective bi-layered tegument that shields them from the host's immune system. This transformation leads to the formation of schistosomula, resembling worm-like creatures, which travel through the venous circulation to the heart and then the lungs. There, they reside and undergo further maturation over a period of 2–3 weeks.
- As the worms continue to develop, they ultimately reach the left side of the heart. From there, arterial blood carries them to the small vessels of the portal hepatic circulation, where both female and male worms reach maturity. Mating occurs and the adult worms enter the vessels, eventually migrating against the portal flow to specific venous plexuses, such as the mesenteric venous plexus in S. mansoni and S. japonicum, and the perivesical plexus in S. haematobium. After leaving the male, the female deposits eggs in these locations. The adult worms have a lifespan of 3–5 years, although some reports suggest survival for up to 30 years.
- There is a predeliction for the inferior mesenteric vein which is why the rectum, sigmoid and descending colon are typically affected.
- The presence of eggs triggers an inflammatory response, leading to irregular thickening of the mucosa, the development of granulomatous polyps, and eventual fibrosis of the intestinal wall.

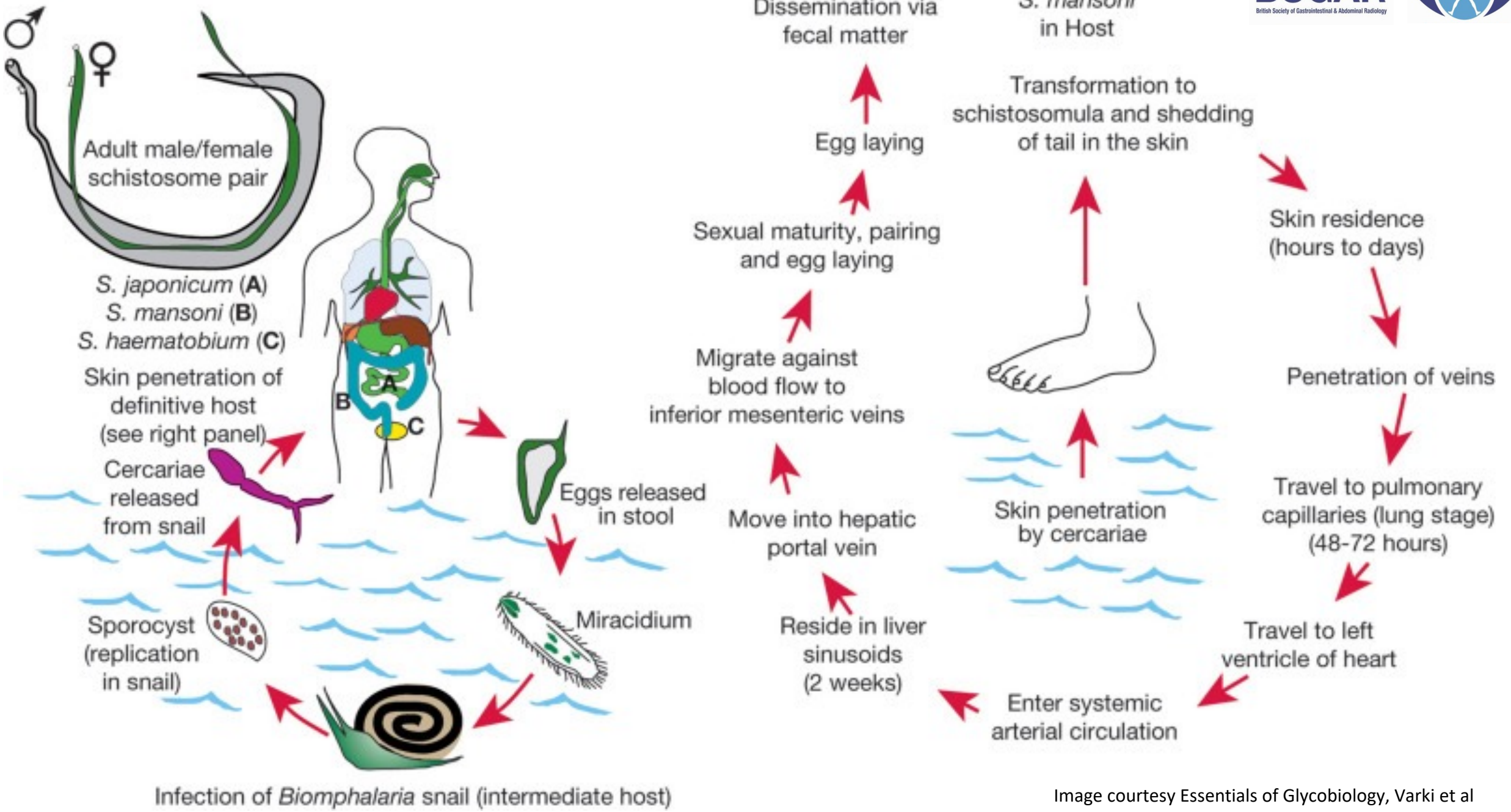


Image courtesy Essentials of Glycobiology, Varki et al

Literature Review: Schistosomiasis gastrointestinal tract involvement (prognosis and complications)

- Gastrointestinal manifestations of schistosomiasis include appendicitis, ulcers, mural thickening and calcification, colorectal polyps, mucosal ulceration and haemorrhoids
- Schistosomiasis associated appendicitis is prone to gangrene and perforation. Appendix masses are unusual. Chronic intestinal schistosomiasis is associated with an elevated risk of developing colon polyps, typically sessile and less than 10 mm. The polyposis itself is inflammatory in nature and does not predispose to colon cancer.
- Colonic calcification in curvilinear, tram-track or nodular distribution. Tram-track pattern is due to egg deposition and calcification in the parallel submucosa and subserosal layers.
- S. hematobium, the primary cause of urinary schistosomiasis, is carcinogenic in humans, leading to squamous cell carcinoma of the urinary bladder (a relatively uncommon form of bladder cancer). There is some limited evidence to suggest the potential carcinogenicity of S. japonicum, associated with colorectal cancer, as well as hepatocellular carcinoma formation. There is insufficient evidence regarding the carcinogenicity of S. mansoni in humans.
- Colorectal cancers linked to Schistosomiasis typically manifest as adenocarcinomas.
- Haemorrhoids may be linked to chronic inflammation induced by egg stimulation, exacerbating local venous reflux and impacting the structure of the anal cushion.

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Discussion and salient features of our case

There was significant diagnostic challenge initially in our case due to:

- The background of tuberculosis confounding interpretation of imaging findings.
- Difficulty in acquiring urinary tract samples due to the calcification of the urethra as well as urinary bladder.
- Schistosomiasis japonicum being a species native to the Philippines, unlike most other species that affect humans.
- Bowel wall calcification is unusual in Schistosomiasis, even in cases of hepato-intestinal schistosomiasis, unlike in urogenital Schistosomiasis.

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