

Historical Vignette

HV1(1-5)

The man with acromegaly who inspired the Shrek animated project.

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Abstract

Maurice Tillet was a professional wrestler, recognized as World Heavyweight Champion by the American Wrestling Association, who inspired the Shrek animated design, and developed the initial symptoms of acromegaly in his twenties. Maurice Tillet, known as “The French Angel”, was photographed by Irving Penn, an American photographer who paid great attention to details, known for fashion, portrait, and still life photography. The dysmorphic features detectable by the observation of these photographs are patently suggestive of acromegaly.

Key-Words: Acromegaly, Dysmorphic features, High stature, Hyperprognatism, Thickened lips, Frontal bossing.

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Acromegaly is a rare chronic, progressive endocrine disorder usually due to a growth hormone (GH)-secreting pituitary adenoma and characterized by progressive anatomical disfigurement, mainly involving face and extremities, and metabolic dysfunctions determined by increased circulating GH and insulin-like growth factor 1 (IGF-1) levels (1,2). A 43-year-old man photographed in 1946 by Irving Penn was the professional wrestler Maurice Tillet (1903-1954), known by the nickname “the French Angel” who inspired the Shrek animated design (**Figure 1**).

On August 1, 1944 “the French Angel” was recognized as World Heavyweight Champion by the American Wrestling Association defeating Steve ‘Crusher’ Casey. Two more photos were taken in 1945 by Irving Penn (**Figure 2 A-B**); in both of them Tillet was photographed with the American supermodel Dorian Leigh.

Figure1. Maurice Tillet at a photo shoot with Irving Penn, 1946. Lot Photographs, Irving Penn, 2 October 2012 New York.

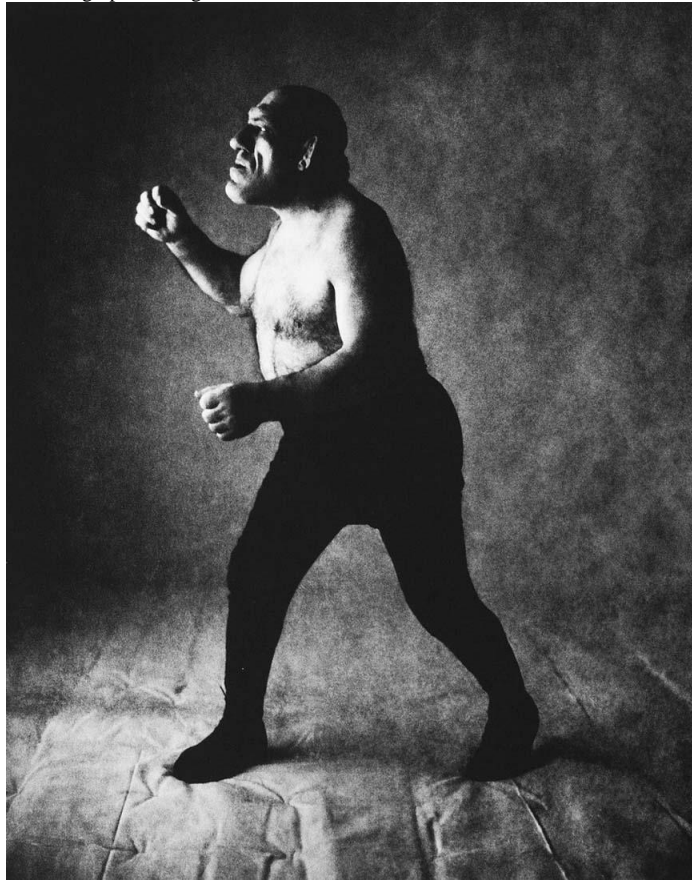


Figure 2 A-B. Maurice Tillet with the American supermodel Dorian Leigh, 1945 by Irving Penn pixels.

A



B



Irving Penn (1917-2009) was an American photographer who paid great attention to details, known for fashion, portrait, and still life photography, and who experimented with many printing techniques (3). Penn's career included work at Vogue magazine, and independent advertising work for clients as Issey Miyake and Clinique. His work has been exhibited worldwide and continues to inspire the art of photography.

Maurice Tillet was born in 1903, a completely average, healthy child, in the Ural Mountains, Russia, to French parents. He aspired to become an actor, spoke about 14 languages and was quite gifted in prose but at the age of 17 developed the initial symptoms of acromegaly.

Tillet was tall but not a giant, a condition in which GH excess develops in childhood before the closure of the epiphyses. However, he can be included in the subset of patients with very young-onset acromegaly who nowadays would warrant a genetic screening for germline mutations of the aryl hydrocarbon receptor interacting protein (AIP) gene, which are known to predispose the appearance of young-onset pituitary adenomas. Acromegaly occurs sporadically in the vast majority of patients but can also be part of a familial disease (familial isolated pituitary adenoma, or isolated familial somatotropinomas) and/or of a syndromic disease occurring concomitantly with other endocrine tumours, such as in MEN1, MEN4, Carney complex, McCune-Albright and SDHx-related pituitary adenomas(4). Nevertheless, we have no information on Tillet's family history regarding pituitary diseases or personal history of other endocrine abnormalities.

The dysmorphic features detectable by the observation of these photographs are patently suggestive of acromegaly. In particular, high stature, frontal bossing, thickened lips, nose enlargement, kyphosis, jaw thickening with hyperprognathism and dental diastases, hands and feet enlargement with thickened and stubby fingers, can all be clearly observed in these and in several other photographs of Tillet.

Acromegaly is characterized by insidious onset and slow progression and is often diagnosed five to more than ten years after onset. In the case of Tillet diagnosis was delayed and his body, his face in particular, were disfigured significantly. Unable to endure the constant derision and humiliation, Tillet left France. Looking for a new identity to fit his disfigurement, Tillet moved to the United States where he made a living by becoming a professional wrestler, and where he was nicknamed the "freak ogre of the ring". His "villain character" was an instant success with the crowds, becoming one of the largest draws in professional wrestling, as in the case of the boxer world heavyweight champion Primo Carnera(5), but his career as a professional wrestler was substantially influenced by acromegaly, so much so that in the second half of the 1940s he had a notable decline and lost several matches, becoming less requested. On the other hand, GH excess is

associated with severe comorbidities with disabling consequences, including cardiovascular, metabolic, respiratory, neoplastic and musculoskeletal complications, which significantly impact on patients' quality of life and mortality risk (1,2). Moreover, the potential progressive expansion of a GH-secreting pituitary tumour left untreated can lead to severe visual field defects, as well as to other pituitary hormone secretion deficiencies which could have also negatively impacted on Tillet's life and career. However, no information is available regarding the management of the disease. In this regard, it is worth mentioning that the first successful pituitary tumour surgical removals in acromegaly patients were performed just in the first decades of the twentieth century, a time when radiotherapy was the first choice treatment for this condition and when medical therapies, such as dopamine agonists, somatostatin analogues and GH receptor antagonist, were not yet available (6).

Tillet returned to France where he died prematurely at the age of 51 from heart disease, a well-known complication of active acromegaly, on April 4th 1954 (7).

Conflicts of Interest: There is no potential conflict of interest, and the author has nothing to disclose. This work was not supported by any grant.

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Received August 1, 2018, accepted August 3, 2018, published on line Nov 11, 2019