a clinical problem

(the clinical case for this issue was prepared by victor cassar pullicino.)

A 45 year old nun was referred to hospital for investigation. She complained that for five years she had become progressively easily fatigued, with marked weakness and lassitude, to the extent that now she had marked difficulty in waking up in the morning and as it was continuous in nature it necessitated bed rest at various times throughout the day. These symptoms were enhanced and accompanied by a feeling of faintness after meals. She had lost her appetite with consequent loss of weight and complained of vague gastrointestinal upsets including occasional vomiting, abdominal pain and alternating episodes of constipation and diarrhoea. For the past year especially, she experienced an increasing difficulty in coping with the duties in convent due mainly to dyspnoea and a fainting feeling on moderate exertion and in fact on two occasions syncope during mass.

In the past she had an appendicectomy at 14 years of age and suffered from pulmonary T.B. when 25 years old for which she was subsubsequently given chemotherapy. She had been well after this but five years ago after a cholecystectomy operation she developed an increasing persistent tanning of her skin. She had found this strange because although her operation was in summer, she had had such a remarkably slow recovery after the operation that she had not gone out much and was not in the habit of sunbathing either.

On examination she looked ill, fatigued and slightly anaemic but was not cyanosed, dyspnoic or orthopnoic. There was no finger-clubbing or jaundice and no lymphadenopathy. She was generally pigmented especially in the neck, axilla, palmar creases and groin, with melanotic patches on the tongue and buccal mucosae. There was also a generalised loss of body hair especially marked in the axillary and pubic regions. She had a rapid small pulse, a blood-pressure of 90/60 and a temperature of 97°F.

The apex beat was impalpable, heart sounds were faint with an abrupt third heart sound but no murmur could be heard. Her cervical veins were distended with a paradoxical increase in the jugular venous pressure on inspiration. On inspection her abdomen was distended and there were two well-defined scars: a whitish appendcectomy scar which contrasted markedly with the highly pigmented cholescystectomy scar. There was a three finger hepatomegaly, a palpable spleen and ascites. There was also slight dependant oedema.

QUESTIONS:

1. What unusual dietary habit should be specifically asked for?
2. Mention some causes of mouth pigmention.
3. What is the importance of the splenomegaly?
4. What is your differential diagnosis?
5. Is the past history relevant?
6. What is the probable diagnosis?
7. What is so diagnostic of the different appearance of the two scars
8. Can you think of a possible explanation with regard to her symptoms being worsened in the morning and after meals?
9. What are the contributing factors to her episodes of syncope?
10. What two simple investigations may prove to be pathognomonic of her condition?

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answers to clinical problem

1. Salt Craving.
2. Physiological in certain races. Not in Euro-Epeans.
   Peutz Jegher's Syndrome.
   Hemochromatosis.
   Addison's Disease.
   Metallic Poisoning especially occupational i.e. lead, bismuth. arsenic.
   Fordyce's Disease.
3. In cases of high sustained venous pressure, as found in constrictive pericarditis, congestive splenomegaly may be sufficiently pronounced to make the spleen palpable. In the absence of evidence of bacterial endocarditis or of tricuspid valve disease, splenomegly in a patient with congestive heart failure, should arouse suspicion of constrictive pericarditis.
4. Addison's Disease.
   Malabsorption Syndrome.
   Cirrhosis of the liver.
   Infiltrative Diseases — Amyloidosis and Haemochromatosis.
   Constrictive pericarditis
   Tricuspid Stenosis.
   Renal Disease eg: Salt loosing Nephritis
   Cardiomyopathies.
5. The fact that the patient had a contact with T.B.
6. The probable diagnosis is a chronic adrenel insufficiency combined with constrictive pericarditis.
7. Scars that have been present before the onset of the adrenal failure remain unpigmented (appendicectomy scar) in contrast to those that are acquired after the onset of the adrenal failure (cholecystectomy scar).
8. An Addisonian is prone to hypoglycaemic symptoms due to increased insulin sensitivity (tiredness, lethargy, feeling of faintness) especially following extended fasting or following carbohydrate ingestion. Reactive hypoglycaemia after meals may occur, cortisol being one of the physiological antagonists of insulin. These patients are also especially hypoglycaemic after a night's rest and have more than usual difficulty in getting up in the morning.
9. Lethargy, tiredness, exertional dyspnoes and syncope are signs of contrictive pericarditis. However the lethargy and tiredness are more pronounced in this patient due to the superimposition of Addison's Disease. One C.V.S. effect of Addison's is hypotension. So although hypotension can probably give rise to a lessening of the signs of contrictive pericarditis c.g. venous distension, hepatomegaly, ascites, etc., the hypotension itself will predispose to and make the patient more susceptible to syncopal attacks.
10. A chest X-Ray may show calcification in the pericardium indicative of constructive pericarditis whilt a plain abdominal X-Ray may show calcification in the adrenal glands. Both these findings indicate a granulomatous process, in this case due to T.B.