Case report: microcardia secondary to chronic adrenocortical insufficiency

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A 53-year-old male presented with insomnia, loss of appetite and irritability, which seemed to worsen during the winter season. He could not say when these symptoms began and reported that he had simply "put up with them." There was no associated nausea, vomiting or abdominal pain, but he had recently developed a strong aversion to meat, such as roast beast, coupled, it seems, with a craving for popcorn, which he snatched up whenever the opportunity arose. He reported associated symptoms of "tightness" in the heart and swollen feet, and complained that his shoes have been "too tight" for the past several months.

The patient, a nonsmoker who had no previous medical history, was single and took no medications. He lived alone in a remote mountain cabin, where he relied on his dog, Rex,* for companionship. He was self-employed as a handyperson and busied himself with odd jobs such as repairing lights and transporting small household goods for his neighbours. When money was especially tight during the holiday season he was sometimes employed as a Santa Claus, who dressed up for children living in the valley below. Interestingly, the patient reported feeling much better after these seasonal excursions.

Physical examination: The patient was a tall, portly gentleman who was surprisingly agile on his feet and in no apparent distress. He had a mild stutter, demonstrating a slight tendency toward monosyllabic whole-word repetitions — "noise, noise, noise, noise, feast, feast, feast, feast, sing, sing, sing, sing." His skin appeared hyperpigmented, with a slightly greenish hue (Fig. 1). Examination of the head and neck revealed mild exophthalmos, pronounced micrognathia and a slight torticollis, which prompted the patient to complain that "his head wasn't screwed on quite right." There were no palpable lumps in the neck. On auscultation chest sounds were normal, but the cardiac examination revealed faint-to-barely-detectable heart sounds with a medially displaced cephalad apical beat in the second intercostal space. Palpation of the abdomen identified no masses or areas of tenderness. The hepatojugular reflex was not prolonged. Examination of the peripheries revealed cold extremities and excessively long fingernails. An absence of external genitalia was noted, but not discussed.

Laboratory and radiologic investigations were hampered by the absence of a radiologist or lab technician in our remote setting, but a local illustrator was able to provide a clear image of the heart, which could only be described as "2 sizes too small." This finding quickly led to a diagnosis of Addison's disease, with secondary congestive heart failure, and appropriate therapy was promptly initiated, with good results.

Commentary: Microcardia presents to physicians much more commonly as a symptom than a sign. Patients often complain of small-heartedness in themselves or others and in most cases physicians concur, although x-rays rarely do.[1] This case demonstrates a rare case of microcardia, secondary to chronic adrenocortical insufficiency. In less remote settings, suspicions of Addison's disease can be confirmed quickly with a laboratory test to demonstrate insufficient levels of plasma cortisol. Because of the absence of laboratory resources available in this case, a presumptive diagnosis was made on the basis of the classical triad of clinical findings: microcardia, hyperpigmentation and salt craving (the popcorn).[2]

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The patient was treated with daily cortisol and aldosterone replacement therapy and significant improvement in both symptoms and appearance were noted.[3] The curious relief of dyspnea, experienced by the patient after excursions down the mountain side, can be explained by the normopoxic environment of the village relative to the hypoxic setting usually experienced by the patient in his home on the mountaintop. Given the chronic course of primary adrenocortical insufficiency, the patient was advised to make regular excursions to this village. And he does this every Christmas.

*Name changed to protect patient confidentiality.

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Fig. 1: "It's too small."

References

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A *CMAJ* reviewer responds:

I have been given the opportunity to respond to the case report by Weir and Fleming involving a patient with a heart 2 sizes too small. I recently helped care for a patient who presented with identical symptoms, but unlike Weir I had the advantage of some sophisticated diagnostic equipment, including a cog-heavy diffendoppler (you have to watch your hands with this one) and a highly accurate glurksmirkometer, which measures to the nearest yert.

I was troubled by the classic triad of irritability, stuttering and paralytic sour frown, and had the patient assessed by the on-call entomologist. Serologic studies were positive for both spider and termite antibodies, likely due to infections in the brain and teeth, respectively. The patient was immediately put on an intravenous course of insecticide, with good results.

The "tightness" in the heart and the swollen feet called for a trip to the tickerpiccer. Imaging revealed a plum-tomato-sized heart covered with mouldy purple spots and, as suspected, atria and ventricles clogged with unwashed socks. The heart lesions were surgically débrided, and the socks were removed.

The patient did well postoperatively. In fact, his heart grew not 1, not 2, but 3 sizes that day.