Plexiform neurofibroma and demielinisant lesions in a patient with GH deficiency treated with rGH

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Introduction

There have been concerns regarding the safety of recombinant growth hormone (rGH) administration in children with GH deficiency in relation to cancer risk as several experimental studies have revealed a mitogenic and proliferative role for both GH and insulin-like growth factor 1 (IGF-1). A recent systematic review that evaluated the link between rGH therapy and cancer found that standardized incidence ratio (SIR) for cancer was 2.74 (95% CI 1.18-5.41), but the cancer standardized mortality ratio (SMR) was not increased and the issue of rGH therapy safety remains a matter of debate.

Case report

M.D., F, 13 years old

The patient was submitted in our endocrine department for follow-up of GH therapy.

Medical history

February 2009 (7.5 ys)

- H=107cm (-4.5SD); W=18 Kg; BMI=15.72 kg/m2;
- Tanner P1G1;
- Clonidine test- max GH= 4.1 ng/ml
- BA: **5.5** ys
- Cerebral CT: normal

Diagnosed with short stature due to GH deficiency- started treatment with rGH 0.035 mg/kgc/d

June 2010 (13 ys)

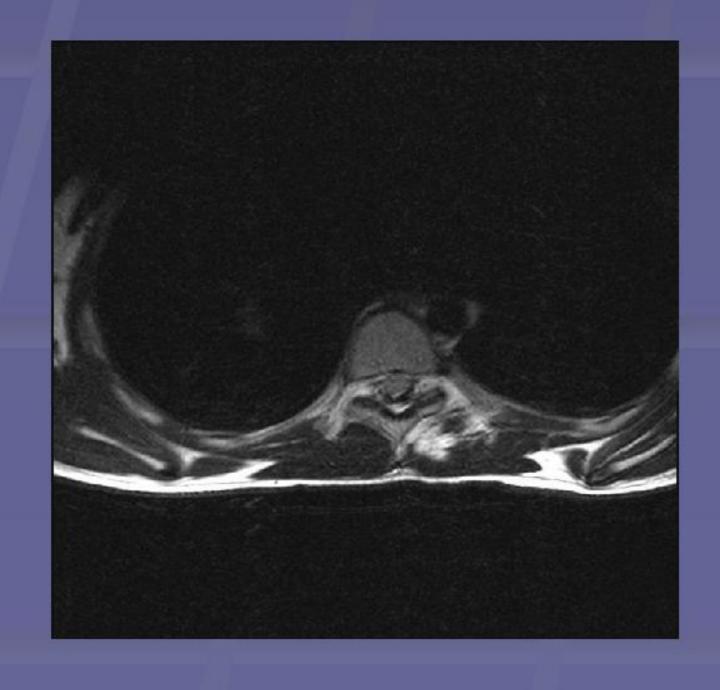
Physical exam

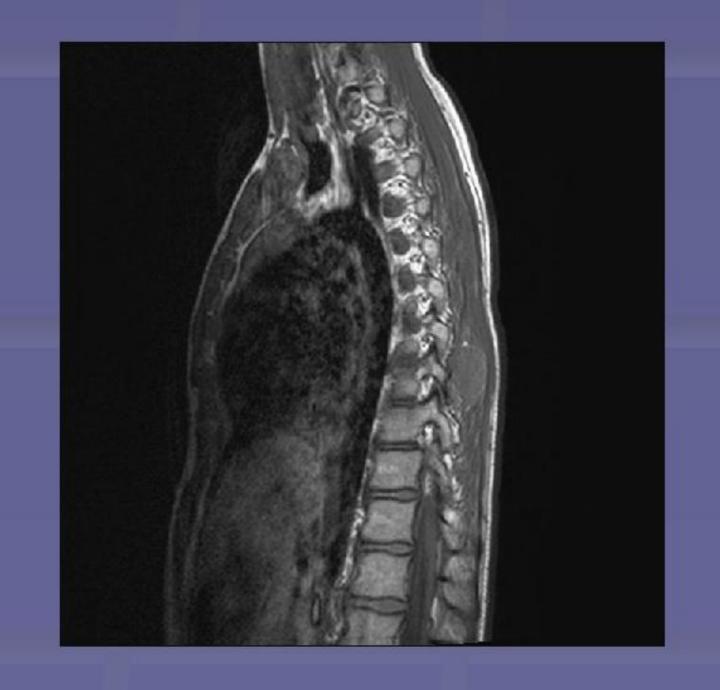
- H=149.9 cm (-0.74SD); W=39 Kg; BMI=17.35kg/m2;
- 96bpm, BP=90/70 mmHg;
- Tanner P3G3;
- subcutaneous left paravertebral tumor in the toracal region reported to be painful by the patient; the tumor had hard consistency, was mobile and had no sign of inflammation.

