Part I—At Fifteen...

He was, his mother always said, the cutest little boy ever, and she had always adored him. So strong, so sturdy, confidently charging through life. At 10, he joined a Little League baseball team, and made the All-Star team in his first year. It wasn’t until quite some time later that she realized something was very wrong.

Looking back, probably the first symptom appeared when he was 11. Promoted up to the next division in Little League, he mostly sat on the bench. His coordination was not as good, and he seemed to have lost his hustle. Of course, it was probably just that he was being expected to do too much, and it was easy to overlook. And he began to grow.

Soon he began to notice girls, but after an initial girlfriend or two, he retreated. Too shy, probably. They moved, and moved again, and his grades slipped. He became more introverted. Troubled teen? Doing drugs? She found no evidence, but worried. And he grew.

He turned 15, and he grew. He was getting close to six feet tall now, and was wearing size 13 shoes. He was skinny, weighing only 150 lbs. His right knee began to swell, and he developed mysterious ulcerated sores on his lower left leg that refused to heal. He began to complain often of feeling sick, and came home from school frequently. Never anything seriously wrong, but he just didn’t feel good.

They revisited the doctor, who picked up on the description of large hands and feet and decided to check blood levels of some hormones. His thyroid was down, his cortisone levels low, his testosterone levels low, and his growth hormone levels high. Taps of his right knee removed up to 300 cc of fluid, but no evidence of anything specifically wrong inside his knee, just that it was being damaged. He looked gaunt and unhealthy. Then he fell and broke his left hip.

Questions

1. What hormonal problem could be causing these symptoms?
2. Why would joint damage be associated with rapid growth and low testosterone levels?
Part II—At Twenty-Five...

It was, Dr. Kidd thought, a most unusual case. The symptoms all indicated that a benign pituitary tumor had put pressure on the pituitary and disrupted its functions. It was an unusual condition, and this was the first case he had ever seen. The boy had been referred to a hospital in Madison, Wisconsin, where they used irradiation (triangulating from the left, the right, and the front) to kill the pituitary and any tumor associated with it. They also had to do a cartilage operation on his knee and put metal pins in both hips to hold the ball and socket jointing together.

The good news was that everything seemed to be working. The patient was 6'4" now and had put on muscle, so he now weighed over 220 lbs. X-rays of the skull showed that the bone saddle the pituitary rested in, which had been pitted, was now healing over, which indicated that the tumor was no longer putting pressure on the bone and therefore had probably been destroyed. His hormone levels had stabilized. Of course, he would need replacement hormones for the rest of his life, especially cortisone, thyroid, and testosterone, and they had used a growth hormone suppressant for a year, but overall Dr. Kidd was happy with his progress.

All of which led to Dr. Kidd’s present quandary. His patient, now 25, was sitting in his office and had just told Dr. Kidd that he was getting married. It had never occurred to Dr. Kidd to discuss it before, but normally patients with these syndromes were sterile, and Eric had little body hair and had told him he never shaved. What should Dr. Kidd do?

Questions

1. Should Dr. Kidd tell Eric that he is probably sterile? Why would he be sterile?
2. Is there anything that they could try to do to stimulate spermatogenesis? Why is the absence of facial hair important?
Part III—At Twenty-Eight...

He was 28 now, and growing a beard. They had injected human chorionic gonadotropin (hCG) for two years in an attempt to produce sperm, but it hadn’t worked. He was big, 6’6”, and weighed 275 lbs. His hands and feet were especially large; he wore 2x gloves and size 18 shoes.

Eric couldn’t wait to get the wires out of his mouth. His jaw had been hurting, especially the joint on the left side. The dentist thought he might have TMJ (temporal mandibular joint dysfunction), and referred him to a dental surgeon. The surgeon had expanded his upper jaw by splitting it into five pieces, pulling the pieces into place, and screwing in small metal plates to hold everything together. The wires were to let everything heal, and 10 weeks of a mouth wired shut had been more difficult than he had thought. However, he was looking forward to getting rid of the frequent headaches.

Questions

1. Why was he growing facial hair? Hint: hCG acts like FSH and LH, which are pituitary hormones that trigger a variety of sexual developments and secondary sexual characteristics, whereas testosterone is more of an end-level hormone.

2. Why did he need jaw surgery?
Dr. Lee sat down at her desk with a weary sigh and rubbed her stiff neck. Sometimes she wondered whether it was worth it all. She worked such long hours, and it was so hard to watch helplessly as a patient died. She’d just lost an old dear friend, and there wasn’t a thing she could do to stop the relentless spread of her disease.

She picked up a fat folder, full of lab tests, notes from office visits, and other documents. Ah yes, this was one of her challenging patients. The guy’s hormones were seriously whacked, and trying to keep all of them in balance plus dealing with his high blood pressure was a bit like juggling china plates while riding a unicycle. As if that wasn’t enough, he kept coming up with odd things. One morning his right eye stopped working, which led to the diagnosis of an aneurysm in an artery immediately beneath the pituitary. The aneurysm was probably due to radiation damage to the blood vessel walls in the area of the tumor. They had gone in and surgically wrapped it to hold it together, but his recovery had taken months. Fourteen months later his right eye began to work again, although he lost most of his peripheral vision. A stroke he had two years later may have been due to radiation or due to high blood pressure, but meant frustratingly long months of physical rehab. And of course, just to make things interesting, he had to have the stroke while he was in a jungle somewhere in Costa Rica. He came home from China testing positive for tuberculosis, from the Marshall Islands with beaver fever, and from Costa Rica with parasitic botflies burrowing under his skin. Botflies! Here in Illinois they only infected sheep. Maybe she was becoming a veterinarian.

Well, back to work. Let’s see...her patient was 45 years old, 6’6”, 310 lbs. At 5’2” she was small to begin with, but he always made her feel tiny. He had two cute adopted daughters. She smiled, thinking of his youngest, Mara, who had given her a rather enthusiastic hug the other day when Dr. Lee had been picking up her daughter Beth from the preschool both girls attended. One of the nice things about small town life was getting to know your patients, and Mara just loved Beth and tried to take care of her at their preschool.

Now Eric’s wife was complaining he wasn’t sleeping enough. Eventually they had figured out that his sleep was getting seriously disrupted by frequent nighttime trips to the bathroom. It’s quite normal for a 45-year-old male to make one nighttime visit and dump 350 cc of urine, but Dr. Lee had asked him to record the frequency of his visits and the quantity of urine produced. Her nurse had attached the chart he had just sent. She looked it over.
Yes! She mentally gave herself a high five. Finally she had a problem she could solve; this was what she spent all of those years of training to do. The diabetes insipidus was so obvious when you looked at this data. All she had to do was to write one prescription!

**Questions**

1. Why are his kidneys so active at night? Note: diabetes insipidus is *not* diabetes mellitus, so the answer is not high blood sugar levels or an insulin deficiency. Diabetes merely means high urine production. Look for a specific hormonal problem involving the pituitary.

2. It also turns out that he has unpredictable shock responses, so that a small cut needing three stitches left him pale and in shock while breaking his left wrist in at least 15 places did not. What hormone could be involved?

3. If the pituitary is so important, and if Eric is not getting replacements of hormones the pituitary makes (except for ADH), why isn’t he dead? To answer this question, think through the cascade of events from hypothalamus to effect.

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