gastrointestinal activity in myxoedema—from chronic constipation to the most severe ileus—has been reported. The milder forms are reversible with thyroid hormone replacement, but more severely affected patients die of other complications of hypothyroidism before treatment has had time to be effective. Only one other report[2] has attributed death to the ileus.

The pathological findings in our case suggest that the ileus was due to a peripheral neuropathy affecting the gut. Myxoedema is known to be associated with a polyneuropathy,[3] the changes in peripheral nerves suggesting that Schwann cell disease, possibly caused by mucopolysaccharide deposition, is partly or wholly responsible for the disorder. Although no mucopolysacchardie was found in this case, Byrom[4] showed that it disappears within days of thyroid hormone administration. The axons, however, were abnormal, showing a severe autonomic neuropathy affecting mainly the extrinsic nerves in the colon, but also suggesting intrinsic nerve involvement.

That peripheral neuropathy can affect the alimentary canal as well as the limbs is well recognised, as in diabetes mellitus.[5] Anatomical changes in bowel innervation are in fact common, but clinical symptoms are rare, probably because slight changes in the nerve supply to the limbs are enough to produce parasthesiae, but to produce a megacolon damage has to be almost complete. The pathological changes in this case suggest that the ileus might have been reversible with thyroid hormone replacement had the patient survived.

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Klinefelter's syndrome with hypogonadotrophic hypogonadism

Patients with Klinefelter's syndrome have raised circulating follicle stimulating hormone (FSH) levels. Luteinising hormone (LH) levels are usually increased also, but they may be within the normal range when the testosterone secretion rate approaches normal.[1] We describe here a patient who showed the clinical features of Klinefelter's syndrome associated with low testosterone levels, undetectable LH, and FSH levels that were not raised. When the patient's age is considered the FSH levels were actually below normal.[2]

Case report

A 70-year-old man with an eight-year history of diabetes mellitus presented with gangrene of the left foot. Conservative treatment was unsuccessful and a below-knee amputation was subsequently performed. On examination he was mildy obese and appeared hypogonadal with a high-pitched voice, a eunuchoidal habitus (span 187 cm, height 174 cm, and upper:lower segment ratio of 0·8:1), smooth skin, and fine, soft, facial hair. There was bilateral (2 × 2 cm) gynaecomastia, body hair was sparse, both testes were soft (1 × 0·5 cm), and his penis was 5 cm in length (stretched). Visual fields and sense of smell were unimpaired. There were signs of peripheral vascular disease in the right leg but no carotid bruits were present. The fundi were not seen owing to bilateral cataracts. On direct questioning he said that he had developed only a small amount of pubic and axillary hair in his teenage years and that he had never shaved. There was no family history of endocrinopathy.

A clinical diagnosis of Klinefelter's syndrome was confirmed by a positive buccal smear and a karyotype of 47,XXY from peripheral blood. The serum testosterone level was 1·04 nmol/l (0·29 ng/ml), serum LH <25 μg LER-907/1, and serum FSH 230 μg LER-907/1. A luteinising hormone-releasing hormone (LHRH) test showed normal FSH and LH responses (see table). A thyrotrophin-releasing hormone test (400 μg intravenously) resulted in normal thyrotrophin and prolactin responses. Serum cortisol was 640 nmol/l (23·2 μg/100 ml) (normal 200-660 nmol/l (7·0-23·9 μg/100 ml)). Radiographs of the sella turcica were normal, as was a brain scan.

No androgen replacement therapy was considered to be warranted, and after he recovered from the amputation the patient was discharged from hospital.

Comment

There are only two other reports describing patients with chromatin-positive Klinefelter's syndrome associated with low gonadotrophin levels. The first describes a 74-year-old man with XY/XXY mosaicism who had low circulating LH and testosterone levels. No comment was made about the FSH levels.[3] The second report describes an 18-year-old man with XO/XY/XXY mosaicism in whom the circulating FSH levels were undetectable. Serum testosterone levels were in the low normal range and serum LH levels varied between normal and high values.[4]

The cause of the bihormonal gonadotrophin deficiency in our patient is obscure and a congenital abnormality cannot be excluded. A pituitary tumour seems unlikely in view of the normal skull radiographs, visual fields, and provocative test results. Diabetes mellitus, which presumably predisposed to the peripheral vascular disease, could theoretically have affected the microvasculature of the hypothalamic-pituitary axis, but it is unlikely that this would have resulted in a "selective" gonadotrophin deficiency (although growth hormone reserve was not tested), particularly as the responses to LHRH were normal. Possibly our patient's condition represents a variant of Klinefelter's syndrome with a reduction in the release of LH and FSH occurring secondary to prolonged hypersecretion, as suggested by Rabinowitz *et al* in their case report mentioned above.[4]

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Responses to LHRH test (100 μg given intravenously)

Time (min):	−30	0	15	30	45	60	90	120	180	Normal values*
FSH (μg LER-907/1)	230	200	310	380	430	470	510	670	870	469±55
LH (μg LER-907/1)	<25	<25	53	79	101	119	170	190	210	97±66
Testosterone (nmol/l)	1·04								0·73	14·4±6·0

*Normal basal values (±SD) for men aged 70-79 years.[2]
Conversion: SI to traditional units—Testosterone: 1 nmol/l ≈ 0·288 ng/ml.