Laboratory tests

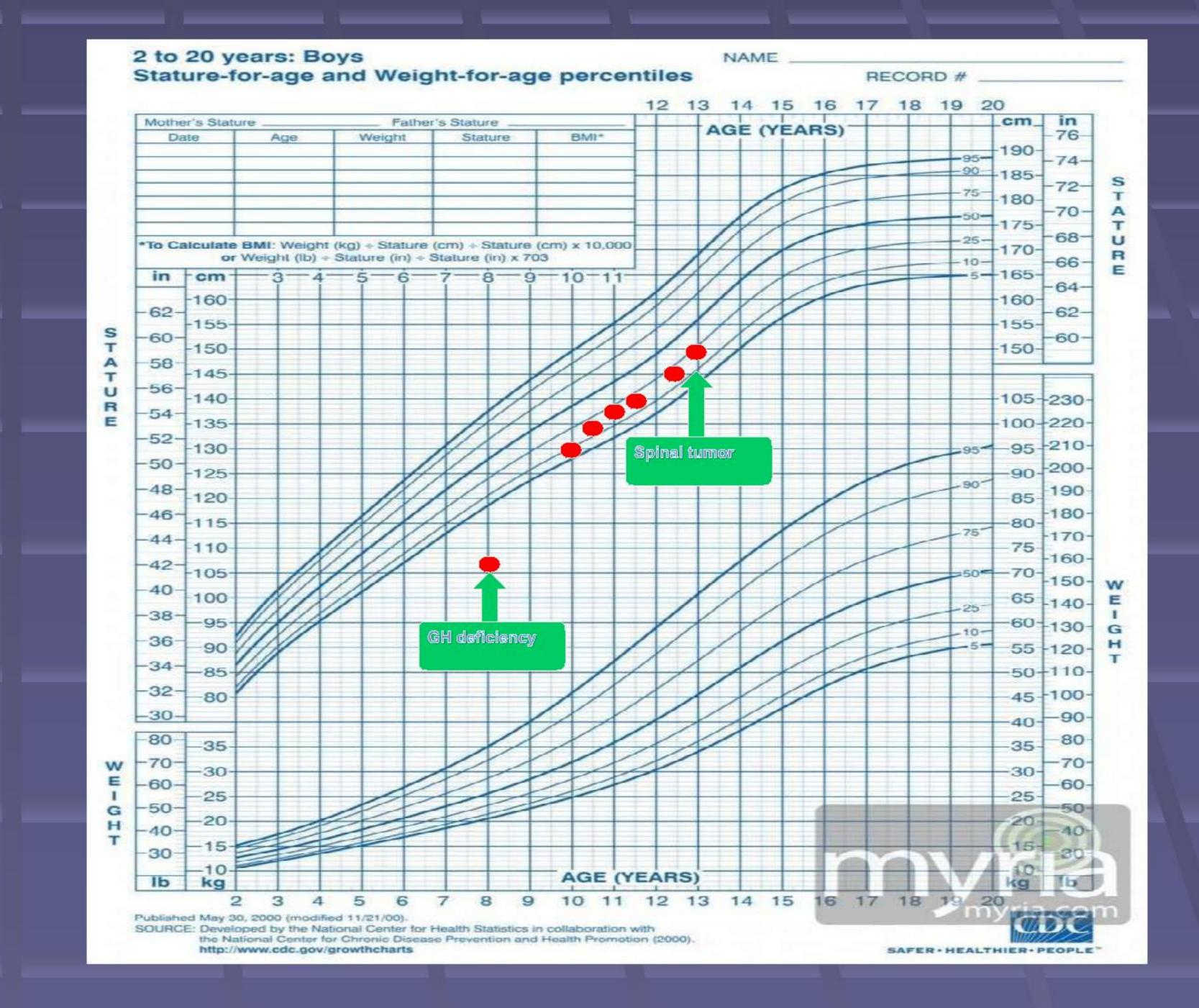
- IGF1 = 330 ng/ml (143-693 ng/ml)
- TSH=3.88 microIU/ml (0.4-4.4 microIU/ml);
- FT4= 1.05 ng/dl (0.89-1.76 ng/dl)
- BA = 13 ys;
- Spinal MRI left paramedian toracal tumor located from T7 to T9 with extension in the left VIIIth foramen that raised the suspicion for a toracal neurofibroma with intra and extraforamen extension.

Figure 1. Spinal MRI





- Sent to neurosurgery
- Biopsy revealed a plexiform neurofibroma



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Taking into account the patient's family complaint of child`s poor scholar performance and memory loss, a cerebral MRI was done revealing some demielinisant supratentorial lesions; consequently, the patient is currently under evaluation in the pediatric neurology department.

Spinal MRI showed no signs of restant tumor or relapse.

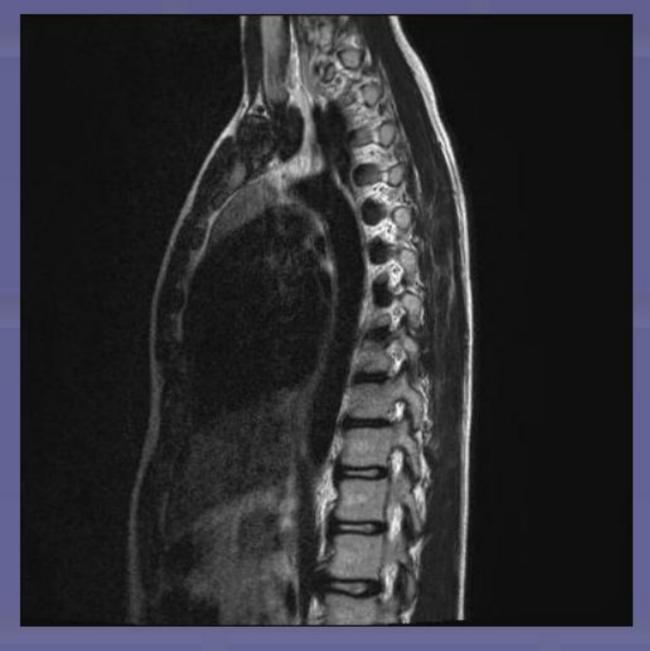


Figure 2. Check-up MRI

Particularity

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In this case, a keypoint resides in the fact that the tumor was discovered early due to the periodic follow-up for GH therapy but another issue might be the continuance of the GHtherapy.

As well it will be important to find out if the two conditions are related to each other and part of a syndromic disease.

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